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A RARE CASE OF SINONASAL TERATOCARCINOMA - A CASE REPORT

Otorhinolaryngology

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ABSTRACT

Sinonasal malignancies (SNTCS) comprise about 1% of all and 3% of head and neck malignancies. (1) They are rare and have various histological subtypes. The most common type is squamous cell carcinoma followed by adenoid cystic carcinoma. (2,3) Teratoma is the most common germ cell tumour, which consists of mature & immature or a mixture of both elements. They are commonly seen in gonads. Sinonasal teratomas are very rare and mostly benign. Malignant teratomas of the sinonasal region are extremely uncommon & rarely reported in literature. In this article, we have described the presentation and management of sinonasal teratocarcinoma in a 66-year-old male.

KEYWORDS

Sinonasal, Teratocarcinoma, Epistaxis, Rhinology

INTRODUCTION

Sinonasal teratocarcinosarcoma is a rare and aggressive malignancy of the nasal cavity and paranasal sinuses, histopathologically characterized by a combination of teratomatous and carcinosarcomatous elements, involving both epithelial and mesenchymal components. (2) This complex histology contributes to its rapid progression and invasive nature. Clinically, SNTCS presents with nonspecific symptoms such as nasal obstruction, epistaxis, facial pain, and swelling, with advanced cases involving orbital or cranial invasion. Treatment is multimodal, with surgery being the mainstay of treatment, often followed by radiation therapy, and occasionally chemotherapy for recurrent or metastatic disease. Despite aggressive management, prognosis remains poor due to the high recurrence and metastasis rates.

Case Presentation

A 66-year-old male presented to the Outpatient Department with complaints of left-sided nasal block, intermittent nasal bleed, and nasal discharge, mucoid in consistency for 2 months. The patient had no comorbidities or substance abuse, and no significant family history. Diagnostic nasal endoscopy (Figures 1, 2) showed a pinkish fleshy mass occupying left nasal cavity between the septum and middle turbinate. It did not move with respiration and no cough impulse was seen. On probing the mass, soft to firm in consistency, bleeds on touch and insensitive to touch. The nasopharynx was found free. Right nasal cavity was normal. No paranasal sinus tenderness noted. Oral cavity, throat, Ear and neck examinations were unremarkable.

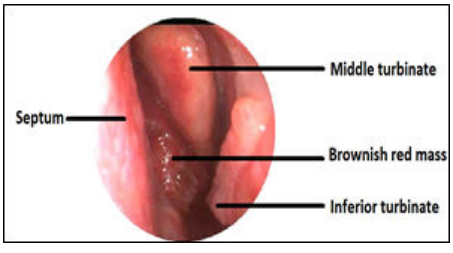


Figure 1

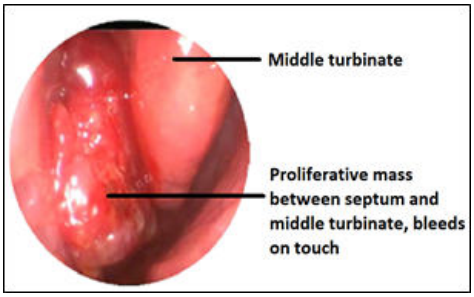


Figure 2

Investigations

Basic blood investigations were done, Hemoglobin - 11.2 g/dl, total count - 8920 cells/cumm, serum electrolytes were within normal limits, Blood urea nitrogen (BUN) was 5 mg/dl and creatinine was 1.5 mg/dl (mildly elevated). In view of elevated creatinine, Contrast-enhanced Computed Tomography (CECT) of nose and paranasal sinuses was not done. Plain CT PNS was done (Figures 3, 4) and showed a hypodense lesion in the left nasal cavity, between the septum and middle turbinate with no involvement over adjacent bony structures or paranasal sinuses with no intraorbital or intracranial extension.

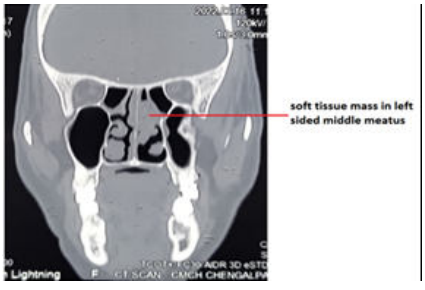


Figure 3

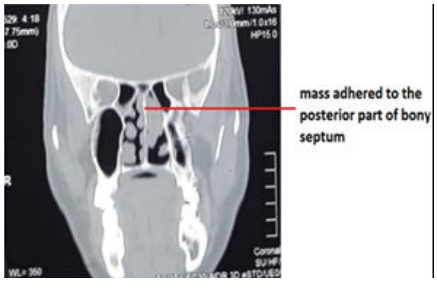


Figure 4

Figures 3, 4: CT Nose and Paranasal sinuses showing the soft tissue mass with homogenous opacity occupying the left side nasal cavity around middle meatus region and attached to the posterior part of bony septum.

Based on these clinical and radiological findings, the differential diagnosis included

1) Infected nasal polyp
2) Pyogenic granuloma
3) Rhinosporidiosis
4) Inverted papilloma

5) Malignant sinonasal mass

A potassium hydroxide (KOH) smear from the left nasal cavity was sent for the detection of Rhinosporidium, but no fungal elements were identified. Clinically, the nasal mass lacked the typical characteristics of a "mulberry-like polypoidal mass" seen in rhinosporidiosis, as well as the "exophytic, polypoidal, frond-like projections with firm to friable consistency" characteristic of inverted papilloma. Additionally, a computed tomography (CT) scan of the nose and paranasal sinuses revealed that the mass was attached to the posterior part of the bony septum, with no involvement of the paranasal sinuses or adjacent bony structures. Based on these findings, the working diagnosis were an infected nasal polyp, a pyogenic granuloma or a sinonasal malignancy. Consequently, planned for Endoscopic excision of the nasal mass under general anaesthesia.

Treatment

The patient underwent Endoscopic excision of a left nasal mass under General Anaesthesia. Intraoperatively, the mass was located between the septum and the middle turbinate, with its attachment at the posterior end of the septum. The mass was excised in its entirety (Figure 5) and sent for histopathological examination (HPE).

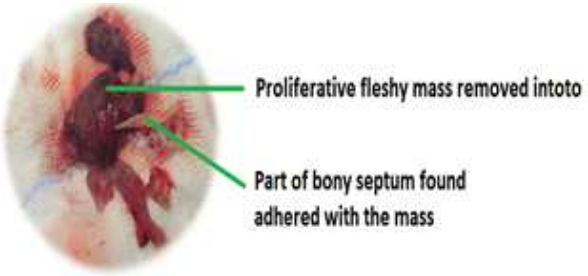


Figure 5: The gross specimen of the left nasal mass.

Histopathological examination (Figures 6, 7) revealed respiratory mucosa with an underlying neoplasm arranged in sheets and nests, composed of cells with hyperchromatic nuclei and scant cytoplasm, with foci of necrosis. These findings were suggestive of poorly differentiated malignancy. Immunohistochemistry (IHC) (Figures 8, 9, 10, 11) was performed using markers such as CK, CD45, p63, Ki67, INSM1, synaptophysin, and chromogranin. CK positivity indicated the presence of epithelial cell lineage, while CD45 negativity confirmed the non-hematopoietic origin of the cells. The p63 marker indicated the presence of myoepithelial cells, and INSM1 focal positivity confirmed the presence of neuroendocrine cells in the head and neck region. The Ki67 labelling index was 60%, and both synaptophysin (an integral membrane glycoprotein) and chromogranin were negative, which further supported the diagnosis of a neuroendocrine tumour. These IHC findings confirmed the diagnosis of sinonasal malignant teratocarcinoma. Following this diagnosis, the case was discussed in a multidisciplinary team (MDT) meeting. Based on the decision, a PET-CT with MR brain fusion was performed (Figures 12, 13, 14). Tumour markers, including serum AFP, LDH, and beta HCG, were measured and found to be within normal limits. The patient was advised to undergo radiation therapy and subsequently completed postoperative intensity-modulated radiation therapy (IMRT) of 60 Gy in 30 fractions over two months. The patient has been on regular follow-up for the last 2 years, with no evidence of disease recurrence.

Immunohistopathology

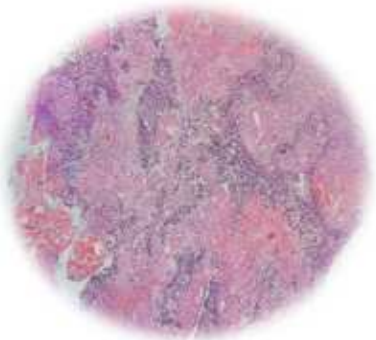


Figure 6: Low power view showing viable tissue and necrotic areas the tumor cells are arranged as sheets intermingled with necrotic and hemorrhagic areas.

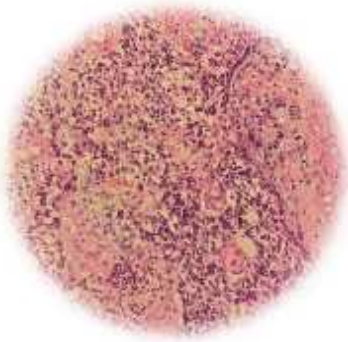


Figure 7: High power view, tumor cells are arranged as sheets and nests. Individual tumor cells showing marked pleomorphism, mitotic figure,hyperchromatic nuclei with eosinophilic cytoplasm shows scant to moderate resembling squamous cells.

Immunohistochemistry

Figure 8: INSM1

Figure 9: CD 45

Figure 10: Ki 67

Figure 11: P63

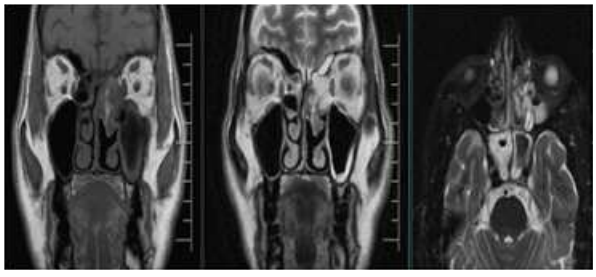
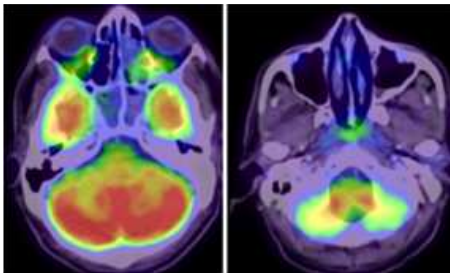


Figure 12: PETCT - Non FDG Avid mucosal thickening in left ethmoidal air cells and bilateral sphenoidal sinus



Figures 13, 14: Axial view of Nose and Paranasal sinus in PET CT - Showed no uptake

DISCUSSION

Dr. Shanmugaratnam et al. first introduced the term "malignant teratoma" as "teratoid carcinosarcoma." (2) Later, Heffner and Hyams coined the term "teratocarcinosarcoma." This is an aggressive tumour that typically presents in the 4th to 5th decade of life, with a male-to-female ratio of 7:1. The most common site of sinonasal malignant teratocarcinoma is the nasal cavity, followed by the ethmoid and maxillary sinuses. Foong et al. also described the aggressive nature of the malignancy and relevant immunohistochemistry markers. (3)

The primary presenting symptoms include nasal obstruction and epistaxis. Grossly, these tumours appear as friable to firm, brownish-red masses that bleed upon contact. Histologically, they are characterized by a combination of mesenchymal and epithelial tissue components with variable growth patterns. The epithelial components include glandular or ductal structures lined by ciliated columnar epithelium, with transitional areas to non-keratinizing squamous epithelium. Clear cells may also be observed. Additionally, these tumours often exhibit features of both squamous carcinoma and adenocarcinoma.

The mesenchymal components include fibroblasts or myofibroblasts, both benign and malignant, benign cartilage with an immature appearance, chondrosarcoma, or osteogenic tissue. Teratoid elements, such as “fetal-appearing” squamous epithelium (clear cells), organoid structures, neurofibrillary matrix, and neural tissue in the form of neural rosettes, are also present. The "fetal-appearing" squamous epithelium is a characteristic histologic finding, indicative of the teratoid nature of the tumor. Immunohistochemical staining typically shows cytokeratin (CK) and epithelial membrane antigen (EMA) positivity in the epithelial components, while neuroepithelial components stain positive for NSE, CD99, chromogranin, synaptophysin, glial fibrillary acidic protein, and S-100 protein. Mesenchymal components are positive for vimentin, and in some cases, may react to myogenic markers or smooth muscle actin. In our case, histopathological examination revealed immunohistochemical staining that showed focal positivity for CK, p63, INSM1, and synaptophysin. CD45 and chromogranin were negative, with a Ki67 labelling index of 60%. A Ki67 labeling index of 60% indicates a high proliferation rate, contributing to the tumor's aggressive nature. (5) The mainstay of treatment for sinonasal teratocarcinoma involves surgical excision followed by radiotherapy, particularly for tumors without extensive invasion. (4.6)

CONCLUSION

- Sinonasal teratocarcinoma (SNTCS) are rare aggressive tumours of sinonasal region with poor prognosis.
- Histopathological examination and immunohistochemistry are required for accurate diagnosis.
- Posterior end of septum is the commonest site of origin, followed by maxillary and ethmoid sinuses.
- Treatment comprises of Excision followed by radiotherapy and occasionally chemotherapy.

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