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Case Report

Malignant peripheral nerve sheath tumor (MPNST) of the nasal cavity-A rare presentation

Gowthami Nagendhran[1]*, Mohd. Rafey[1], SH Arif[1], Mahboob Hasan[1], Syed Iqra Usman[1]

[1]Dept. of Pathology, Jawaharlal Nehru Medical College, AMU, Aligarh, Uttar Pradesh, India

Abstract

Malignant peripheral nerve sheath tumors are malignant tumors arising from or differentiating from the peripheral nerve sheath cells. These tumors are more common in the upper extremities and trunk. MPNSTs arising in the head and neck region are rare, among which the nasal cavity and paranasal sinuses are the rarest locations. Diagnosing these tumors is challenging due to nonspecific clinical, radiological findings and histomorphological overlap; however, immunohistochemistry helps to distinguish them. Here, we present a case of a male patient in his 50s with a malignant peripheral nerve sheath tumor located in the nasal cavity.

Keywords: Malignant peripheral nerve sheath tumor, Nasal cavity, Head & neck.

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1. Introduction

Malignant peripheral nerve sheath tumor is an extremely rare malignant tumor of the nasal cavity and paranasal sinuses.[1,2] MPNSTs account for 5-10% of all malignant soft tissue tumors, with about 8-16% occurring in the head and neck region. The nasal cavity and paranasal sinuses are the rarest in the head and neck region.[3]

MPNSTs are more common in young adults and middle-aged adults. These tumors pose topographical problems and present a clinicoradiological and histological polymorphism.1 Approximately half of the cases are associated with neurofibromatosis type 1. The optimal treatment includes surgical excision and adjuvant radiotherapy; however, the role of chemotherapy remains controversial.[4]

2. Case Report

A male patient in his 50s presented as an outpatient in the department of otorhinolaryngology with complaints of nasal obstruction and dyspnea for 3 months, bleeding nose on and off for 2 months. Clinical examination revealed a firm, whitish mass filling the right nasal cavity with a nasal septum deviated towards the left. Then, the patient was advised for radiological investigations. Contrast-enhanced Computed Tomography of the nose and paranasal sinus revealed a hypodense soft tissue density lesion filling the right anterior and mid ethmoidal air cells, right maxillary sinus, and right nasal fossa extending up to right choana with left-sided deviated nasal septa (**Figure 1**).

Then, the biopsy was taken from the abnormal area and sent to the histopathology section in the Department of Pathology, JNMC, AMU. The specimen was put in 10% BNF overnight, processed, and the slide was examined. Hematoxylin and eosin-stained sections on low-power examination showed cellular and relatively hypocellular areas of atypical spindle cells arranged in short fascicles along with areas of necrosis, hemorrhage, and inflammatory infiltrate (**Figure 2**). High-power examination of the same section revealed ovoid to spindle-shaped atypical pleomorphic cells having hyperchromatic spindle-shaped nuclei and vacuolated eosinophilic cytoplasm. (**Figure 3**) Atypical mitotic activity was noticed. Based on morphology, possible differentials were poorly differentiated carcinoma,

Corresponding author: Gowthami Nagendhran
Email: gowthamiparimala@gmail.com

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leiomyosarcoma, melanoma, and MPNST. A battery of immune markers was applied, including P40, Pan ck, SMA, HMB-45, S-100 and H3K27me3. S100 showed strong diffuse cytoplasmic positivity in tumor cells (**Figure 4**); however, p40 (**Figure 5**), Pan ck, SMA and HMB-45 were negative in tumor cells. Further loss of H3K27me3 (**Figure 6**) confirmed the diagnosis of Malignant peripheral nerve sheath tumor (MPNST). The patient was then transferred to the radiation oncology department for further management.

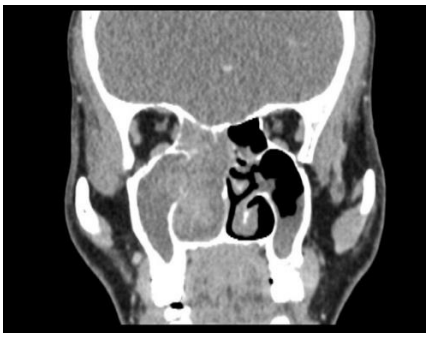

Figure 1: Contrast enhanced CT scan (Coronal view) shows hypodense soft tissue lesion filling maxillary sinus, right nasal cavity and right ethmoidal sinus along with sinonasal polyposis.

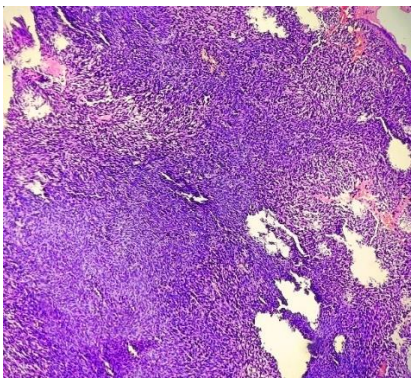

Figure 2: Low power examination shows cellular and relatively hypocellular areas showing proliferation of spindle cells. (H&E-10X)

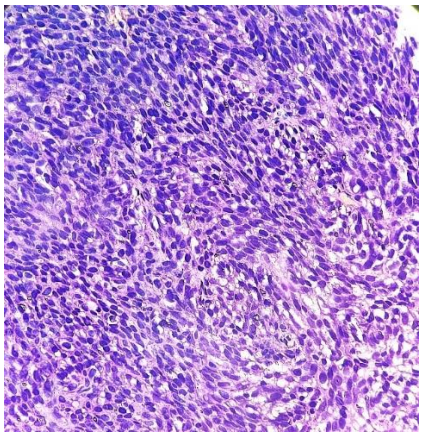

Figure 3: High power examination shows proliferation of atypical pleomorphic spindle cells with hyperchromatic nucleus, vacuolated eosinophilic cytoplasm. (H&E-40X)

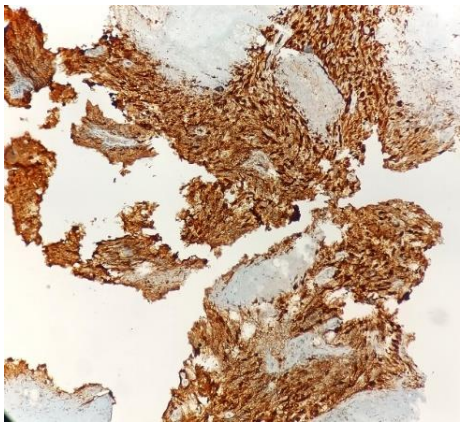

Figure 4: Low power examination shows diffuse cytoplasmic positivity for S100 in tumor cells. (IHC-10X)

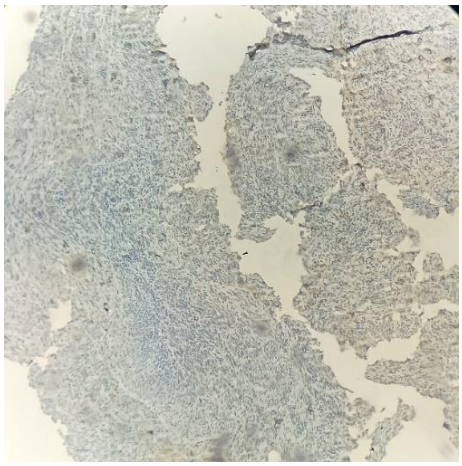

Figure 5: Low power examination shows negativity for p40 in tumor cells (IHC-10X)

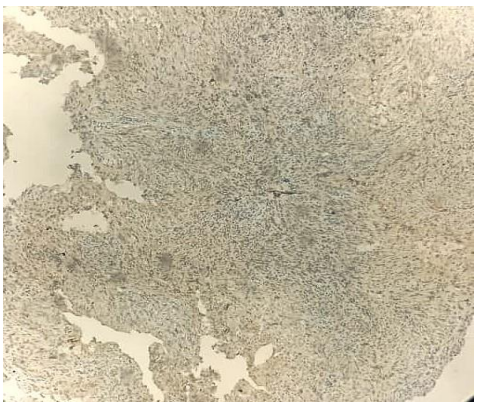

Figure 6: Low power examination shows loss of H3K27me3 in tumor cells (IHC- 10X)

3. Discussion

Malignant peripheral nerve sheath tumors or MPNSTs are malignant spindle cell tumors that arise from nerve sheath cells: Schwann cells, perineural fibroblasts, or fibroblasts. MPNSTs are rare, comprising 5-10% of sarcomas and a global incidence of 0.001% affecting individuals between the third and fourth decades.[1-3] They occur mainly in the roots of the limbs and trunk, less commonly in the head & neck

region. However, the location of nasal cavities and paranasal sinuses is infrequent. It is isolated in 30% of cases, while 70% are associated with type 1 neurofibromatosis (NF1).[4] Neurofibromatosis type 1 (Von Recklinghausen's disease) is an autosomal dominant disorder predisposing individuals to various benign and malignant tumors. The characteristic features include café- au- lait macules, lisch nodules, intertriginous freckling and neurofibromas. Malignant transformation is usually associated with deep neurofibromas.[4]

Symptoms of MPNSTs developing in the nasal cavity and paranasal sinuses are non-specific, mainly related to mass effect.[2] The symptoms may include unilateral nasal obstruction, epistaxis, hyposmia, hypoaesthesia, atypical pain or swelling of the nasal region, headache, rhinorrhea, and dyspnea.[3,4] CT and MRI images are fundamental to evaluating tumors in this anatomical area. CT is useful for assessing tumor extension and eventual metastasis. While MRI can reveal the nerve of origin, the topographical relationship of the tumor with neighboring structures like vascular, muscular structures, and fat plane involvement. MRI distinguishes the lesion from fat tissue better than CT.[5,6]

The diagnosis is made mainly by histopathological and immunohistochemical examination. Histological examination shows dense fasciculated proliferation of spindle-shaped cells with abundant cytoplasm and hyperchromatic, tapered, elongated nuclei.[1] Most often, the tumors are composed of relatively hypocellular areas alternating with hypercellular areas, showing perivascular attenuation and mottled appearance on low-power examination. The myxoid background is seen in 10% of cases.[3] Histomorphologically, these tumors, particularly in the head and neck location, can be confused with spindle cell carcinoma, melanoma, and spindle cell sarcomas like monophasic synovial sarcoma, leiomyosarcoma, and fibrosarcoma.[8] Here comes the role of immunohistochemistry in distinguishing MPNSTs from their mimicers. MPNST shows positivity for S100 and SOX10 in 30-50% of cases. Loss of H3K27me3 is seen in 50% of cases. Other IHCs like SMA, CD34, P40, and P63 are negative.[2,3] Spindle cell carcinoma reveals strong cytoplasmic positivity for Pan ck, while melanomas are positive for HMB-45. Similarly, synovial sarcoma, leiomyosarcoma are positive for CD-99 and SMA respectively.

Early diagnosis is the key to successful treatment, leading to more prolonged survival. A clinical diagnosis of a mass lesion in the nasal cavity should be confirmed by biopsy for definitive diagnosis.[7] MPNSTs are often underrecognized, and the initial diagnosis is missed.1 These tumors have a poor prognosis and a high chance of local recurrence ranging from 40-65%.[4] The treatment of choice includes wide surgical resection with or without adjuvant radiotherapy.[2,4,0,10] However, complete resection of tumors in the head and neck region is not always possible due to its proximity to vital structures in whom adjuvant radiotherapy is recommended. The role of chemotherapy in this tumor is controversial.[1-4] Poor prognostic factors include association with NF1, large size, high grade, metastasis, head and neck location, positive surgical margins, and rhabdomyosarcomatous differentiation. In head and neck MPNST, 5-year survival is 15-47%.[2,4]

4. Conclusion

In conclusion, malignant peripheral nerve sheath tumors (MPNSTs) of the nasal cavity are extremely rare, posing significant diagnostic and therapeutic challenges. Due to their aggressive nature and potential for local invasion and recurrence, early diagnosis and a multimodal treatment approach, including surgical resection with clear margins, radiotherapy, and, in some cases, chemotherapy, are crucial for better outcomes. This case highlights the importance of considering MPNST in the differential diagnosis of nasal cavity tumors, along with the diagnostic importance of immunohistochemistry. It underscores the need for further studies to establish optimal management strategies for this rare malignancy.

5. Source of Funding

None.

6. Conflict of Interest

None.

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