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Case Report

Sinonasal teratocarcinosarcoma: a case report

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ABSTRACT

Sinonasal teratocarcinosarcoma (TCS) is a very rare malignant neoplasm of sinonasal tract with intermixed teratomatous, carcinomatous and sarcomatous elements. While the diagnosis is largely based on tissue analysis and immunohistochemistry, the mode of management demands further study. Surgical resection with or without chemotherapy and radiation therapy is currently the most accepted treatment regimen. Locally aggressive, while also associated with metastatic lesions, SNTCS is not easily resectable owing to its location and possible intracranial extension. Due to its aggressive nature over one-third of TCS tend to recur leading to treatment failure with a mean survival time of 1.9 years. Possible differentials include squamous cell carcinoma, olfactory neuroblastoma, adenocarcinoma, malignant mixed tumor of salivary gland type, undifferentiated carcinoma, malignant craniopharyngioma, mucoepidermoid carcinoma, transitional carcinoma of Schneiderian type and adenosquamous carcinoma. In this report, we present a case of TCS in a 55 years old male patient who presented to us with complaints of hyposmia, blurring of vision, diplopia and epiphora.

Keywords: Chemotherapy, Sinonasal tract, Teratocarcinosarcoma, Telecanthus

INTRODUCTION

Sinonasal teratocarcinosarcoma (TCS) is a very rare malignant neoplasm with intermixed teratomatous, carcinomatous and sarcomatous elements. It is characterized by combined features of immature or malignant teratoma and carcinosarcoma.[1,2] TCS almost exclusively arises in the sinonasal tract.[3]

This highly malignant rapidly aggressive growth was first described by Shanmugaratnam et al in 1983 and was aptly termed as 'Teratocarcinosarcoma' by Hefner and Hyams in 1984.[4,5] TCS occurs predominantly in males (male:female ratio of 8:1) with >90% of the patients present at 35 years and above, with a mean age of 61 years.

Due to its aggressive nature, over one-third of TCS tend to recur leading to treatment failure with a mean survival time of 1.9 years.[6,7] In this report, we present a case of TCS in a 55 years old male patient.

CASE REPORT

A 55 years old male patient presented in the outpatient department with right nasal obstruction since, 3 months. There was significant proptosis of the right eye, which was associated with diplopia and restriction of eye movements. History revealed hyposmia, blurring of vision, diplopia and epiphora from right eye since, 2 weeks. On examination, the patient had telecanthus, broadening of bridge of nose and swelling of right lateral aspect of nose (Figure 1A). On anterior rhinoscopy, a mass was seen filling the right nasal cavity, obliterating the right nasofacial fold (Figure 1B).

The mass was sensitive to touch, covered in necrotic material and did not bleed on touch. It pushed the septum to the left and reached up to the vestibule. Contrast enhanced computed tomography study showed a heterogenously enhancing lesion in the right nasal cavity extending into right ethmoidal sinus superiorly; eroding the bony nasal septum and extending into left nasal cavity

and left ethmoidal sinus. The lesion extends posteriorly into right choana and laterally erodes the medial wall of right maxillary sinus, obstructing right OMC (Figure 2). Superolaterally, the lesion extends to erode crista galli and cribriform plate with extradural extension into right anterior skull base with loss of fat planes and perilesional oedema with respect to the right frontal lobe. With a subfalcine herniation of right frontal lobe into left lobe, the mass measured 7.4×3.6×6 cm. The patient underwent a medial maxillectomy together with excision of space occupying lesion by an anterior craniotomy.

A frozen section and squash cytology sent intra operatively was suggestive of an esthesioneuroblastoma. Definitive tissue diagnosis showed a poorly differentiated malignant neoplasm with primitive neuroepithelial foci, high grade carcinoma and high-grade sarcoma.

Further confirmed by positive Pan CK, p40 (carcinomatous areas), vimentin (spindle cell areas) and synaptophysin (patchy positive in neuroepithelial cells) in immunohistochemistry. The case was reviewed on 10th post-operative day to find that the patient was symptomatically better. CT scan of chest was done 3 weeks post operatively revealed heterogeneously enhancing near circumferential asymmetric wall thickening of upper esophagus at D1-D3 vertebral level. An endoscopic biopsy of the oesophageal lesion revealed squamous cell carcinoma. Initial palliative radiation was administered in the post-operative cavity and to the oesophageal lesion as well. Considering the unique presentation, the patient was advised concurrent chemotherapy for the oesophageal lesion and referred to the department of medical oncology.

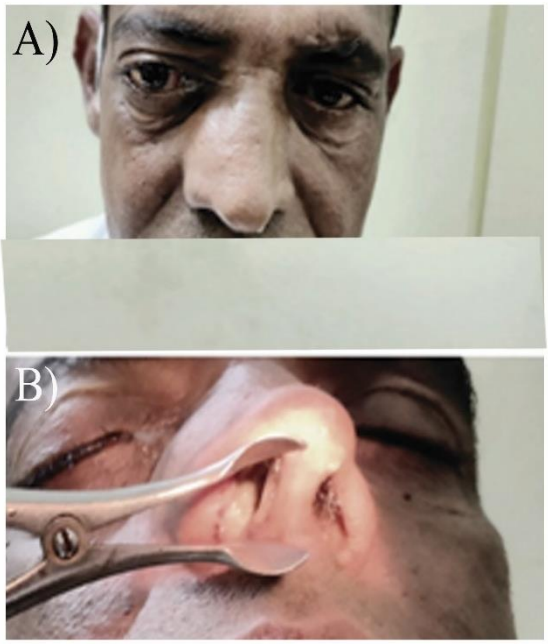

Figure 1: Pre-operative photograph (A) showing right nasal mass extending upto the vestibule; and (B) proptosis of right eye.

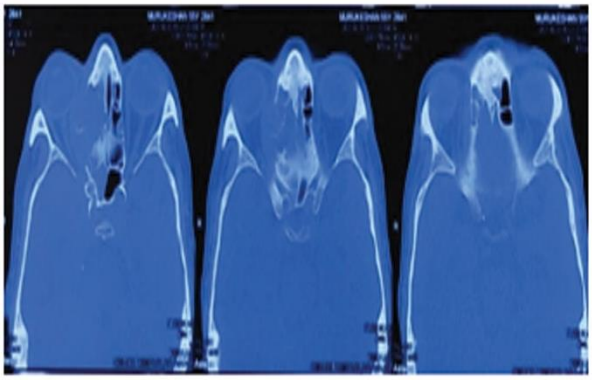

Figure 2: Contrast enhanced computed tomography image showing lesion filling the right nasal cavity.

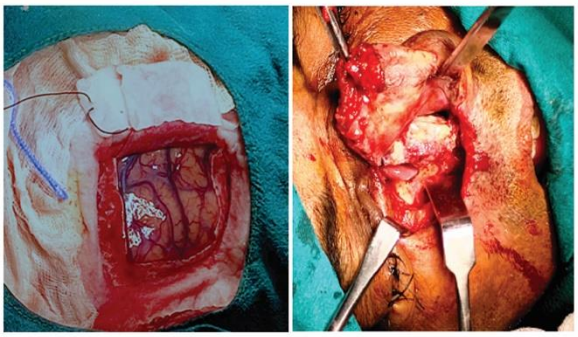

Figure 3: Intra-operative images showing craniofacial resection and excision of lesion. Dural defect was patched.

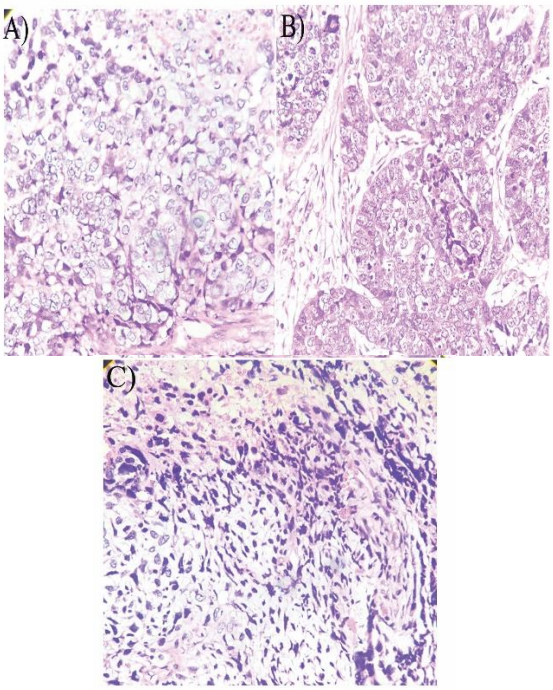

Figure 4: Photomicrographs showing (A) immature squamous (teratoma) component; (B) carcinoma (keratinization) component; and (C) Sarcoma component (H and E, at 40X).

DISCUSSION

TCS is intermixed with teratomatous, carcinomatous and sarcomatous elements. The most common presenting symptoms are epistaxis and nasal obstruction. The patient in this report was presented with a history of nasal obstruction. The diagnosis requires adequate sampling. The conditions that interfere in the diagnosis of TCS include possible squamous cell carcinoma, olfactory neuroblastoma, adenocarcinoma, malignant mixed tumor of salivary gland type, undifferentiated carcinoma, malignant craniopharyngioma, mucoepidermoid carcinoma, transitional carcinoma of Schneiderian type and adenosquamous carcinoma.[4] Therefore, positive staining for PanCK, p40 (carcinomatous areas), vimentin (spindle cell areas) and synaptophysin (patchy positive in neuroepithelial cells) by immunohistochemistry is recommended. The patient in this case report was positive for PanCK, p40, vimentin and synaptophysin. Surgical resection is the first-choice treatment of SNTCS, but it is difficult to ensure an adequate safety margin because of the anatomical location. Therefore, the local recurrence rate is about 40% among surgical patients.[8]

Post-operative radiation therapy is effective to prevent local recurrence.[9] Radiotherapy followed by radical and complete resection is considered the ideal approach to treating a SNTCS. Chemotherapy has been considered for recurrent or extensive cases. Due to the rare nature of the tumor, it is difficult to collect reasonable evidence of an improved cure rate following chemotherapy.

Patients who did not receive postoperative radiotherapy had a higher rate of recurrence as compared to those who had radiotherapy.[10,11] TCS are aggressive tumours and consequently more than half of the patients die within three years. (Heffner/Hyams).

A high recurrence rate of approximately 37% further confirms their aggressive nature. SNTCS is known to be locally aggressive but lung metastases have occasionally been reported.[12] Survival rates at 3 and 5 years is estimated at about 30% and 20% respectively.[5]

CONCLUSION

Given the vast number of differentials that may present with similar symptoms, a greater index of suspicion is required to clinch this rare diagnosis. It is highly likely that smaller tissue sampling may not lead us in the right direction, due to the absence of all the different elements in the tissue sample tested. Adequate tissue for biopsy, histopathological evaluation and immunohistochemistry needs to be done so as not to miss out on the disease. Aggressive chemo-radiation can prove life-saving, if administered timely.

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