International Journal of Otorhinolaryngology and Head and Neck Surgery
Jagtap SK et al. Int J Otorhinolaryngol Head Neck Surg. 2023 Apr;9(4):310-313
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20230762

Case Series

A case series of rare sinonasal tumors

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Received: 07 January 2023
Revised: 13 March 2023
Accepted: 14 March 2023

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ABSTRACT

Sinonasal tumors often mimick inflammatory diseases. If misdiagnosed, results are life threatening. This article comprises of study done on 5 patients who presented with nasal mass. They underwent clinical examination, diagnostic nasal endoscopy, radiology and histopathological examination (HPE). Thorough evaluation helps the surgeon diagnose rare cases thus preventing detrimental outcomes. Herein we describe our experience with rare tumors like glomangiopericytoma, embryonal rhabdomyosarcoma, small cell sinonasal carcinoma, inverted papilloma and sinonasal undifferentiated carcinoma (SNUC).

Keywords: Glomangiopericytoma, Embryonal rhabdomyosarcoma, Small cell sinonasal carcinoma, Inverted papilloma, SNUC

INTRODUCTION

Sinonasal tumors represent less than 3% of head and neck cancers and 0.8% of all human cancers.[1] Approximately 55% originate in the maxillary sinuses, 35% in the nasal cavity, 9% in the ethmoid and 1% in the frontal and sphenoid sinuses.[2] The symptoms are identical to those of inflammatory diseases.[1] Due to low incidence of these tumors, there are difficulties in definitive diagnosis and adequate treatment. The aim of our study is to describe rare sinonasal tumors like glomangiopericytoma, embryonal rhabdomyosarcoma, small cell sinonasal carcinoma, inverted papilloma and SNUC.

CASE SERIES

Case 1

A 62-year-old female had pinkish red mass in left nasal cavity since 3 months. Magnetic resonance imaging of paranasal sinuses (MRI-PNS) (Figure 1) showed a vascular lesion involving left nasal cavity. Digital subtraction angiography (DSA) showed feeders from left maxillary artery, which was embolized. Endoscopic excision (Figure 2) was done. HPE was suggestive of glomangiopericytoma. The patient is in follow up since 1.5 years.

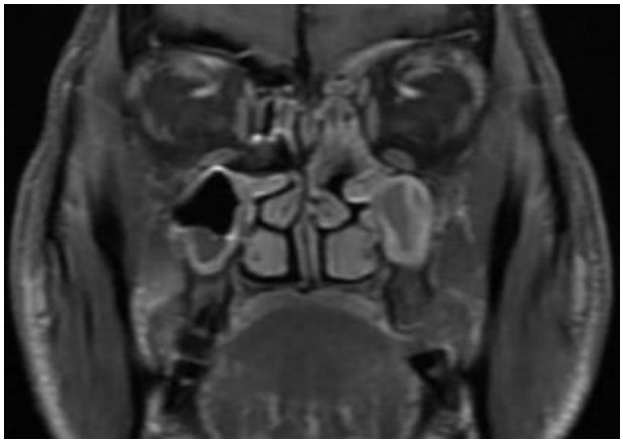

Figure 1: MRI PNS (Coronal view) showing hyperintense lesion with post contrast enhancement measuring 53×14×37 mm in left nasal cavity.

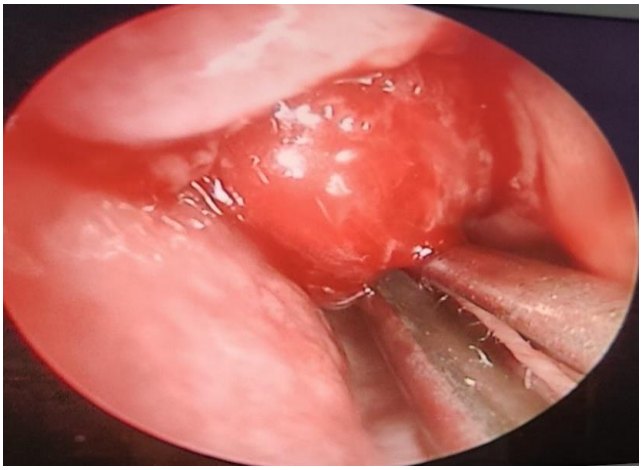

Figure 2: Endoscopic excision of glomangiopericytoma.

Case 2

A 50-year-old female had reddish left sided nasal mass and intermittent epistaxis since 2 months. CT PNS (Figure 3) and MRI (PNS + brain) (Figure 4) showed tumor in left nasal cavity with bone destruction extending in left basifrontal lobe. HPE revealed small cell carcinoma (SCC). The patient was referred for radiation therapy and chemotherapy.

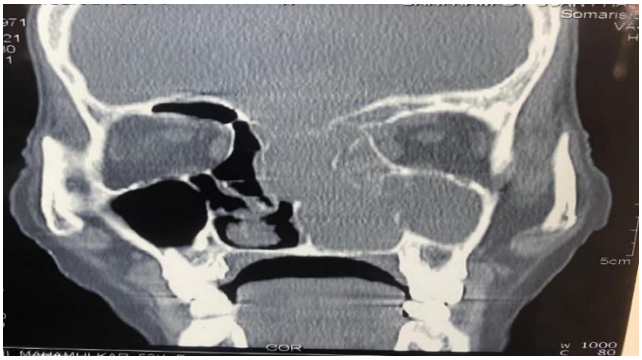

Figure 3: CT PNS (coronal view) showing extensive soft tissue opacification involving left nasal cavity and paranasal sinuses with erosion of underlying bone.

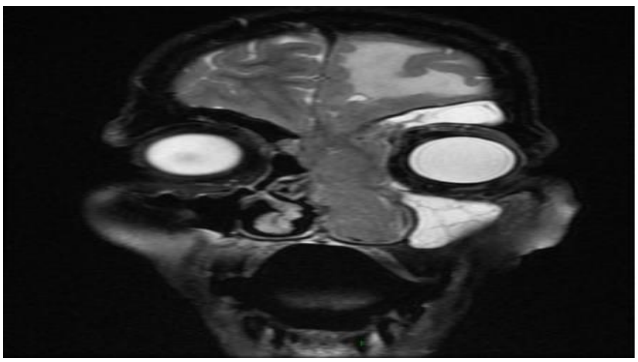

Figure 4: MRI PNS + brain (coronal view) showing extension of the SCC into the left frontal lobe.

Case 3

A 34 years old male had right nasal mass, epistaxis, proptosis since 1 month. CT (Figure 5) and MRI (PNS + brain) (Figure 6) showed lesion involving right sinuses causing bony destruction, extending in the right extraconal compartment. HPE was suggestive of embryonal rhabdomyosarcoma. He is receiving chemotherapy; VAC regimen (Inj. Vincristine 2 mg IV, inj. actinomycin 2.5 mg IV, inj. cyclophosphamide 1 gm IV).

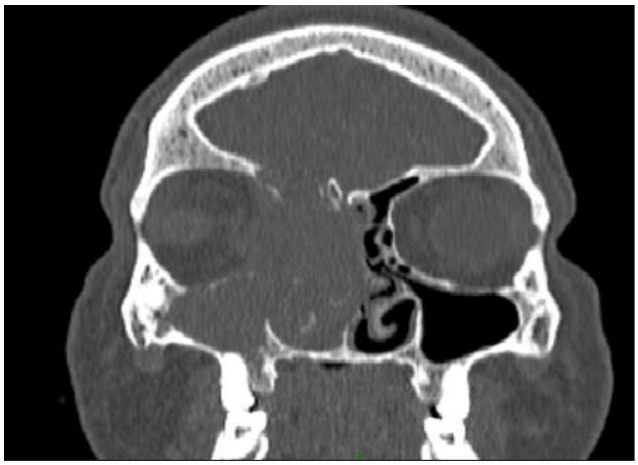

Figure 5: CT PNS (coronal view) showing right nasal mass causing erosion of adjacent bone and extension into right extraconal compartment.

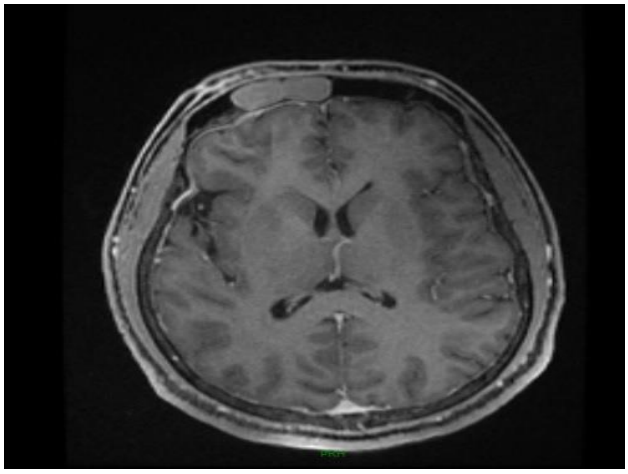

Figure 6: MRI brain (axial view) of extension of the tumor upto the frontal lobe.

Case 4

A 45-year-old male came with single, pale mass occupying the right nasal cavity. Nasal endoscopy was suggestive of sinonasal polyposis. CT PNS (Figure 7) showed mass arising from right maxillary sinus causing widening of the osteomeatal unit. Endoscopic exicison was done (Figure 8). HPE revealed diagnosis of inverted papilloma.

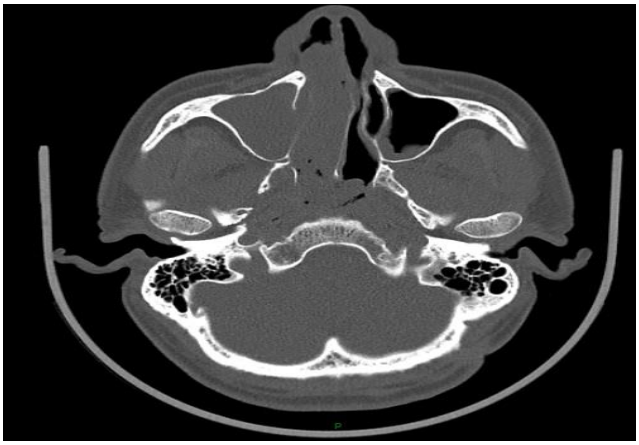

Figure 7: CT PNS (axial view) showing polypoidal mass extending from right maxillary sinus with significant widening of the right osteomeatal unit.

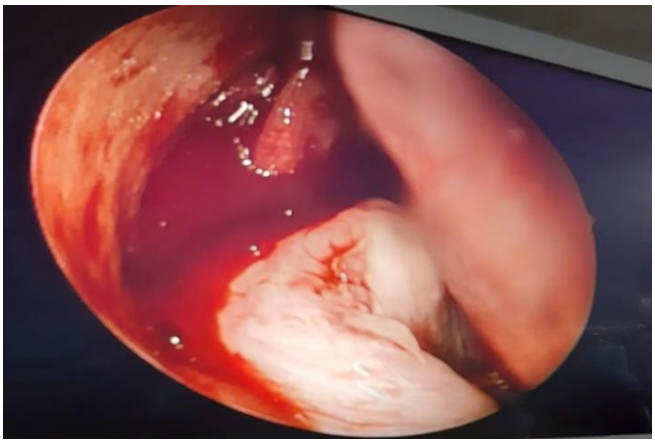

Figure 8: Endoscopic excision of inverted papilloma.

Case 5

A 78-year-old male, came with proliferative mass in left nasal cavity since 15 days. CT PNS reaveled a mass arising from left maxillary sinus causing bony erosion. HPE (Figure 9) was suggestive of SNUC. Patient was advised to undergo chemo-radiation therapy .

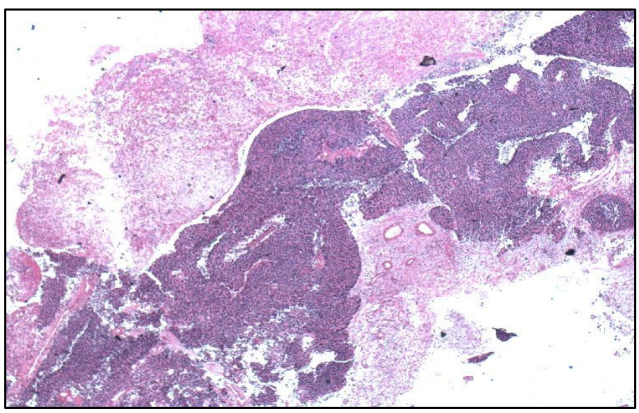

Figure 9: HPE (low power 10x) of SNUC.

DISCUSSION

Glomangiopericytoma

Glomangiopericytoma (GPC) is a rare tumor arising from the periyctes surrounding capillaries and accounts for less than 0.5% of all sinonasal tumors.[3] It mimics the macroscopic appearance of common inflammatory polyps. Local invasion, regional destruction and metastatic spread have been reported. There is good prognosis after surgical excision.[4] We had similar experience in our case.

SCC

SCC is one of the most aggressive types of neuroendocrine carcinomas. In a study by Wang et al the median survival time of this group of patients was 11 months, nearly half of patients have tumor recurrence and/or distant metastasis.[5] There is high incidence of locoregional failure and distant metastasis. A multidisciplinary treatment approach with aggressive complete surgical excision (if feasible), followed by multiagent chemotherapy and radiotherapy, is recommended.[6] We had similar experience in our case.

Rhabdomyosarcomas

It is a soft tissue malignancy of pediatric population, and extremely rare in adults. Turner et al published a retrospective review showing the 5-year relative survival was 49.1%.[7] Prognosis is poorer in adults than children. Histologically, three types are distinguished: embryonic, alveolar and pleomorphic. The embryonic type is most common in ENT, mainly affecting children under 5 years of age and has the best prognosis. Alveolar type mainly affects children over 5 years of age, adolescents and young adults, and has poorer prognosis. The pleomorphic form is rarer, affects adults and has poor prognosis.[8]

Surgery followed by radio-chemotherapy is the choice in locally advanced and metastatic forms. [6] We had a similar experience in our case.

Inverted papilloma

Inverted papilloma is a benign epithelial growth in the underlying stroma of the nasal cavity; Allergy, chronic sinusitis and viral infections are possible causes. It is known for its invasiveness, tendency to recur and association with malignancy. It comprises of 0.5-4% of all primary nasal tumor. It usually presents in male patient of fifth and sixth decade of life.[9] The recurrence rates of 40–80% are unacceptably high. In our case even after 6 months there has not been recurrence so afar.

SNUC

SNUC is described by Frierson et al., as an aggressive neoplasm that was clinico-pathologically distinct from

other poorly differentiated malignancies of the nasal cavity and sinuses.[10] They are highly invasive tumors involving multiple sinuses as well as adjacent structures with high rate of regional recurrence and distant metastasis. The treatment includes multimodality therapy, including surgical resection and adjuvant chemo-radiotherapy. The prognosis of SNUC is poor, death often occurs within short periods following diagnosis.[11,12] Most of the findings are consistent with our case.

CONCLUSION

Sinonasal masses may be due to development anomalies, infectious diseases, inflammatory processes, trauma or neoplasm. Due to lack of specificity in the presentation of rare tumors, one must be attentive to the diverse number of diseases of this region, a thorough clinical examination with radiology and histopathology is necessary for specific diagnosis and prevent complications. By reporting these rare cases we are contributing to data pool of nasal tumors where lack of reporting is major obstacle.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Jagtap SK, Nemade SV. A case series of rare sinonasal tumors. Int J Otorhinolaryngol Head Neck Surg 2023;9:310-3.