

Original Research Article

 Received
 : 30/09/2024

 Received in revised form
 : 05/10/2024

 Accepted
 : 16/10/2024

Keywords: Nasal Cavity, Paranasal Sinuses, SNUC.

Corresponding Author: **Dr. O H Radhika Krishna,** Email: othulururadhika@gmail.com

DOI: 10.47009/jamp.2024.6.5.106

Source of Support: Nil, Conflict of Interest: None declared

Int J Acad Med Pharm 2024; 6 (5); 562-575



LESIONS OF NASAL CAVITY & PARANASAL SINUSES: A PROSPECTIVE STUDY

Swarupa Ravuri¹, Sreelakshmi Korra², Swapna Kotha³, O. H Radhika Krishna⁴, Chandrika Boleneni⁵

^{1, 2}Assistant Professor, Department of Pathology, MNJ institute of Oncology and Regional Cancer Center, Red Hills, Lakdikapul, Hyderabad, Telangana, India

³Assistant Professor, Department of Radiodiagnosis, MNJ institute of Oncology and Regional Cancer Center, Red Hills, Lakdikapul, Hyderabad, Telangana, India

⁴Professor, Department of Pathology, MNJ institute of Oncology and Regional Cancer Center, Red Hills, Lakdikapul, Hyderabad, Telangana, India

⁵Senior Resident, Department of Pathology, MNJ institute of Oncology and Regional Cancer Center, Red Hills, Lakdikapul, Hyderabad, Telangana, India

Abstract

Background: Malignant lesions of the nasal cavity and paranasal sinuses are rare, accounting for less than 3% of upper aerodigestive tract malignancies. Due to their heterogeneous nature and anatomical complexity, diagnosing and treating these lesions presents significant challenges. This prospective study aims to evaluate the histopathological spectrum and treatment outcomes of rare malignant lesions at a tertiary cancer referral center. Materials and Methods: A prospective study was conducted from January 2020 to June 2023, involving 57 patients with confirmed malignant lesions of the nasal cavity and paranasal sinuses. Data collected included clinical presentation, histopathological findings, immunohistochemical markers, and treatment protocols. The outcomes of surgery, radiotherapy, and chemotherapy were analyzed. Result: The study identified 89.5% of lesions as common malignancies (e.g., non-Hodgkin's lymphoma, nasopharyngeal carcinoma), while 10.5% were classified as rare malignancies (e.g., malignant melanoma, sinonasal teratocarcinosarcoma). Immunohistochemical analysis played a crucial role in diagnosis. Recurrence rates were 28%, and treatment-related complications occurred in 17.5% of cases. Conclusion: Malignant lesions of the nasal cavity and paranasal sinuses require a multidisciplinary approach for accurate diagnosis and treatment. Early intervention, guided by histopathological and immunohistochemical findings, improves survival outcomes. Regular follow-up is critical for detecting recurrence.

INTRODUCTION

Malignant tumors of the nasal cavity and paranasal sinuses are uncommon and present significant diagnostic and therapeutic challenges due to their proximity to vital structures such as the orbit, skull base, and cranial nerves.^[1] These tumors account for less than 3% of all head and neck malignancies and tend to be diagnosed at an advanced stage due to non-specific early symptoms, including nasal obstruction and epistaxis.^[2] Their histological heterogeneity further complicates diagnosis and treatment planning, necessitating advanced diagnostic modalities like immunohistochemistry (IHC) and imaging techniques like MRI.^[3]

Sinonasal malignancies exhibit a wide spectrum of histopathological subtypes, including squamous cell carcinoma, adenoid cystic carcinoma, and rarer entities such as sinonasal undifferentiated carcinoma (SNUC), malignant melanoma, and sinonasal teratocarcinosarcoma.^[4] Early diagnosis is often hindered by the anatomical complexity of the paranasal sinuses, allowing for tumor growth before symptoms become apparent.^[5]

The increasing use of MRI and advanced histopathological tools, including IHC markers, has greatly improved the identification of specific tumor types, thereby guiding more precise treatment plans.^[6] However, treatment outcomes remain poor, with high rates of recurrence and metastasis, particularly in aggressive subtypes such as SNUC and sinonasal teratocarcinosarcoma.^[7]

This study aims to expand the existing knowledge by presenting a comprehensive prospective analysis of rare malignant tumors of the nasal cavity and paranasal sinuses, including their clinical presentation, imaging characteristics, and treatment outcomes.

MATERIALS AND METHODS

Study Design: This prospective study was conducted at a tertiary cancer referral center from January 2020 to June 2023. The study population comprised 57 patients who presented with histopathologically confirmed malignant lesions of the nasal cavity and paranasal sinuses.

Inclusion Criteria

Patients with histopathologically confirmed malignant lesions of the nasal cavity or paranasal sinuses.

Exclusion Criteria

Patients with benign sinonasal tumors or those without histological confirmation of malignancy.

Data Collection: Data on patient demographics, clinical symptoms, imaging findings (CT and MRI), histopathology, immunohistochemical markers, and treatment protocols were recorded. Tumor types were classified based on histological and IHC findings.

Imaging Techniques: MRI scans were used for detailed tumor characterization, particularly for assessing soft tissue involvement, tumor extension, and proximity to critical structures such as the orbit and skull base. CT scans were used to evaluate bony involvement, including bone erosion or destruction. **Treatment Protocol:** Patients received treatment according to their specific tumor type and disease stage. Treatment modalities included surgical resection, radiotherapy, chemotherapy, or a combination. Surgical resection was performed in cases where the tumor was localized, with adjuvant therapies used in more advanced or metastatic cases.

RESULTS

Interpretation: The majority of patients were in the age group of 31-50 years, with a male predominance (63.2%), reflecting trends typically seen in sinonasal malignancies.

Interpretation: Nasal obstruction was the most common presenting symptom (70.2%), followed by epistaxis (49.1%) and facial swelling (38.6%).

Table 1: Demographic Characteristics of the Study Population (N = 57).			
Characteristic	Frequency (n)	Percentage (%)	
Age Group (years)			
18-30	8	14.0%	
31-50	25	43.9%	
51-70	20	35.1%	
>70	4	7.0%	
Gender			
Male	36	63.2%	
Female	21	36.8%	

Table 2: Clinical Symptoms at Presentation (N = 57)

Table 2. Children Symptoms at Tresentation $(1 - 57)$			
Symptom	Frequency (n)	Percentage (%)	
Nasal Obstruction	40	70.2%	
Epistaxis	28	49.1%	
Facial Swelling	22	38.6%	
Diplopia	14	24.6%	
Anosmia	10	17.5%	

Table 3: Imaging Findings in Malignant Lesions (N =	: 57)
-----------------------------------------------------	-------

Imaging Modality	Common Findings	Number of Cases (n)	Percentage (%)
CT Scan	Bony Erosion/Destruction	26	45.6%
MRI	Intracranial Extension	16	28.1%
MRI	Orbital Invasion	12	21.1%
CT Scan	Lymph Node Involvement	8	14.0%

Interpretation: Bony erosion was the most frequent imaging finding (45.6%). MRI revealed intracranial extension in 28.1% of cases, indicating the aggressive nature of these tumors.

Table 4: Histopathological Findings in Malignant Lesions (N = 57)			
Malignancy	Number of Cases (n)	Percentage (%)	
Non-Hodgkin's Lymphoma	16	28%	
Nasopharyngeal Carcinoma	11	19.3%	
Squamous Cell Carcinoma	8	14%	
Sinonasal Undifferentiated Carcinoma	5	8.7%	
Neuroblastoma	3	5.3%	
Rhabdomyosarcoma	3	5.3%	
Ewing's Sarcoma	3	5.3%	
Adenoid Cystic Carcinoma	2	3.5%	
Rare Malignancies	6	10.5%	

Interpretation: Non-Hodgkin's lymphoma was the most frequent diagnosis, followed by nasopharyngeal carcinoma and squamous cell carcinoma. Rare malignancies comprised 10.5% of the cases.

Table 5: Treatment Modalities (N = 57)			
Treatment Modality	Number of Cases (n)	Percentage (%)	
Surgery Alone	20	35.1%	
Surgery + Radiotherapy	25	43.9%	
Surgery + Chemotherapy	5	8.8%	
Chemotherapy Alone	4	7.0%	
Palliative Treatment	3	5.2%	

Interpretation: Surgical resection combined with radiotherapy was the most common treatment approach (43.9%). Palliative treatment was administered in advanced, inoperable cases.

Table 6: Treatment Outcomes and Complications (N = 57)			
Outcome	Number of Cases (n)	Percentage (%)	
Local Recurrence	16	28.1%	
Distant Metastasis	8	14.0%	
2-Year Survival	30	52.6%	
Treatment-related Complications	10	17.5%	

Interpretation: The recurrence rate was 28.1%, with distant metastasis occurring in 14% of cases. The overall 2-year survival rate was 52.6%, and treatment-related complications were seen in 17.5% of patients.

Imaging and Histopathological Findings: 1. Malignant Melanoma

An 80-year-old male presented with a mass in the left nasal cavity. Histopathological examination and immunohistochemistry confirmed the diagnosis of malignant melanoma. The tumor was positive for HMB-45 and S100 markers, and negative for PCK, CK7, synaptophysin, and LCA. The high Ki-67 index suggested a high proliferative rate, indicating aggressive tumorbehavior.

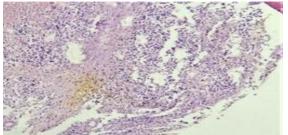


Figure 1:a Malignant Melanoma H&E 10x

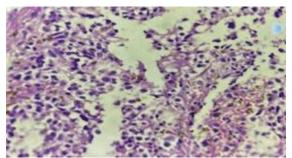


Figure1:b Malignant Melanoma H&E 40x 40x10x

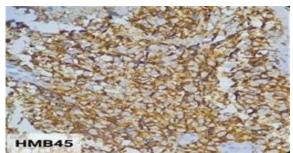


Figure 1:c Malignant Melanoma IHC HMB45 positive

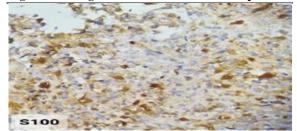


Figure 1:d Malignant Melanoma IHC S100 Positive

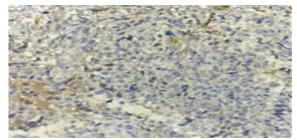
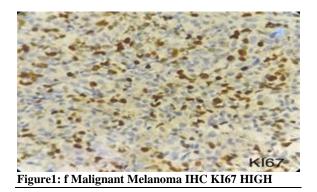


Figure1: e Malignant Melanoma IHC PCK negative

564



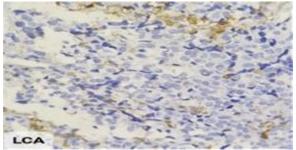


Figure1: g Malignant Melanoma IHC LCA negative 2. Meningioma

A 65-year-old male presented with a nasal cavity mass, which was confirmed as an ectopic meningioma, a rare occurrence in sinonasal malignancies. Immunohistochemistry showed the tumor was positive for EMA and negative for PCK, SMA, CD34, ER, and PR, supporting the diagnosis of ectopic meningioma. These tumors account for less than 1% of non-epithelial sinonasal tumors.



Figure: 2a Meningioma H&E 10X

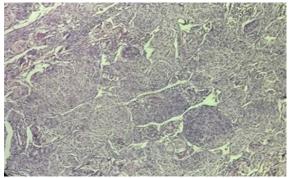


Figure:2b Meningioma H&E 40X



Figure :2c Meningioma IHC EMA POSITIVE

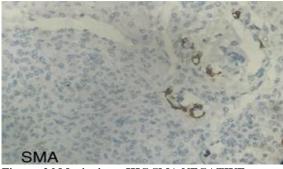


Figure :2d Meningioma IHC SMA NEGATIVE

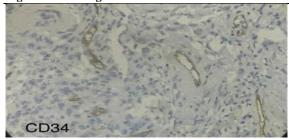


Figure2: e Meningioma IHC CD34 NEGATIVE

3. Chordoma

Chordomas are rare tumors that can present in unusual locations, including the nasal cavity. A 4year-old female presented with a nasal mass, which was confirmed as a chordoma based on histological findings. Immunohistochemistry demonstrated positivity for S100 and EMA, with a low Ki-67 index (5%), suggesting a slow-growing tumor.



Figure :3a Chordoma H&E 10X

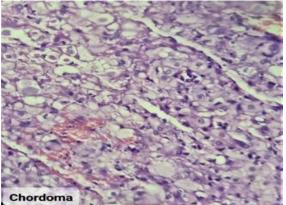


Figure 3b:Chordoma H&E 40X

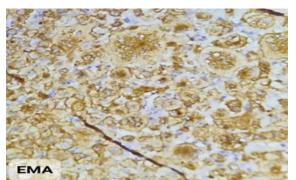


Figure :3c Chordoma IHC EMA 40X POSITIVE

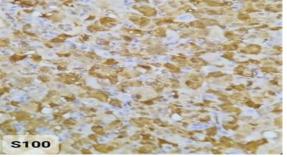


Figure :3d Chordoma IHC S100 40X POSITIVE

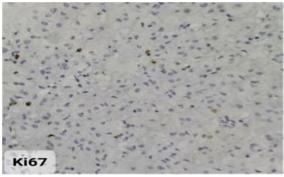


Figure :3e Chordoma IHC KI67 LOW

4. Sinonasal Adenocarcinoma

A 37-year-old male presented with a nasal mass, which was identified as intestinal-type sinonasal adenocarcinoma. Immunohistochemical staining showed positivity for CK20, CDX2, villin, MUC2, and SATB2, confirming the intestinal differentiation. Non-intestinal variants of sinonasal adenocarcinoma typically lack these markers, further supporting the diagnosis.

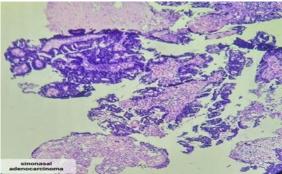


Figure :4a Sinonasal Adenocarcinoma H&E 10x

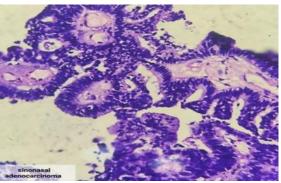


Figure:4bSinonasal Adenocarcinoma H&E 40x

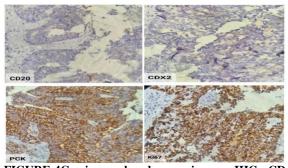


FIGURE:4C sinonasal adenocarcinoma IHC CD 20,,CDX2 NEGATIVE,PCK POSITIVE KI67 - HIGH.

5. Clear Cell Odontogenic Carcinoma

A 40-year-old male presented with a left nasal mass. Histopathological examination revealed clear cell odontogenic carcinoma, a rare tumor. Immunohistochemical staining showed strong positivity for P63, confirming the odontogenic origin of the tumor.

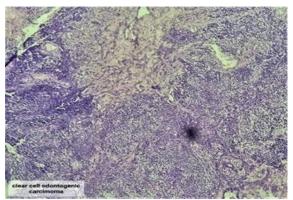


Figure:5a Clear Cell Odontogenic Carcinoma H&E 10x

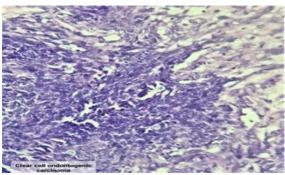


Figure:5bClearCellOdontogenic Carcinoma H&E 40x

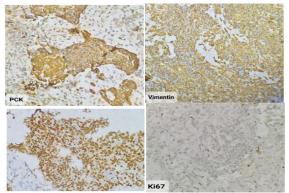


Figure: 5C Clear Cell Odontogenic Carcinoma IHC PCK ,VIMENTIN, P63 POSITIVE KI67 LOW

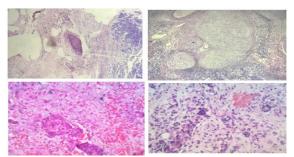


Figure: 6A. Sinonasal teratocarcinosarcoma H&E 40x showing mesenchymal, squamous, neuroepitelial elemements.

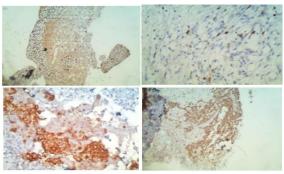


Figure: 6b. Sinonasal teratocarcinosarcoma IHC S100, PCK, CD56 POSITIVE.

6. Sinonasal Teratocarcinosarcoma

Sinonasal teratocarcinosarcoma is a rare and highly aggressive tumor of the nasal cavity. A 55-year-old male presented with a large, destructive mass. Histological analysis showed a heterogeneous neoplasm with features of carcinoma, sarcoma, and immature teratoma. Immunohistochemistry revealed positivity for S100, PCK, and CD56, consistent with the diagnosis.

Histological and Immunohistochemical characterization of common nasal and paranasal malignant lesions:

1. Adenoid Cystic Carcinoma

Adenoid cystic carcinoma is a slow-growing but highly infiltrative tumor, often involving the sinonasal region. The tumor was positive for P63 on immunohistochemistry, confirming the diagnosis.

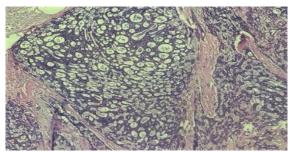
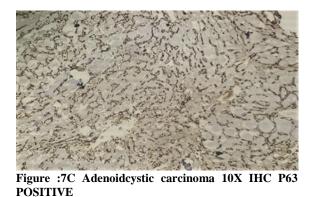


Figure :7a Adenoidcystic carcinoma H&E 10x



Figure:7b Adenoidcystic carcinoma 40XIHC CK7 positive

567



2. Neuroblastoma

Neuroblastoma is a rare malignant tumor that typically arises in the sympathetic nervous system and can occasionally present in the nasal cavity and paranasal sinuses. The case in question involves a pediatric patient who presented with a rapidly growing mass in the nasal cavity, leading to significant nasal obstruction and facial swelling. MRI and histopathological examination confirmed diagnosis the of neuroblastoma. Immunohistochemistry revealed the tumor's positivity for markers such as CD56, synaptophysin, and chromogranin, which are indicative of neuroendocrine origin.

This tumor is highly aggressive and often requires multimodal therapy, including surgical resection, chemotherapy, and radiotherapy. The proximity of the tumor to critical structures such as the orbit and skull base complicates treatment planning. In this case, MRI revealed extensive tumor invasion into the surrounding tissues, necessitating a comprehensive approach to both local and systemic control of the disease.

IHC Markers

9yr/M	50yr/F	55yr/F
Sinonasal mass	Nasal cavity mass	Nasal cavity mass
NSE,CD 56 -	Synaptophysin-	PCK, P63,
positive	Positive.	Synaptophysin -
Vimentin - weak	Vimentin - weak	positive
positive	positive in few	
	cells	
PCK, Myogenin,	PCK,LCA,P63 -	Ki 67 - high
Desmin - Negative	Negative	

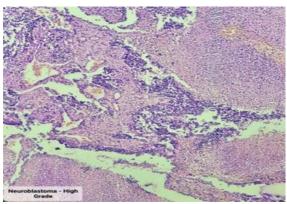


Figure :8a Neuroblastoma H&E 10x.

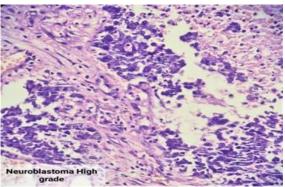


Figure :8b Neuroblastoma H&E 40 x.

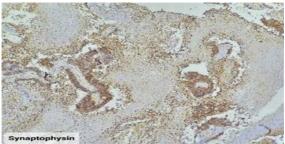


Figure :8c Neuroblastoma 10 x IHC Synaptophysin Positive.

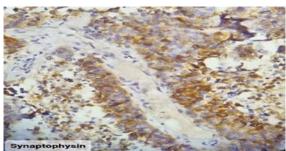


Figure:8d Neuroblastoma 40 x IHC synaptophysin positive

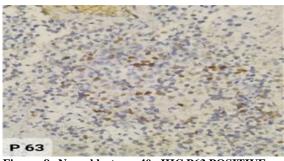


Figure :8e Neuroblastoma 40x IHC P63 POSITIVE

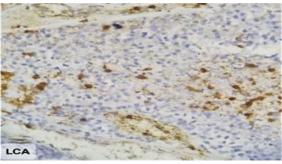


Figure 8f Neuroblastoma 40 x IHCLCA negative.

3. Ewing's Sarcoma

Ewing's sarcoma is a rare tumor that can involve the sinonasal tract. In this case, a young female presented with a sinonasal mass that was confirmed as Ewing's sarcoma. Immunohistochemistry showed positivity for CD99 and Vimentin, confirming the diagnosis.

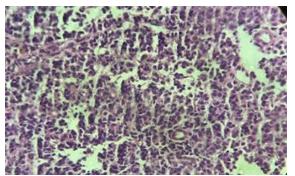


Figure 9:a Ewings sarcoma H&E40 x.

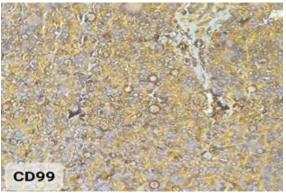


Figure 9:b Ewings sarcoma40 x IHC CD99 positive

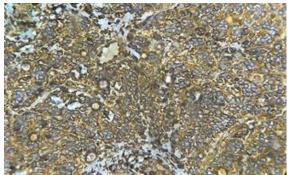


Figure 9:c Ewings sarcoma40 x IHC VIMENTINE positive

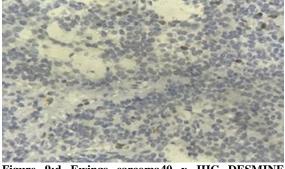


Figure 9:d Ewings sarcoma40 x IHC DESMINE negative.

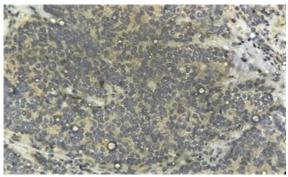
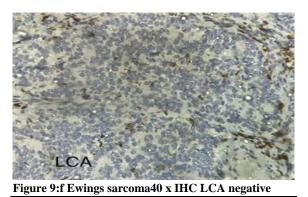
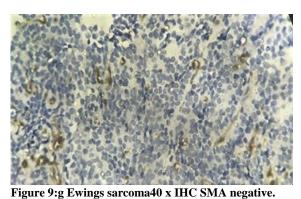


Figure 9:e Ewings sarcoma40 x IHC CD56 negative





CD99, Vimentin Positive PCK,CD56, Synaptophysin, Negative

4. Rhabdomyosarcoma (RMS)

SMA, Desmin

Rhabdomyosarcoma (RMS) is a highly malignant soft tissue sarcoma that arises from skeletal muscle progenitors. It is more common in children and adolescents and can occasionally present in the nasal cavity and paranasal sinuses. The case here involves a pediatric patient presenting with a rapidly enlarging nasal mass, leading to significant nasal obstruction and facial swelling.

Histopathological examination confirmed the diagnosis of embryonal rhabdomyosarcoma, a common subtype of RMS, known for its aggressive behavior and potential to invade surrounding tissues. Immunohistochemistry showed positivity for

muscle-specific markers such as desmin and myogenin, which are characteristic of RMS.

MRI demonstrated extensive local invasion into the paranasal sinuses and adjacent soft tissues, with the tumor encroaching on the orbit and skull base. Multimodal therapy, including surgery, chemotherapy, and radiotherapy, is typically required for RMS due to its aggressive nature.

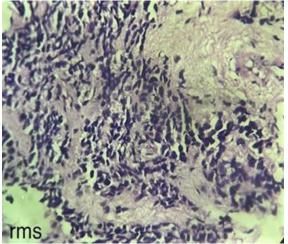


Figure 10:aRhabdomyosarcoma H&E

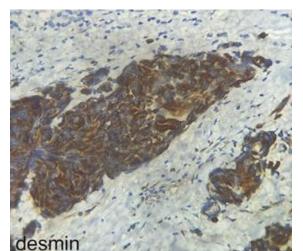


Figure 10:bRhabdomyosarcoma40x DESMIN positive.

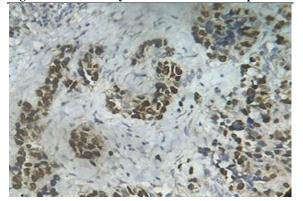


Figure 10:C Rhabdomyosarcoma40x MYOGENIN positive.

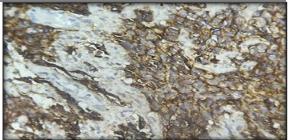


Figure 10:D Rhabdomyosarcoma40x synaptophysin negative.

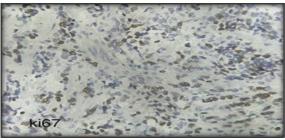


Figure 10:E Rhabdomyosarcoma40X KI67

Desmin, Myogenin, Vimentin	Positive
CD99, P63, Synaptophysin	Negative

5.Non-Hodgkin's Lymphoma:

- Non-Hodgkin's Lymphomas: (16 cases 28%) The majority of NHLs of the head and neck occur in extranodal sites such as the paranasal sinuses, nasal cavity, oral cavity, salivary glands and laryngopharynx.
- NHLs of the sinonasal tract are uncommon malignancies.
- Non-Hodgkin's Lymphomas that included B cell type -DLBCL, Plasmablastoma, NK cell type, T-cell Lymphoblastic lymphoma

Sn. No	Non-Hodgkin's Lymphomas	No. of cases	Percentage(%)
1	B cell lymphoma	11	19.2%
	DLBCL	09	14%
	- Plasmablastic lymphoma	02	3.5%
2	T cell lymphoma	01	1.7%

3	NK cell lymphoma	03	3.5%
4	T- cell Lymphoblastic lymphoma	01	1.7%
Total		16	28%

5A . Non-Hodgkin's Lymphoma B CELL TYPE Non-Hodgkin's lymphomas, particularly of B-cell type, are common malignancies in the sinonasal region. Of the 16 cases observed, diffuse large Bcell lymphoma (DLBCL) was the most frequent subtype. Immunohistochemistry showed positivity for CD20 and BCL6 markers. In one case, a 5-yearold male presented with a T-cell lymphoblastic lymphoma, with immunohistochemical markers confirming the diagnosis.

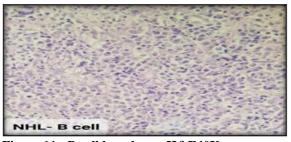


Figure 11:aB cell lymphoma H&E40X.

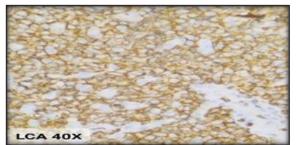


Figure 11:B B cell lymphoma 40XIHC LCA positive.

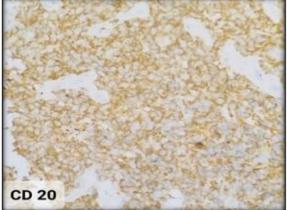
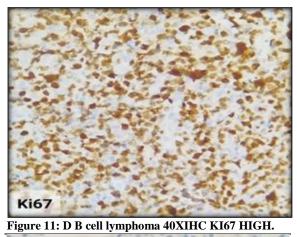


Figure 11:C B cell lymphoma 40XIHC CD 20 POSITIVE.



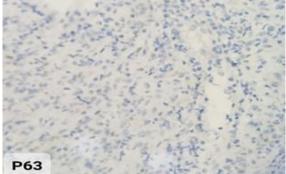


Figure 11 E :B cell lymphoma 40XIHC P63 nagative.

5B .Diffuse Large B-Cell Lymphoma (DLBCL)

Diffuse Large B-Cell Lymphoma (DLBCL) is the most common subtype of non-Hodgkin lymphoma, known for its aggressive behavior and rapid growth. In this case series, 9 patients presented with sinonasal involvement, displaying symptoms such as nasal obstruction, facial swelling, and occasional epistaxis.

Histopathological examination and immunohistochemical staining confirmed the diagnosis of DLBCL, with the tumor cells showing strong positivity for CD20, a B-cell marker, and BCL6. The proliferation index, as measured by Ki-67, was elevated, suggesting a high-grade malignancy.

MRI imaging revealed mass lesions within the nasal cavity, with invasion into the paranasal sinuses in several cases. The extent of the disease often required a combination of chemotherapy (R-CHOP regimen) and radiotherapy, with variable outcomes depending on the stage at diagnosis.

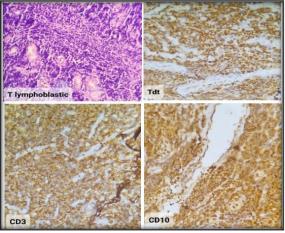


FIGURE12A.T Lymphoblastic Lymphoma H&E 40x &IHC TDT,CD3,CD10 POSITIVE.

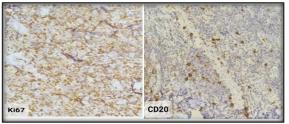


FIGURE12b:TLymphoblasticLymphomaIHCki67 POSITIVE CD20 negative.

5. T-Lymphoblastic Lymphoma

A 5-year-old male presented with a rapidly enlarging nasal mass, leading to nasal obstruction and facial swelling. Histopathological examination confirmed the diagnosis of T-lymphoblastic lymphoma, a rare and aggressive form of non-Hodgkin lymphoma, most commonly seen in pediatric patients.

Immunohistochemical analysis demonstrated strong positivity for CD3 and TdT, consistent with T-cell lineage. This type of lymphoma is characterized by its rapid progression and requires prompt and aggressive treatment, often involving chemotherapy. MRI imaging showed a large mass in the nasal cavity with involvement of the surrounding soft tissues, indicating the aggressive nature of the disease. Early diagnosis and a multimodal treatment approach, including chemotherapy, were critical in managing this case.

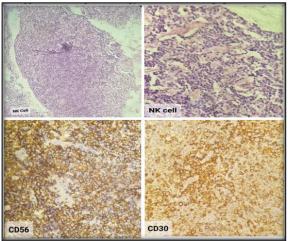


FIGURE13A. NK CELL LYMPHOMA H&E40x &IHC CD56,CD30 POSITIVE.

6. Natural Killer (NK) Cell Lymphoma

Natural Killer (NK) cell lymphoma is a rare and highly aggressive form of non-Hodgkin lymphoma, predominantly affecting the nasal cavity. It is more common in Asian populations and is often associated with Epstein-Barr virus (EBV) infection. In this case, the patient presented with a nasal mass, which was causing significant nasal obstruction and pain. Histopathological facial and immunohistochemical examination confirmed NKcell lymphoma. Immunohistochemistry showed strong positivity for CD56 and CD3, with EBVencoded RNA (EBER) in situ hybridization, confirming the association with Epstein-Barr virus. MRI imaging revealed a destructive mass in the nasal cavity with invasion into adjacent structures, including the paranasal sinuses. Due to the aggressive nature of NK-cell lymphoma, the treatment plan included chemotherapy combined with radiotherapy.

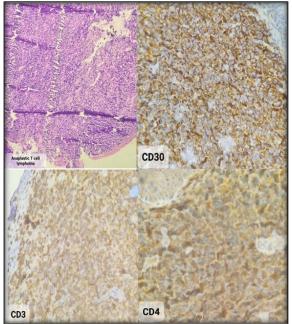


FIGURE14A.AnaplasticLymphomaH&E40x&IHCCD 30,CD3,CD4 POSITIVE.

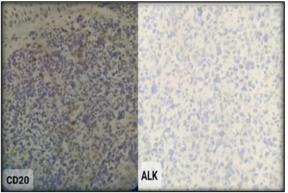


FIGURE 14B. AnaplasticLymphom IHCCD20, ALK NEGATIVE.

7. Anaplastic T-Cell ALK-Negative Lymphoma

Anaplastic T-cell lymphoma, particularly the ALKnegative subtype, is an aggressive form of peripheral T-cell lymphoma. A 46-year-old female presented with a nasal mass and associated symptoms of nasal obstruction and facial pain. ALK-negative anaplastic large cell lymphoma (ALCL) has a poor prognosis compared to the ALKpositive variant, and its presentation in the sinonasal region is rare.

Histopathological and immunohistochemical studies confirmed the diagnosis, showing positivity for CD30 and T-cell markers, with negativity for ALK (anaplastic lymphoma kinase), which defines this subtype. ALK-negative ALCL is known for its aggressive course, and treatment typically involves chemotherapy, although outcomes are less favorable than in ALK-positive cases.

MRI revealed a significant mass in the nasal cavity with invasion into adjacent structures, necessitating an aggressive treatment regimen. Early diagnosis and a multidisciplinary approach to management were critical for this case.

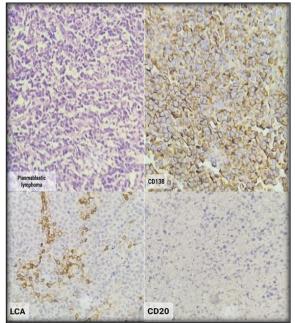


FIGURE15A PLASMA BLASTIC LYMPHOMA H&E 40X ,IHC CD138 POSITIVE ,LCA, CD20 NEGATIVE.

8. Plasmablastic Lymphoma

Plasmablastic lymphoma is an aggressive subtype of non-Hodgkin lymphoma, typically associated with immunocompromised patients, especially those with HIV infection. In this study, two cases of plasmablastic lymphoma presented with masses in the nasal cavity. The patients exhibited symptoms of nasal obstruction and facial swelling.

Histopathological analysis revealed large, atypical cells with plasmablastic morphology, and immunohistochemical staining showed positivity for CD138, MUM1, and EMA, confirming the plasmablastic phenotype. Notably, CD20 was negative, which is characteristic of this lymphoma subtype. EBV-encoded RNA (EBER) was also positive, indicating an association with Epstein-Barr virus.

MRI scans showed extensive soft tissue masses with invasion into adjacent structures, including the paranasal sinuses. The aggressive nature of plasmablastic lymphoma necessitated immediate initiation of chemotherapy, typically involving regimens such as CHOP or EPOCH.

Sinonasal undifferentiated carcinoma (SNUC):

WHO definition: Undifferentiated carcinoma lacking evidence of differentiation such as squamous, glandular or neuroendocrine differentiation by histology and immunohistochemistry

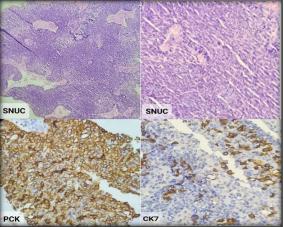


FIGURE 16A Sinonasal Undifferentiated Carcinoma H&E 40X IHC PCK POSITIVE, CK7 NEGATIVE.

9. Sinonasal Undifferentiated Carcinoma (SNUC)

Sinonasal undifferentiated carcinoma (SNUC) is a rare and highly aggressive malignancy arising from the nasal cavity and paranasal sinuses. It is characterized by rapid growth, local invasion, and a poor prognosis. A patient in this case presented with a large sinonasal mass, leading to significant nasal obstruction and facial pain.

Histopathological examination revealed undifferentiated malignant cells, consistent with SNUC. Immunohistochemical staining showed no specific squamous or glandular differentiation, which is typical of this carcinoma type. MRI imaging confirmed the presence of a large, irregular mass with extension into the orbit and skull base, further highlighting the aggressive nature of this disease.

SNUC requires a multimodal treatment approach, typically involving surgery followed by radiotherapy and chemotherapy. Early diagnosis and intervention are critical in managing this aggressive tumor, although outcomes remain poor due to its high recurrence rate.

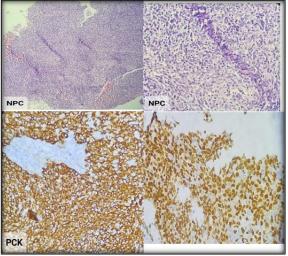


FIGURE 17A Nasopharyngeal Carcinoma H&E 40X IHC PCK,p63 POSITIVE.

10. Nasopharyngeal Carcinoma (NPC)

Nasopharyngeal carcinoma (NPC) is a malignant tumor that originates in the epithelial lining of the nasopharynx. It is particularly common in regions such as Southeast Asia and is associated with Epstein-Barr virus (EBV) infection. In this case, the patient presented with a mass in the nasopharynx, accompanied by symptoms of nasal obstruction, epistaxis, and cervical lymphadenopathy.

Histopathological examination confirmed the diagnosis of nasopharyngeal carcinoma, with immunohistochemical staining showing positivity for EBV-encoded RNA (EBER) and epithelial markers such as CK5/6. MRI imaging revealed a large tumor mass extending into the nasal cavity and paranasal sinuses, with possible involvement of the skull base.

Nasopharyngeal carcinoma is treated primarily with radiotherapy, often combined with chemotherapy in advanced cases. Due to its location, NPC frequently presents at advanced stages, necessitating a multimodal treatment approach.

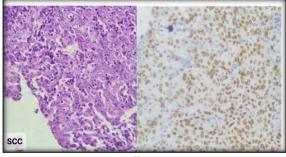


FIGURE 18A squamous cell Carcinoma H&E 40X IHC P63 POSITIVE.

11. Squamous Cell Carcinoma (SCC)

Squamous cell carcinoma (SCC) of the nasal cavity and paranasal sinuses is one of the most common malignancies arising in this region. In this case, the patient presented with a nasal mass accompanied by symptoms of nasal obstruction, epistaxis, and facial pain.

Histopathological examination confirmed the diagnosis of squamous cell carcinoma, with the tumor displaying typical keratinization and intercellular bridges. Immunohistochemistry showed strong positivity for cytokeratin (CK) and p63, markers that are characteristic of SCC.

MRI imaging demonstrated a large, irregular mass extending into the paranasal sinuses, with evidence of local invasion into the surrounding soft tissues. Treatment involved surgical resection followed by radiotherapy, with chemotherapy considered in more advanced cases.

DISCUSSION

This study highlights the challenges of diagnosing and treating rare malignant lesions of the nasal cavity and paranasal sinuses. These tumors, though rare, are highly aggressive and frequently diagnosed at advanced stages due to non-specific early symptoms and their anatomical complexity.^[8]

Imaging and Diagnosis: MRI proved invaluable in evaluating soft tissue involvement, tumor invasion into surrounding structures, and guiding surgical resection. CT scans were essential for detecting bony destruction, particularly in cases of malignant melanoma and sinonasal undifferentiated carcinoma.^[9] For example, the MRI of the sinonasal teratocarcinosarcoma case showed extensive orbital invasion, which greatly influenced the surgical approach.^[10]

Histopathology and Immunohistochemistry: Histopathological examination confirmed the diagnosis of rare entities such as chordoma and sinonasal teratocarcinosarcoma. Immunohistochemical markers like HMB-45 and S100 were essential in diagnosing malignant melanoma.^[11] These techniques enabled more accurate diagnosis, helping tailor specific treatment strategies for each patient.^[12]

Treatment and Outcomes :Multimodal treatment, including surgery, radiotherapy, and chemotherapy,

574

was necessary in most cases. Despite aggressive treatment, recurrence rates were high (28.1%), particularly sinonasal in patients with undifferentiated sinonasal carcinoma and teratocarcinosarcoma.[13] Treatment-related complications, such as infections and cranial nerve damage, occurred in 17.5% of patients, underlining the importance of close postoperative monitoring.^[14] Prognosis: The overall prognosis for patients with sinonasal malignancies remains poor, with a 2-year survival rate of 52.6%. Early diagnosis and aggressive treatment are key to improving outcomes, but novel therapies, such as targeted therapies and immunotherapies, may offer new hope for patients with treatment-resistant malignancies.^[15]

CONCLUSION

This prospective study provides valuable insights into the clinical, imaging, histopathological, and treatment outcomes of rare malignant lesions of the nasal cavity and paranasal sinuses. While surgery, radiotherapy, and chemotherapy remain the standard treatment modalities, recurrence rates and complications are common, highlighting the need for novel treatment approaches. Early diagnosis using advanced imaging and immunohistochemistry plays a critical role in improving survival rates in these patients.

• Tumors of nasal cavity and paranasal sinuses are rare cancers, with a prevalence of less than 1%.

• A multimodal and multidisciplinary approach should be pursued for accurate diagnosis of these tumors.

• Therapeutic approach, surgery with pre or post op chemo - radiation can be decided only after definite histological diagnosis.

• The possibility of rare and uncommon malignancies in these locations must be remembered.

REFERENCES

- Bhattacharyya N et al. Incidence and Outcomes of Sinonasal Malignancies. Otolaryngol Head Neck Surg. 2017;157(5):899-906.
- 2. Turner JH et al. Patterns of Care and Survival for Sinonasal Cancer: A National Cancer Database Analysis. Head Neck. 2018;40(8):1704-1714.
- Lund VJ et al. Tumors of the Nasal Cavity and Paranasal Sinuses: Current Perspectives. Curr OpinOtolaryngol Head Neck Surg. 2016;24(2):134-141.
- 4. Dulguerov P et al. Management of Nasal Cavity and Paranasal Sinus Carcinomas. Cancer. 2001;92(2):270-281.
- Ferris RL et al. Immunotherapy for Head and Neck Cancer: Recent Advances and Future Directions. Oral Oncol. 2020;102:104502.
- Goepfert RP et al. Sinonasal Teratocarcinosarcoma: A Retrospective Review of 14 Cases. Head Neck. 2017;39(9):1811-1817.
- Garden AS et al. Adenoid Cystic Carcinoma of the Sinonasal Tract: Long-Term Outcomes After Radiation Therapy. Cancer. 2013;119(13):2239-2246.
- Amit M et al. Sinonasal Undifferentiated Carcinoma: Long-Term Outcomes. Head Neck. 2018;40(2):541-547.
- Hanna EY et al. Multidisciplinary Management of Sinonasal Malignancies: Craniofacial Resection Outcomes. Cancer. 2011;117(5):1106-1115.
- Hsueh WD et al. Advances in Sinonasal Imaging for Malignant Lesions. Am J Rhinol Allergy. 2019;33(3):239-246.
- 11. Lee J et al. Advances in Targeted Therapy for Sinonasal Malignancies. J Clin Oncol. 2021;39(4):451-461.
- Ferris RL et al. Advances in Targeted and Immunotherapy for Head and Neck Cancer. Oral Oncol. 2020;102:104502.
- 13. Van Zante A et al. Recurrence and Survival in Adenoid Cystic Carcinoma of the Sinonasal Tract: Long-Term Analysis. Laryngoscope. 2017;127(2):380-386.
- 14. Goepfert RP et al. Sinonasal Melanoma: Multimodal Treatment Outcomes. Cancer. 2020;126(12):2675-2683.
- Jager MJ et al. Recent Advances in Immunotherapy for Sinonasal Malignancies. Lancet Oncol. 2022;23(2):151-164.