Section: Rheumatology & Immunology

Case Series

A CASE SERIES OF VASCULITIS MIMICS FROM A TERTIARY CARE CENTRE IN KERALA

Sreedevi Suresh[1], Sunija B[2], Deepthi S V[2], Jacob Antony[3], Srikantan S[3], Sreepriya[4], Yamuna Pillai[5], Sreena Sreekumar T[2]

[1]Assistant Professor, Division of Rheumatology, Government Medical College, Thiruvananthapuram, Kerala, India
[2]Assistant Professor, Department of Internal Medicine, Government Medical College, Thiruvananthapuram, Kerala, India
[3]Professor, Department of Internal Medicine, Government Medical College, Thiruvananthapuram, Kerala, India
[4]Associate Professor, Department of Radiodiagnosis, Government Medical College, Thiruvananthapuram, Kerala, India
[5]Assistant Professor, Department of Medical Gastroenterology, Government Medical College, Thiruvananthapuram, Kerala, India

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Corresponding Author:
Dr.Sreedevi Suresh
Email: drsreedevi.vishnu@gmail.com

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ABSTRACT

Vasculitis is a group of challenging conditions grouped under various calibre of vessels involved i.e. large, medium, small and variable vessel vasculitis. Vasculitis mimics is another entity that shares common clinical features involving multiple or specific organs. Hence accurate knowledge of both entities is essential to order for appropriate set of investigations that helps to identify both. Differentiating between vasculitides and its mimics is very crucial for choosing the right therapeutics and thereby avoiding long term immunosuppression. Here we discuss five uncommon vasculitis mimics according to vessel size. Large vessel vasculitis mimic described here is congenital Middle Aortic Syndrome in a child who presented with young onset hypertension. The medium vessel vasculitis mimics - Segmental Arterial Mediolysis and Fibromuscular Dysplasia depicted here had multiple visceral aneurysms as seen in other medium vessel vasculitides. Symmetrical Peripheral Gangrene mimicked medium vessel vasculitis, and the imaging showed Atherosclerosis. Meningococcal infection which had cutaneous manifestations mimicked small vessel vasculitis.

INTRODUCTION

Vasculitis is a group of diseases with heterogenous manifestations which are due to inflammation and fibrinoid necrosis in the blood vessels thereby causing ischemia and leading to end organ damage.[1] According to the 2012 International Chapel Hill Consensus Conference, vasculitis has been classified according to the type of vessel involved as large, medium, small and other types including single organ, variable vasculitis secondary to other autoimmune conditions and infection induced vasculitides.[2] Vasculitis should be diagnosed promptly, and optimum management must be instituted to avoid organ failure, simultaneous work up for mimics is equally warranted to prevent unwanted exposure to harmful immunosuppressive drugs.[3,4]

Vasculitis mimics: Mimics of vasculitis include infections, malignancy, thromboses and non-inflammatory conditions like vasculopathies.[4] Infectious process can simulate vasculitis of any vessel size. Vasculopathies usually mimic large and medium vessel vasculitis.[5] Large vessel vasculitis most commonly manifest as claudication, symptoms of accelerated hypertension, unequal limb pulses, absent limb pulses, stroke, organ ischemia & radiologically as arterial abnormalities including stenosis, aneurysms, dissections & rupture.[6] Medium vessel vasculitis usually presents as refractory young onset hypertension, arterial aneurysms, stenosis, dissection, renal infarctions, stroke, transient ischemic attacks & gangrene.[6] Small vessel vasculitis has multitudes of cutaneous manifestations like ulcer, gangrene, petechiae and purpura.[7] Vasculitis mimics can have similar presentations which makes distinction from true vasculitis difficult, hence awareness of these entities are also equally important to make a favourable decision regarding diagnosis and therapeutics.[8]

In this case series we describe 5 cases that initially were considered as vasculitis by their clinical features. But later they were proven to be vasculitis mimickers by adequate clinical, imaging and

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laboratory evidence. Case 1 is an Idiopathic Middle Aortic Syndrome (MAS) which closely resembles Takayasu arteritis, a large vessel vasculitis, as the child presented with young onset hypertension. Case 2 and 3 are Fibromuscular Dysplasia (FMD) and Segmental Arterial Mediolysis (SAM) which simulated medium vessel vasculitis namely Poly Arteritis Nodosa (PAN) due to the presence of aneurysms in them. Cases 4 and 5 are that of symmetrical peripheral gangrene due to ischemic dilated cardiomyopathy and meningococcal infection which imitated medium and small vessel vasculitis due to development of digital gangrene.

Case 1

11-year-old girl presented with abdominal pain, claudication and giddiness for 6 months. She did not give any history of fever, dysuria, headache, weakness of limbs, dyspnoea, chest pain, decreased urinary output, vomiting or altered bowel habits. On examination, pulses were diminished in the lower limbs. She was detected to have hypertension of 160/90 mm of Hg in the right upper limb with a systolic pressure difference of more than 10 mm of Hg between both upper limbs. Apical impulse was felt down and out to the 5th left intercostal space. A systolic murmur was present in the left sternal border & interscapular area. Initial blood investigations were normal except for raised ESR (50mm/hr) and CRP (23mg/l). Takayasu arteritis being the most common large vessel vasculitis in young females was suspected owing to young onset hypertension with unequal limb pulses, hypertension and decreased pulses in the lower limbs with raised ESR and CRP. Ultrasound abdomen and pelvis showed haematocolpos & after drainage inflammatory markers started to fall and became normal. A detailed work up for causes of hypertension in young was done & echocardiogram showed left ventricular hypertrophy. CT aortogram [**Fig.1a-f**] showed coarctation of descending thoracic aorta, narrowed abdominal aorta, bilateral renal artery stenosis and multiple collaterals which were in favour of middle aortic syndrome. There were no clinical signs of genetic syndromes like café au lait spots or neurofibromas. PET CT scan was done to assess the vascular uptake in vessels, but didn't pick up activity in any of the vessels. Hence Takayasu arteritis was less likely and the inflammatory markers reverting to normal after drainage of hematocolpos was also against a diagnosis of vasculitis. Hence a diagnosis of Idiopathic MAS was made.

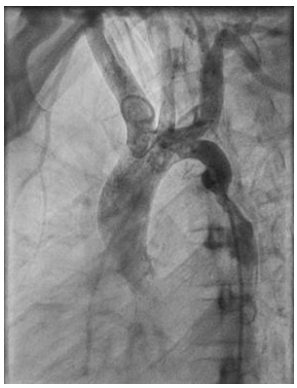

Fig. 1a

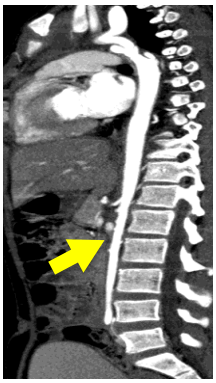

Fig. 1b

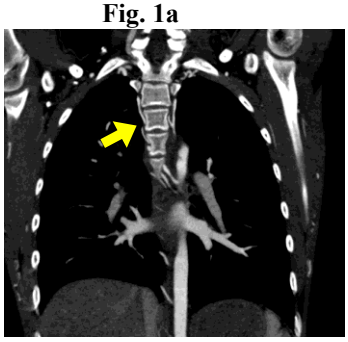

Fig. 1c

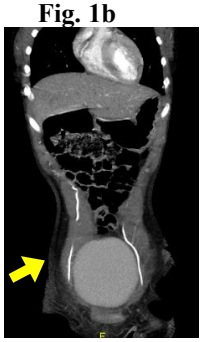

Fig. 1d

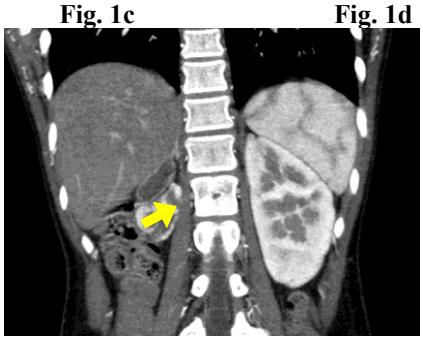

Fig. 1e

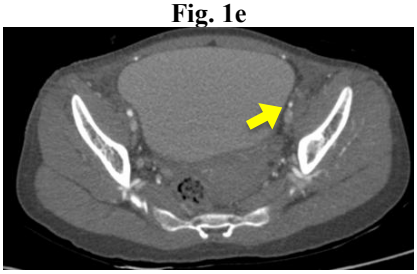

Fig. 1f

Fig. 1a - pre coarctation; **1b** - arrow shows narrowed abdominal aorta; **1c&d** - arrows showing collateral vessels; **1e** – arrow showing renal artery stenosis; **1f** - arrow shows narrowed iliac vessel

Case 2

31-year-old male with no prior comorbidities presented with gradually progressive dyspnoea and abdominal pain for 3 months. There were no swelling of legs, orthopnoea, paroxysmal nocturnal dyspnoea, chest pain, palpitation or cough. He denied any history of fever, vomiting or dysuria. On admission pulse rate was 90 /min with right upper limb BP being 190/110. All peripheral pulses were well felt equally. Bilateral renal bruit was present. JVP was elevated. He had pallor. Chest examination showed fine basal crepitations, so a provisional diagnosis of acute

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pulmonary oedema with accelerated hypertension was made. Work up with vasculitis as one of the differentials for young onset hypertension was done. He denied joint pain, oral ulcer, hair loss, photosensitivity, neurological deficit or gangrene. Hemogram showed anaemia of (Hb-9 gm/dl) with ESR 20 mm/hr. Liver function tests & electrolytes were normal. He had Urea/Creatinine of 73/4.3 with a Urine P:C ratio of 1.3. Echocardiogram showed no regional wall motion abnormality, but had mild mitral valve prolapse. Ultrasound abdomen showed hyperechoic kidneys with maintained corticomedullary differentiation. Renal artery doppler showed rising broad based systolic peak in bilateral main hilar arteries & reduced peak systolic velocity in bilateral main hilar and segmental arteries. Further CT renal angiography [**Fig 2**] revealed bilateral renal artery extrarenal aneurysmal dilatation with homogenous wall thickening which had a short segment diffuse distal arterial narrowing. Approximately 3 cm from origin from aorta, Right Renal Artery has a fusiform dilatation of 3 cm with a calibre of 2 cm, distal to which it divides into 2 branches outside renal parenchyma before entering it. There was circumferential homogenous thickening of renal artery and its branches measuring 4.1 mm with a significant progressive narrowing as the renal artery further courses and divides. Segmental branches not well visualized. Right kidney showed delayed homogenously reduced enhancement pattern. Left renal artery showed mild narrowing 2.3 cm from origin and then circumferential thickening same as the right. The left kidney had normal size position axis but delayed homogenously reduced enhancement pattern. Since there were renal artery aneurysms one of the differentials considered was Polyarteritis Nodosa (PAN), but distal location of aneurysms with more stenotic lesions in both renal arteries & normal inflammatory markers led to a final diagnosis of Fibromuscular dysplasia (FMD). Visceral artery aneurysms are found in ANCA associated vasculitis also, but in the absence of ANCA (Anti neutrophilic cytoplasmic antibodies) & other systemic features, it was less likely. The APLA (Antiphospholipid antibody) & ANA (Antinuclear antibody) tests were negative. Renal biopsy showed thrombotic microangiopathy. He is being managed with multiple anti-hypertensives and is being planned for renal artery stenting.

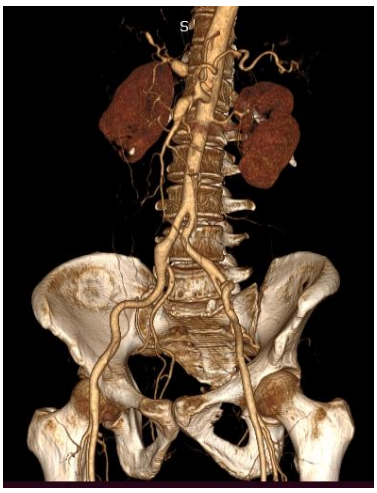

Fig 2: CT angiogram showing bilateral renal artery extrarenal aneurysmal dilatation with homogenous wall thickening which had a short segment diffuse distal arterial narrowing. Right renal artery has a fusiform dilatation of 3 cm from origin from aorta, with significant progressive narrowing. Left renal artery showed mild narrowing 2.3 cm from origin and then circumferential thickening same as the right. It has a beaded narrow constricted segment followed by fusiform dilation, largest dilated segment 7.1 mm.

Case 3

46-year-old male presented with 1 week history of postprandial abdominal pain in the epigastric region with no radiation & it aggravated on the day of hospitalisation. There was no history of chest pain, palpitation, sweating, fever, loose stools, vomiting, loss of appetite or weight. No events of skin rash, constipation/obstipation, or jaundice were reported. He was a teetotaller. On examination he was conscious & oriented with all peripheral pulses palpable equally bilaterally. BP was 110/80 mm of Hg in right upper limb and pulse rate was 80/min. No features of hypermobility/organomegaly/bruit/mass were present. There was epigastric tenderness. Chest was clear. All the basic blood investigations, amylase, lipase & Ultrasound abdomen were normal. ESR and CRP were 34 mm/ hr and 13 ng/ml respectively. Since there was history of bleeding per rectum, sigmoidoscopy was done which showed rectal erosions, circumferential ulcers, pseudo polyps & mucosal bleed in ascending colon, transverse colon and erosions in hepatic flexure, with an impression of probable right sided ischemic colitis. CT Angiography [**Fig 3**] was done to look for any vascular causes which revealed fusiform dilatation of common hepatic artery measuring 10x16 mm. SMA (Superior Mesenteric Artery) before branching showed fusiform dilatation with an intimal flap of 13x12 mm & a true lumen of 9.5 mm, suggestive of dissection. Right renal artery showed a saccular aneurysm 5.4x4.2mm just after its origin. There was circumferential symmetric short segment bowel wall thickening in ascending colon with mild surrounding fat stranding - possibly due to inflammation. Autoimmune work up showed negative ANA &

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ANCA. Since the patient was a middle-aged male with aneurysms in mesenteric vessels, a possibility of PAN was considered. The overall clinical picture, including acute presentation in the absence of other systemic manifestations, markers of inflammation being not so elevated and imaging findings that had predominantly dissections rather than micro-aneurysms and stenosis in mesenteric vessels - led to a differential diagnosis of Segmental Arterial Medio lysis (SAM). He was stabilized using symptomatic measures with amelioration of complaints with the plan to undergo embolization if symptoms recur since often the turn of events can be fatal due to massive uncontrollable bleed and hemodynamic instability.

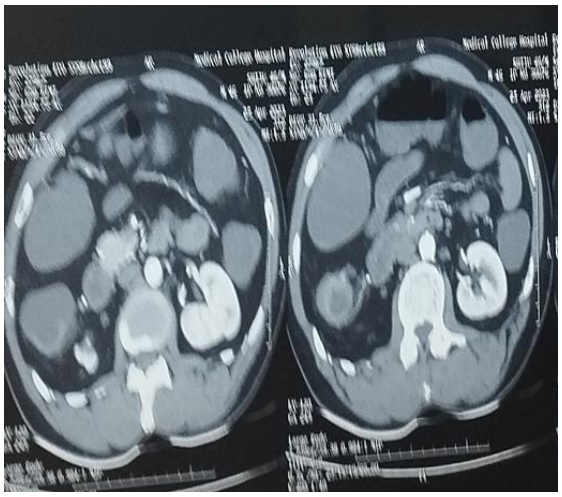

Fig 3: Fusiform dilatation of common hepatic artery measuring 10x16 mm. SMA before branching showed fusiform dilatation with an intimal flap of 13x12 mm & a true lumen of 9.5 mm, suggestive of dissection. Right renal artery showed a saccular aneurysm 5.4x4.2mm just after its origin.

Case 4

40-year-old male came with history of fever lasting for 10 days 2 months back. 1 month back he developed productive cough, occasional epistaxis and hemoptysis, dyspnea, orthopnea and paroxysmal nocturnal dyspnea & had gradual swelling of legs for 2 weeks. With these symptoms he was hospitalized. He had hypotension for which ionotropic support was given. Within a day he developed discoloration of tips of all toes of right foot and second toe of left foot, which was initially reddish, but progressed to become black in 4 days [**Fig 4a**]. With this history he was referred to our Centre since he couldn't continue the treatment there due to financial constraints. There was no history of testicular pain, hematuria, hematemesis, ear discharge, hallucination, delusion or eye redness. He was detected to have hypertension for 2 years & was on irregular medications. He was a chain smoker and alcoholic. O/E- pulse rate-102/min, all the peripheral pulses were felt equally. BP in the right upper limb-160/102 mm of Hg. Respiratory rate, SpO2 & temperature were normal. There was clubbing of fingers & bilateral pitting pedal oedema with gangrene of all toes on right foot and 2nd toe of left foot with desquamation of skin of both feet. He had left parasternal heave with a down and out apical impulse beyond the left 5th midclavicular line. Hemogram, LFT & electrolytes were normal. Creatinine was 2.3. Troponin I was 260, NT pro BNP was 17800 pg/ml (normal value-<100pg/ml). ESR and CRP were raised, 45 and 32 respectively. Hypertensive heart disease with heart failure and left ventricular systolic dysfunction was the provisional diagnosis. Since he was a young male with hypertension, epistaxis & gangrene, primary medium to small vessel vasculitis (like polyarteritis nodosa and cryoglobulinemic vasculitis) or secondary vasculitis secondary to connective tissue diseases were thought of as differentials. ANA, APLA& ANCA were negative. Echocardiogram showed dilated left ventricle, regional wall motion abnormality with hypokinesia of mid and basal inferior, anteroseptal, inferolateral, apical & inferior septal segment with moderate systolic dysfunction, grade 2 diastolic dysfunction, mild mitral regurgitation with ejection fraction of 32%, severe pulmonary artery hypertension and severe tricuspid regurgitation. Venous and arterial dopplers of both lower limb vessels (to look for thrombotic causes as occlusion) were normal. Ultrasound abdomen showed mildly hyperechoic kidneys, bilateral pleural effusion and ascites. CT angiogram of lower limbs [**Fig 4b**] showed atherosclerotic wall thickening with circumferential intermittent calcification of infra-renal abdominal aorta causing 35% luminal narrowing. IMA (Inferior Mesenteric Artery) at origin had ostial narrowing & 50 % luminal narrowing with a complete normal contrast opacification of rest of the IMA. He had symmetrical peripheral gangrene (SPG) of lower limb toes - causes of which includes infective, autoimmune, cardiac and vasoconstrictor drugs. He was diagnosed with symmetrical peripheral gangrene due to LV dysfunction (due to ischemic cardiomyopathy) as other causes of vasculitis and infections were ruled out. Chronic smoking and alcoholism, with history of vasopressor use in the previous hospitalization record, low cardiac output due to left ventricular dysfunction with atherosclerosis of infrarenal abdominal aorta vessels which would have reduced the blood supply to acral parts- might have been the triggers of gangrene. Gangrene is often not seen in large vessel vasculitis or atherosclerosis as collaterals are formed that provide adequate blood supply to the limbs. He was managed with diuretics, anti-anginal drugs, antiplatelets, anticoagulation, statins and anti-hypertensives.

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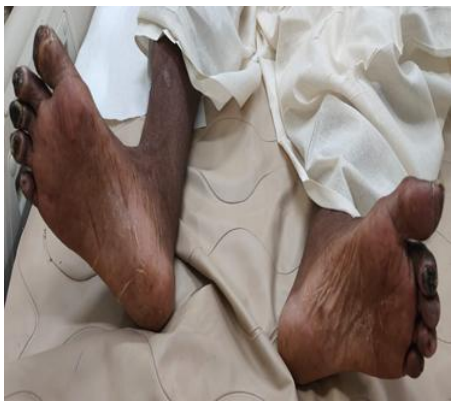

Fig 4a - Gangrene of toes

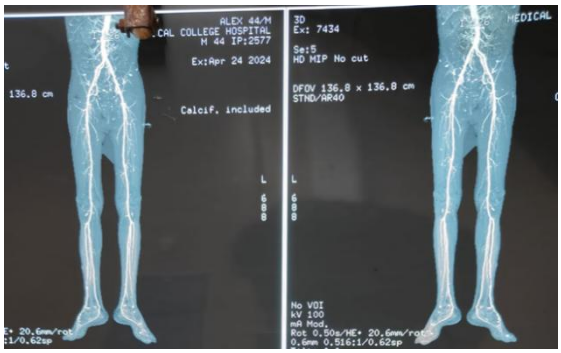

Fig 4b - CT angiography of both lower limbs showing atherosclerotic wall thickening with circumferential intermittent calcification of infra-renal abdominal aorta causing 35% luminal narrowing. IMA at origin had ostial narrowing with 50 % luminal narrowing with a complete normal contrast opacification in the rest.

Case 5

28-year-old male with no prior comorbidities presented with high grade fever for 5 days, breathlessness for 4 days, bluish discoloration of extremities and altered behaviour for 3 days. On receiving in emergency department, his pulse rate was 100/', BP in right upper limb was 128/80 mm of Hg. He was febrile with a temperature of 101^0 F. Dry gangrene of both hands and feet was present (**Fig. 5a,b**). All peripheral pulses of both upper and lower limbs were palpable and equal. Patient was drowsy with signs of meningeal irritation & bilateral extensor plantar reflex. Pupils were equal and reactive to light. Rest of the systemic examinations were normal. Xray chest showed bilateral pleural effusion. Since it was a young male with fever, meningeal signs and gangrene, other than infectious causes, medium to small vessel vasculitis was also thought of as a differential. Total counts were high (13,000 mm^3), rest all investigations were normal. Immunological work up with ANA, ANCA, APLA & cryoglobulins were negative. Blood culture grew Neisseria meningitidis. He was treated with antibiotics & thereafter the gangrene didn't progress. So here symmetrical peripheral gangrene was due to meningococcal sepsis.

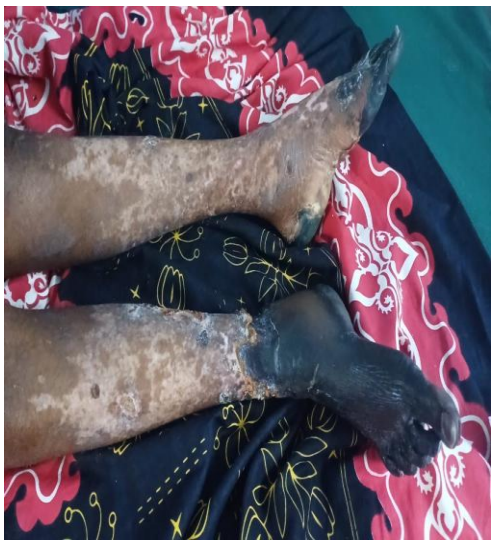

Fig 5a - Gangrene of feet

Fig 5b - Gangrene of fingers

DISCUSSION

Case 1 discusses the salient differences between idiopathic MAS and Takayasu's arteritis (TA) [**Table 1**]. Most of the cases of MAS show segmental or diffuse narrowing of the abdominal and/or distal descending thoracic aorta, with varying involvement of renal and visceral branches. It is implicated as one of the causes of hypertension in children and adolescents.[9,10] TA primarily occurs in individuals under the age of forty.[3-5] Patients may initially present with nonspecific complaints like malaise, arthralgia, weight loss, and fever & later with symptoms of ischemic complications. Hypertension is one of the common symptoms in both entities. Refractory hypertension can manifest as stroke, hypertensive encephalopathy, congestive heart failure & end stage renal disease. In MAS femoral pulses may be absent or diminished. Systolic murmur, abdominal bruit or other audible bruits may be heard due to extensive enlarged collateral formation. Other specific signs such as café au lait spots in patients with neurofibromatosis or

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hypercalcemia and Elfin facies in Williams syndrome should be searched for in any case of MAS. In TA, subclavian artery narrowing is a common anomaly leading to limb claudication, diminished pulses, and inconsistent blood pressure measurements between the upper extremities whereas vertebral arteritis can manifest with a range of neurological symptoms. Coronary ostial involvement can lead to fatal myocardial infarction.[6,7] Idiopathic MAS almost always involves abdominal aorta in the perirenal segment with ostial predilection of visceral arteries. TA also involves proximal ostial portion of arteries.[21]

MAS was initially described by-Sen et al. [11]The causes can be congenital or acquired. It is postulated to be due to defective fusion of embryonic aorta.[12] Congenital causes are congenital rubella syndrome, neurofibromatosis, mucopolysaccharidosis, genetic syndromes like Williams syndrome, Turner syndrome, and Alagille syndrome. [13-16] One of the acquired causes of MAS includes TA. Rumman et al. in his systemic review reported 630 cases of MAS in children of which 63% were idiopathic,15% mendelian,17% were due to inflammatory disorders.[17] The various vessel involvement and predilection in MAS & TA is summarized in **Table 2**. More cerebrovascular, carotid and subclavian involvement with more prevalence of aneurysms in TA helps us to distinguish between idiopathic and inflammatory causes. Surgical techniques or endovascular procedures are carried out to control hypertension in idiopathic MAS.[23] High-dose glucocorticoids, effective in inducing remission along with steroid sparing agents are used in TA. Revascularization through surgical procedures or endovascular interventions, such as balloon angioplasty, stenting, and stent graft replacement, may also be attempted during the period of remission which may improve the outcome.

Table 1: Idiopathic MAS v/s TA

	Idiopathic Middle aortic syndrome	TA
Etiology	Idiopathic, congenital, genetic syndrome due to inappropriate fusion of dorsal aorta	Granulomatous inflammatory vasculitis which starts from adventitia and media of large vessels and results in intimal hyperplasia and stenosis
Age	Pediatric population	Young adults
Symptoms[22]	Claudication, refractory hypertension, CCF, renal failure, leg weakness, absent or weak lower limb pulses	Claudication, Refractory hypertension, CVA, CAD
Systemic symptoms	-	+/-
Examination	Systolic murmur with abdominal bruit	Aortic regurgitation murmur, bruit in carotid, subclavian & renal artery
Vessel distribution	AA> Renal artery>SMA>Coeliac > DTA	SCA>Thoracic Aorta>CCA> AOA > AA>Renal artery
PET CT scan	No vessel wall uptake	Vascular uptake in affected vascular territories
CT aortogram	Narrowing of abdominal aorta distal to arch with visceral vessel & renal artery stenosis, multiple collaterals with no enhancement of aortic wall	Circumferential thickening of aortic wall with enhancement of wall, vessel wall oedema
Treatment	Anti-hypertensives, surgical measures	Steroids & other immunosuppressants, antihypertensives, surgical measures once disease is in remission

CCF- Congestive Cardiac Failure, CVA- Cerebrovascular accident, CAD- Coronary Artery Disease, AA- Abdominal aorta, SMA- Superior Mesenteric Artery, DTA- Descending thoracic aorta, SCA- Subclavian artery, CCA- Common carotid artery, AOA- Arch of aorta, RAS- Renal Artery Stenosis

Table 2: Vessel involvement in Idiopathic MAS & Takayasu's arteritis [18,19,20]

Vessel involved	Idiopathic MAS	TA
DTA[18]	8%	10%
Abd. Aorta[19]	98%	90%
1.Suprarenal	29%	13%
2.Aneurysm	5%	25%
3.Unspecified	35%	70%
Bilateral RAS[20]	62%	51%
SMA	27%	26%
Coeliac	20%	19%
Common iliac	1%	1.9%
Cerebrovascular	2%	6.7%
Subclavian	3%	13%
Carotid	1.5%	8.6%

Case 2 shows a young male with FMD who presented with hypertensive emergency for which PAN and TA are differential from vasculitis point of view. He had bilateral renal artery aneurysms with stenosis. Aneurysms are described in large vessel vasculitis like Giant cell arteritis (GCA), TA, medium vessel vasculitis like PAN, Kawasaki disease, small vessel vasculitis like ANCA(Anti neutrophilic cytoplasmic antibody) associated vasculitis and SLE (Systemic Lupus Erythematosus).[8] TA is more commonly reported in young women and has predilection toward the juxta ostial portion of common carotid, subclavian and renal artery, though it can involve any part of aorta. Stenotic lesions are more common than aneurysms but the latter can occur in TA in 33%.[6,21] Circumferential wall thickening in arteritis due to

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vessel inflammation could be picked in MR angiography as vessel wall oedema in T2 and fat-suppressed sequences and mural contrast enhancement on T1 sequences.[6,21] GCA can also present as thoracic aneurysms,but presents in patients above 50 years. They usually give history of temporal headaches, visual loss or jaw claudication.[22-24] GCA typically affects cervical-cephalic arteries proximally to the entry point of the cranial dura as intracranial arteries lose internal elastic lamina and vasa vasorum as they penetrate dura. Only 10-15% of patients have isolated extracranial involvement, with the most common locations being the aorta and the upper limb arteries(subclavian and axillary artery).[6,21,24] Doppler ultrasound of the temporal artery may reveal the characteristic "halo sign" which has variable sensitivity and specificity. CTA in GCA often reveals long tapered stenoses in large vessels, but these findings are not pathognomonic. [24,25]

PAN was another differential which is a necrotizing pan arteritis that commonly affects young males at around 50 years. It is seen in the branching points of medium-sized vessels with lesions in different stages of development and is initially characterized by focal segmental inflammation that leads to weakening of wall thereby forming multiple small aneurysms and thrombosis later replaced by fibrosis culminating in vessel wall thickening and stenosis resulting in organ infarct especially kidney.[26] PAN can affect any vascular bed of any organ except those in the lungs. The renal manifestations described are renal artery stenosis, aneurysms, rupture of aneurysms leading to infarction or hematoma formation. However, it does not cause glomerulonephritis unlike in ANCA-related small-vessel vasculitis. Other systemic features favouring PAN are constitutional & musculoskeletal features, livedo reticularis, mononeuritis multiplex, Hep B positivity.[27] Multiple focal visceral/renal micro aneurysms (1-5 mm in size) are considered virtually pathognomonic of PAN due to involvement of renal arcuate or interlobar arteries.[28] Inflammatory markers are usually raised in all forms of vasculitides. Salient differences between vasculitides & its mimics are given in **Table 3**. Features of ocular, ear, nose & throat involvement, glomerulonephritis, upper and/or lower respiratory tract features with ANCA positivity would fetch a diagnosis of ANCA associated vasculitis (AAV).[29-31]

Hence Case 2 was FMD which is a segmental, non-atherosclerotic non inflammatory vasculopathy occurring most commonly in medium-sized arteries like renal artery, extracranial internal carotid artery and vertebral artery.[32] It can develop due to hyperplasia of medial walls or fibroplasia of media, adventitia or intima. Collagen deposition results in a thickened intima, often with fragmentation or duplication of the internal elastic lamina. The most common clinical manifestations are hypertension, headache and pulsatile tinnitus, hypertensive emergencies like coronary or cerebrovascular accidents, dissection of affected vessels and renal infarction.[33] These complaints occurring in younger individuals may point towards vasculitis. Medial fibroplasia is the most common type of FMD and has the classical 'string of beads' appearance on angiography due to alternating areas of stenosis and aneurysmal dilatation. Intimal FMD, which is the rarest of all types, has angiographic appearance of concentric band-like stenoses and smooth tapered lesions like those seen in large-vessel vasculitides.[34] It presents as unifocal or multifocal stenoses that can progress to renal artery dissection. It usually has bilateral renal involvement with aneurysms in middle to distal portion of renal artery with larger size having normal inflammatory markers.[35] Doppler Ultrasound (DUS) typically reveals increased peak systolic velocities in the mid to distal portions of the affected arteries in the absence of atherosclerosis.[35] The positive predictive value of DUS for diagnosing FMD in carotid and renal arteries are high as 87.7% and 94.2% respectively. Since negative predictive value is only 62%, routine DUS would not be an appropriate test for screening for FMD. CT Angiography (CTA) is recommended to be the imaging modality of choice due to its ability to distinguish between atherosclerotic plaques & FMD lesions & shorter scan times.[36] The characteristic CTA findings include arterial irregularities or a "beading" appearance, focal stenosis, dissections and aneurysms.[36] MRA with gadolinium is an alternative imaging modality which also has a high sensitivity of 97% and specificity of 93% compared with catheter-based angiography for diagnosing renal FMD. MRA not only detects dissection and aneurysms but also scores over other modalities in revealing the 'string of beads' appearance. Renal angiography remains the gold standard for the diagnosis of FMD and PAN.[37] Treatment of hypertension with anti-hypertensives remains the medical option for treatment. For patients with refractory hypertension or in those with severe renal artery or other vascular territory stenoses, revascularization is recommended[38] like percutaneous transluminal balloon angioplasty (PTA) with or without stenting (e.g.: stenoses, dissections, and aneurysms) or coiling (e.g.: aneurysms). Restenosis after PTA, complex lesions or inaccessible sites by PTA may need to undergo surgical procedures.[38] Thus, making an accurate diagnosis is crucial to avoid a lifetime of immunosuppression-related complications.

Case 3 is a case of Segmental Arterial Mediolysis, a non-inflammatory vasculopathy.[39] This entity is not properly understood but it is postulated that repeated vasoconstrictive stimuli is one of the triggers leading to degeneration of smooth muscle cells in media resulting in arterial damage.[40,41] Histopathology of SAM may show vacuole degeneration of smooth muscle cells in media with deposition of fibrin in media-adventitia junction with no inflammatory infiltrate whereas in vasculitis all arterial layers will be inflamed[41]. FMD predominantly affects young women whereas there is no sex predilection in SAM

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with affected individuals in 40–60-years age group.[42] Lesions are commonly distributed in mesenteric vessels of varying frequencies with superior mesenteric, coeliac, hepatic and renal, splenic and inferior mesenteric arteries being the most involved. Cerebrovascular involvement has also been reported.[43] Findings as per case studies are[43] aneurysm (76%), dissection (61%), rupture (46%), stenosis (19%) and occlusion that can present acutely with sudden intra-abdominal haemorrhage and ischemia, like the medium vessel vasculitis, PAN.[43,44] Post-prandial abdominal or flank pain is the most common symptom which is attributed to stenoses and occlusions of mesenteric vessels that cause intestinal ischemia. Gastrointestinal haemorrhage could be due to dissections and aneurysms, [41-43] sometimes leading to life threatening haemorrhage and shock resulting in death.[44] Constitutional symptoms, neurologic & cutaneous manifestations and arthralgias that are present in patients with PAN are often absent in SAM.[42] ESR and CRP was raised in 50-60 % according to literature review of 143 cases due to bleeding or tissue ischemia due to necrosis.[44] SAM should also be kept in mind when aneurysms, stenoses, and occlusions are identified in medium and large vessels, especially when these lesions are limited to one anatomic location.[42] Differential diagnosis for vessel aneurysms has been tabulated [**Table 4**].

Kalva et al. has postulated certain diagnostic criteria for SAM. Clinical criteria were absence of congenital predisposition for dissection or vascular disorders like Loeyz-Dietz syndrome/Marfan's syndrome /Ehler Danlos syndrome, FMD or arteritis, acute presentations- such as abdominal pain or hypotension due to GI bleed, haematuria or stroke, and/or chronic presentations such as abdominal pain, hypertension, haematuria (or asymptomatic). Imaging criteria were the presence of dissection, fusiform aneurysm, occlusion, beaded appearance, wall thickening of the mesenteric or renal arteries with or without organ infarction, absence of associated contiguous aortic dissection or atherosclerosis. Serologic criteria were absence of ANA, ANCA and normal complement levels.[43] CT angiography is the investigation of choice. PET scan could also help in differentiating vasculitis from SAM as there would be no vascular uptake in the latter. Treatment of SAM involves embolization, surgical bypass, stenting or resection of the injured arteries. [45,46]

Table 3: Differences in presentation between vasculitides and its mimics

	SAM	FMD	PAN	AAV	GCA	TA
Age	Any, MC 40-60	20-30	40-60	Any	>50 yrs	15-40 yrs
Gender	M>F	F>M	M+F	M+F	F>M	F>M, 9:1
Constitutional	-	-	+	+	+	+
Hypertension	+	+	+	+	+	+
Cutaneous features	-	-	+	+	+	=
Ocular	-	-	+	++	++	+
CVA	rare	rare	rare	rare	++	++
Pulmonary	-	-	-	++	-	-
Arthralgias/arthritis	-	-	++	++	+	+
Inflammatory markers	normal	normal	high	high	High	high
Vessel inflammation	nil	nil	present	present	present	present

Table 4: Differential diagnosis of vessel aneurysms

	SAM	FMD	PAN	GCA	TA
Anatomic site	Visceral mesenteric	Renal	Intraabdominal	Intracranial	Aorta and its branches
Biopsy	Segmental mediolysis, thrombosis, haemorrhage, aneurysms	Medial fibroplasia, thick collagen bundles alternating with thinned areas of media	Segmental necrotizing panarteritis with many immune cells	Vasculitis with predominant mononuclear cells or granuloma with multinucleate giant cells in inner portion of media, adjacent to internal elastic lamina	Mononuclear cell, giant cell and granulomas, starts from vasorum of adventitia and medio-adventitial junction
Angiography	'String of beads' appearance, stenoses, aneurysms, dissections, thrombosis	Classic 'string of beads' appearance; stenoses and narrowing;	Microaneurysms predominantly at vessel branch points	Extracranial - smooth, occlusion of subclavian, axillary, proximal brachial artery, long tapering stenoses, thoracic aneurysms	Large aneurysms and stenoses in aorta and great vessels

Case 4 presented with symmetrical peripheral gangrene in the setting of reduced ejection fraction. Symmetrical peripheral gangrene (SPG) was first described by Hutchison in 1891.[47] SPG is a rare clinical entity that presents with acute, symmetrical ischemia that involves two or more extremities that results in tissue death leading to gangrene in the absence of large vessel obstruction or vasculitis.[48] Various infective and noninfective causes have been enlisted as etiological factors for SPG.[49] [**Table 5**].

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Table 5: Aetiology of SPG

Infective	Bacterial	Staphylococci, Streptococci, Meningococci, Salmonella paratyphi, E. coli, Klebsiella pneumoniae
	Viral	Covid 19, HIV, Varicella zoster
	Parasitic	Plasmodium vivax, Pl.falciparum, Leptospira
Non-infective	Autoimmune	SLE, Primary vasculitides-cryoglobulinaemic vasculitis, PAN, APLA syndrome
	Envenomation	Snake venom
	Drugs	Warfarin, Propylthiouracil, Vasoconstrictors-Dopamine, Adrenaline
	Cardiac	Low output cardiac failure, MI, Myocardial pseudoaneurysm, Ventricular tachycardia, AV block

In infections, often disseminated intravascular coagulation (DIC), which is a hypercoagulable condition due to sepsis is implicated as the cause for gangrene. This case had heart failure with decreased cardiac output leading to compromised peripheral perfusion to limbs, skin, liver and spleen by vasoconstriction as a compensatory mechanism to ensure adequate supply to vital organs like brain & heart. Low flow state combined with microvascular occlusion from DIC leads to ischemic changes that involve distal extremities and advances, proximally resulting in gangrenous changes as seen in similar case reports.[50] Other aggravating factors include cold-induced vasospasm, vasoconstrictor drugs, Diabetes mellitus, and renal failure. 'Symmetric peripheral gangrene' was coined by Fishberg upon observing two patients who developed this condition in the background of cardiac failure.[51] With decreased peripheral perfusion pressures from 36 to 60 mm Hg, blood flow through digital arteries stops leading to gangrene. [52-54]

Treatments strategies are often unsuccessful since gangrene cannot be reverted. Identifying the cause remains the challenge and addressing it forms the solution. Hemodynamic stability is to be ensured and DIC is to be managed.[48] Vasopressors should be avoided as they can further compromise the blood circulation to these ischemic areas. Administration of fresh frozen plasma for replacement of clotting factors or use of anticoagulants, based on whether bleeding or thrombosis is predominant may be tried.[54] Other medical options tried with varying success rates are sympathetic blockades, vasodilators such as intravenous Nitroprusside, local or intravenous infusion of alpha-blockers (Phentolamine, Chlorpromazine), and infusion of prostaglandin (Epoprostenol).[54,55] But finally surgical debridement or amputation of the gangrenous area after the appearance of a line of demarcation is attempted. Prognosis is poor with mortality reported to be in a quarter of patients.[56,57] Most deaths ensued in 1-3 weeks following gangrene as per case studies.[56] Rest of the patients who survived underwent autoamputation or surgical removal. [57,58]

We observe here in the patient that the imaging of the lower limb vessels had atherosclerosis in aorta which is a non-inflammatory condition which affects larger arteries hence mimics large vessel vasculitides like TA.[59,60] The difference between the two is elaborated in **Table 6**.

Table 6- Atherosclerosis v/s TA

	Atherosclerosis	**TA**
Age	60 yrs	20-40 yrs
CT angiogram findings	Aneurysms>stenosis	Stenosis >aneurysms
ESR/CRP	Normal	Elevated
Life style risk factors	+++	+
PET CT	Eccentric thickening of wall, patchy uptake	Concentric thickening with diffuse uptake
Vessel wall calcification	+++	+
Limb pulses	Lower limbs more involved	Upper limbs more involved
Vessel predilection		
Subclavian artery	+	+++
Thoracic aorta	+	+++
Abdominal aorta	Infrarenal	Suprarenal or renal
Iliac vessels,IMV	++	+

Case 5- is an example of SPG but here the aetiology is an infectious agent, meningococci. As in the above case, there is symmetrical involvement of extremities- the lower limbs being involved first and later progressing to upper limbs if not controlled early. Gangrene is one of the manifestations in medium vessel vasculitides. The above clinical feature in young males makes autoimmune disease associated vasculitis highly likely. Neisseria meningitidis (Meningococcus) is the causative organism for Neisserial meningitis. Asymptomatic pharyngeal colonization occurs in humans, and they are transmitted by respiratory droplets and requires close, direct contact. Initial presentation may be a pharyngitis that develops later into haemorrhagic rash[61] which may be diffuse mottled or purpuric due to blood stream infection. There may be fever and altered sensorium. Gangrene can ensue later in meningococcal sepsis. Fatalities occur due to circulatory collapse due to release of endotoxin. Meningococci can impair anticoagulant (protein C is depleted in sepsis) and fibrinolytic pathway thereby

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leading to thrombus formation in small to medium vessels.[62] It damages endothelial cells in the cutaneous capillaries by direct invasion or immune complex mediated process, induces tissue thromboplastin on inflammatory cells and endothelial cells thereby promoting coagulation in those vessels culminating in DIC(Disseminated Intravascular Coagulation).[63] Hence infectious vasculitis doesn't warrant immunosuppressants but requires appropriate antimicrobials to arrest the spread of gangrene. Hence it must be identified with proper investigations. The history of fever with rapid spread of skin lesions like purpura, petechia, gangrene with other localizing signs like meningeal irritation should make us think of this possibility rather than primary systemic vasculitis.[64]

Purpuric rash in this infectious vasculitis can simulate that of Henoch Schoenlein purpura (HSP) as seen in other case reports.[62] The above patient had palpable purpuric lesions in the lower limbs which later coalesced & finally led to gangrene. Features that are common to both meningococcemia and HSP are prior history of pharyngitis, fever, non-blanchable purpuric rash and arthralgia due to antigen-antibody reaction. There has been reports of HSP after meningococcal vaccine. But arthritis, gastrointestinal vasculitis & nephritis is less common with meningococcal infection than HSP[62]. Fever in HSP is usually low grade & patient turns afebrile after appearance of rash whereas fever is high grade and prominent with rash in meningococcaemia. Systemic symptoms of fever, arthralgia and rash are due to bacterial invasion of blood stream, but later appearance is due to antigen-antibody reaction. Meningitis is seen in meningococcal infection with CSF and skin lesions depicting gram negative intracellular and extracellular diplococci. CSF culture done with chocolate agar or mucosal cultures done with Thayer-Martin media will yield Neisserial growth. Assays can be done to detect meningococcal DNA. HPE of skin lesions shows necrosis of endothelial cells in capillaries and small veins in dermis and subcutaneous tissue with accompanying neutrophilic infiltration and thrombotic occlusion of vessels. Meningococci are seen within the endothelium and thrombi. HSP lesions show leukocytoclastic vasculitis with perivascular infiltrate of neutrophils or lymphocytes with no demonstrable organisms. Further direct immunofluorescence done on lesions show deposition of IgA and complement along the dermo-epidermal junction in HSP whereas with meningococcal infection it is IgG, IgA and C3 with organisms. There is rapid resolution of symptoms following Ceftriaxone administration ideally given for 1 week. HSP may require steroid for severe GI manifestations, arthritis and nephritis, meningococcaemia doesn't usually require it unless adrenal failure occurs.[64]

CONCLUSION

Most of the patients described here were young patients, hence vasculitis was considered the differential in all these entities. Schematic history with examination aided by appropriate imaging and blood investigations have contributed to reaching the correct diagnosis (**Table 7**). Often cases present with non-classic clinical features- herein, we need to rely on newer modalities of imaging like PET CT scan to come a conclusion. Hence a thorough awareness of the mimics is extremely important.

Table 7: Similarities and differences between vasculitides and vasculitis mimickers in case series

Vasculitis mimic	Vasculitis as the close differential	Similar features	Differentiating features
Middle aortic syndrome	TA	Young hypertension	Inflammatory markers normal in MAS
Fibromuscular dysplasia	PAN	Visceral aneurysms	Bilateral renal artery stenosis more in FMD
Segmental arterial mediolysis	PAN	Visceral aneurysms	Aneurysm with dissection in branches of abdominal aorta in MAS
Symmetrical peripheral gangrene-LV dysfunction	Medium and small vessel vasculitis	Gangrene	ANCA negative, absence of vasculitis imaging-wise, normal inflammatory markers in SPG
Segmental peripheral gangrene-meningococcal infection	Medium vessel vasculitis and PAN	Gangrene, fever, purpura	Short duration fever, absence of other constitutional features of medium vessel vasculitides like livedo reticularis, orchitis, arthritis,nephritis in meningococcal infection

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