Sino Nasal Teratocarcinosarcoma-An Interesting Clinical Entity

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Abstract Sinonasal teratocarcinoma is a rare tumour of the paranasal sinuses. It was earlier termed as teratoid carcinosarcoma or teratocarcinoma. It is a highly aggressive tumour with a 5 year survival rate of 45 %. Here we report a case of sinonasal teratocarcinoma who was managed excision of tumour by lateral rhinotomy approach followed by radiotherapy.

Keywords Teratocarcinosarcoma · Immunohistochemistry · Lateral rhinotomy

Case Report

A 43-year-old woman presented to the OPD with complaints of left-sided nasal obstruction and nasal bleeding from the left nostril since the past 3 months. She also had complaints of loss of sensation of smell and increased watering of the left eye. She was a known case of hypothyroidism and hypertension and was on regular treatment. Diagnostic nasal endoscopy showed a reddish mass occupying the whole of the left nasal cavity which bled on touch. Sensation over all quadrants of face was normal. Increased lacrimation was present in the left eye. Because of the bleeding tendency of the tumor, a screening diagnostic angiography was performed to rule out any vascular pathology. However, angiography showed an avascular nasal mass. CT scan of paranasal sinuses with contrast showed complete opacifica-

tion of the left nasal cavity and left frontal recess and ethmoidal sinuses (Fig. 1).

Endoscopic guided left lateral rhinotomy approach was used for surgical exploration. The nasal cavity was exposed and the mass was seen occupying the left vestibule extending posteriorly till the left choana and partially to the right choana, abutting the posterior end of the septum (Fig. 2).

The tumor mass was excised en masse in continuity with the normal nasal mucosa in the extramucosal plane along the sphenoethmoid recess and superiorly till the cribriform plate and fovea ethmoidalis. The surgically excised specimen was sent for histopathological examination (HPE).

The HPE showed features suggestive of both carcinoma and sarcoma (Fig. 3). Immunohistochemistry was performed. The tumor cells expressed desmin, myogenin, cyto-keratin, and SMA, and were immunonegative for Mic-2 and synaptophysin.

Postoperatively, the patient was given a full course of radiotherapy with combination of chemotherapy (cisplatin + 5FU) for 6 weeks, and is on regular follow-up. The patient was completely relieved of her symptoms, and a postoperative CT scan taken 6 months later showed no recurrence. Regular follow-up is being done, and the patient was completely symptom free and no recurrence was detected as evidenced by diagnostic nasal endoscopy, 1 year postsurgery.

Discussion

Sinonasal teratocarcinosarcoma is a very rare tumor. Only 35–40 cases are documented in world literature and was first described by Heffner and Hyams [1]. This neoplasm is more common in males, with a male–female ratio of 4:1. It is seen in age groups between 18 and 79 years [2]. It is commonly

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Fig. 1 Coronal CT scan of paranasal sinus showing mass in the left nasal cavity

seen in the nasal cavity and can extend into the sinuses, mainly ethmoidal or sphenoidal sinuses.

Clinically, these patients generally present with epistaxis or nasal obstruction. These patients can also present with headache, proptosis, and neurological involvement. Local invasion of the tumor is very common and can lead to morbidity and mortality. Metastatic lesions are documented in regional lymph nodes and lungs [3, 4].

Morphologically, the tumor presents as a reddish, friable mass with areas of necrosis. Microscopic examination shows malignant epithelial and stromal elements in varying proportions. The epithelial components include glandular or ductal structures. The stromal components may be formed by benign or malignant fibromyxoid, chondroid, osteoid, myoid, and primitive neural component with or without fibrillary background [5]. Squamous epithelial cells are clear cells resembling normal fetal epithelium. Inadequate sampling may lead to errors in diagnosis. Immunohistochemistry is the gold standard of diagnosis.

Fig. 2 Excision of mass by lateral rhinotomy approach

HPE- HEMATOXYLIN AND EOSIN STAINING

Fig. 3 Histopathology showing features of teratocarcinosarcoma

Treatment options include radical excision followed by radiotherapy or a combination of radiotherapy with chemotherapy [6, 7]. This is followed up with a very meticulous, extended follow-up to monitor any recurrences and treat them accordingly.

Conclusion

Sinonasal teratocarcinoma is a rare tumour of paranasal sinus. Histopathological examination and Immunohistochemistry aids in diagnosis. Surgery followed by radiotherapy with or without chemotherapy is the treatment of choice. Early diagnosis and management and good follow up can give a better prognosis

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