Sinonasal Teratocarcinosarcoma - A case report

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Abstract

Sinonasal teratocarcinosarcoma is a very rare tumor of ambiguous origin. Detailed examination of the biopsy material is essential for diagnosis. It is a very aggressive tumor with a five-year survival rate of 45%. Surgery with radiotherapy has been shown to be effective. We report a case of a 38-year old Indian gentleman with left nasoethmoidal mass eroding the cribriform plate with intracranial extension. Subtotal endoscopic excision of the mass revealed features of a sinonasal teratocarcinosarcoma. Immunohistochemistry confirmed the same. The patient was subsequently treated with radiotherapy. The clinical features, histopathological and immunohistochemistry findings and management of sinonasal teratocarcinosarcoma are discussed.

Keywords Teratocarcinosarcoma • Sinonasal Mass.

Case Report

A 38-year-old gentleman presented with left nasal obstruction of two years duration. Diagnostic nasal endoscopy revealed a polypoidal mass seen in the left sphenothmoidal region. CT PNS revealed mass in the left posterior ethmoid and sphenoid sinuses. Under hypotensive general anaesthesia, he underwent complete endoscopic excision of the same. The histopathological examination of the specimen was reported as olfactory neuroblastoma. Patient was advised to have MRI, immunohistochemistry of the biopsy specimen and further radiotherapy but was lost for follow up.

The same patient presented to us two years later with occasional left nasal bleed on and off of one-week duration. Diagnostic nasal endoscopy revealed a fleshy, friable mass occupying the whole of left nasal cavity pushing the septum to the right. MRI with contrast (Fig. 1) revealed left nasoethmoidal mass eroding the cribriform plate with intracranial extension on the left side. With a suspicion of recurrence of olfactory neuroblastoma, endoscopic excision was planned. Under hypotensive general anaesthesia, endoscopic subtotal excision of polypoidal mass occupying the whole of left nasal cavity, nasopharynx, left maxillary sinus and left frontal sinus was removed leaving behind a sleeve of the tumor in the region of left cribriform plate. Post-operative period was uneventful. Patient was discharged home after the removal of nasal packs. He had further follow-up as an outpatient for post-operative endoscopy and discussion of histopathology report.

The histopathological examination of the specimen showed malignant neoplasm composed of epithelial and mesenchymal components. Islands of primitive neuroepithelial tissue with true rosettes (Fig. 2) and pseudorosettes were seen along with cells having vacuolated cytoplasm and focal areas of necrosis. There were clear squamous cells forming glandular pattern (Fig. 3). The mesenchymal components showed loose connective tissue with chondromyxoid appearance. These findings were consistent with sinonasal teratocarcinosarcoma.

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Fig. 1 Left nasosethmoidal mass eroding the cribriform plate

Fig. 2 Stromal areas with a neuroepithelial rosette

Fig. 3 Clear squamous cells forming glandular patterns

Fig. 4 Immunohistochemistry done on the biopsy specimen was positive for

Fig. 4a. cytokeratin specific for epithelial cells,

Fig. 4b vimentin for mesenchymal cells,

Fig. 4c Neuron specific enolase for neuroepithelial cells

Immunohistochemistry done on the biopsy specimen was positive for cytokeratin specific for epithelial cells (Fig. 4 a), vimentin for mesenchymal cells (Fig. 4 b), neuron specific enolase for neuroepithelial cells (Fig. 4 c), S100, glial fibrillary acidic protein X and ESA.

The patient underwent a full curative course of radiotherapy (6000 cGy) over a period of six weeks. Periodic post-operative endoscopies, initially at fortnightly intervals for two months and then every four weeks for six months were done. At the end of six months, he is symptom free but for skin erythema and dryness of mouth secondary to radiotherapy. There is no tumour recurrence or metastasis at the end of one year

Discussion

Sinonasal teratocarcinosarcoma is a very rare tumor with less

than 40 documented cases in world literature. Heffner and Hyams¹ first categorized it in 1984. Earlier they were termed as teratoid carcinosarcoma, blastomas, teratocarcinoma and malignant teratoma.

They are commonly seen in the nasal cavity and can extend into the sinuses, most commonly the ethmoidal or the sphenoidal sinuses. It usually occurs in the age group between 18 to 79 years (median age is 60 years)¹. It is more common in males than in females (4:1). Commonest presentation is nasal obstruction and epistaxis. Other rare presentations are headache, proptosis or neurological involvement. The symptoms are of short duration suggestive of aggressive nature of the tumor. Mortality is due to local invasion.

Exact origin of the tumor is not known. Morphologically, the tumor presents as a red or reddish-purple friable mass with

areas of necrosis. Histopathological examination is the corner stone of diagnosis. The architecture is varied and different tissue types (malignant epithelial and stromal elements) are present in varying proportions. Well formed glands, immature squamous epithelium, atypical stroma may be seen. The epithelial components show glandular/ductal structures lined by columnar epithelium and islands of squamous cells. The glandular epithelium may or may not be carcinomatous. Squamous epithelial cells are clear cells resembling normal fetal epithelium. The stroma may be formed by benign or malignant fibromyxoid, chondroid, osteoid, myoid and primitive neural component with or without fibrillary background. Seminoma, embryonal carcinoma, yolk sac tumor or choriocarcinoma are not seen as a part of teratocarcinoma. The biopsy specimen should be examined thoroughly, otherwise it is easy to miss the diagnosis⁶. Immunohistochemistry helps in diagnosis. Radical excision followed by radiotherapy has shown good results. Individualized multimodal therapy including a histology-specific chemotherapy has been proposed⁵. Combination of radiotherapy and chemotherapy has been reported very effective in the management of sinonasal teratocarcinoma⁷. 5-year survival rate is around 45%².

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