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REVIEW ARTICLE

Sinonasal Teratocarcinosarcoma: A Rare Clinical Entity

Sanjeev Mohanty, Gopinath Maraignanam, L Somu

ABSTRACT

Sinonasal teratocarcinosarcoma is a rare tumor of the paranasal sinuses, that shows combined features of both malignant teratoma and carcinosarcoma. It was earlier called teratoid carcinosarcoma or blastoma or teratocarcinoma or malignant teratoma. Its pecularity is its highly aggressive nature with a 5 years survival rate of 45% with treatment according to published statistics. Its treatment modality is a combination of surgery and radiotherapy. Here, we report one such case who presented with a history of one episode of torrential bleeding from left nostril and nasal obstruction for 3 months for which total excision of the tumor was done by endoscopic-assisted left lateral rhinotomy approach. Histopathology and immuno-histochemistry confirmed the diagnosis of sinonasal teratocarcinosarcoma and she was subsequently administered radiotherapy postoperatively.

Keywords: Sinonasal mass, Teratocarcinosarcoma, Radiotherapy.

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CASE REPORT

A 43-year-old female presented to the OPD with complaints of left sided nasal obstruction and nasal bleeding from left nostril since the last 3 months. She also had complaints of loss of sensation of smell and increased watering of 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. Epiphora of left eye was noted.

Because of the bleeding tendency of the tumor, a screening diagnostic angiography was done to rule out any vascular pathology. However, angiography showed an avascular nasal mass. CT scan of paranasal sinuses with contrast showed complete opacification of left nasal cavity and left frontal recess and ethmoidal sinuses (Fig. 1).

Endoscopic-guided left lateral rhinotomy approach was used for surgical exploration. 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 sent for HPE.

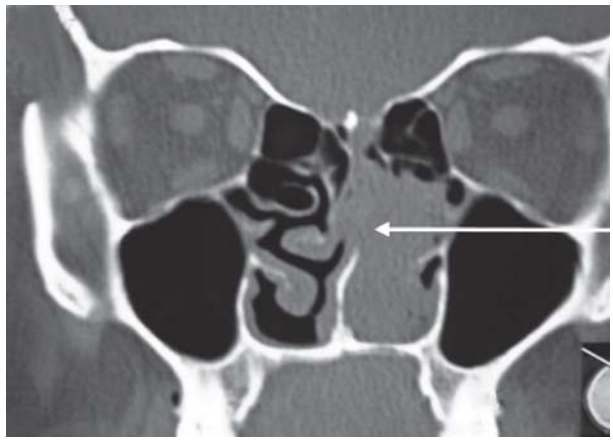

Fig. 1: CT-PNS coronal section showing the mass

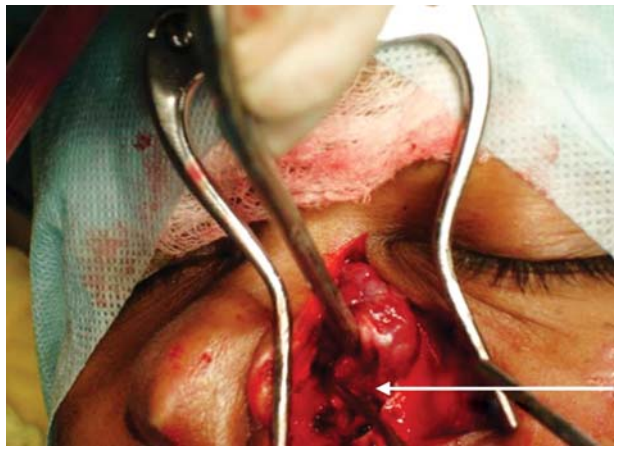

Fig. 2: Mass in the nasal cavity

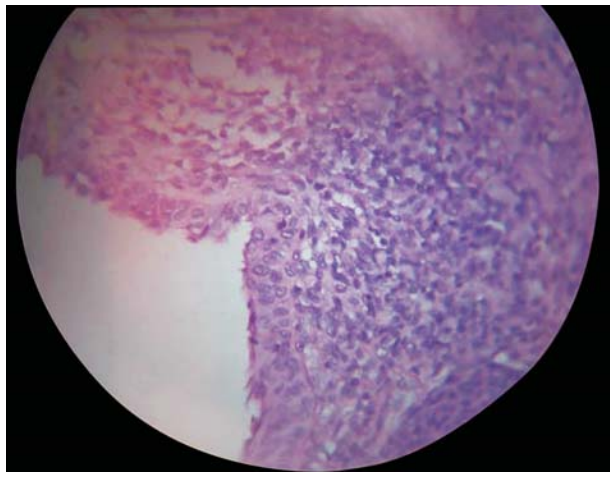

Fig. 3: HPE-hematoxylin and eosin staining

The HPE showed features suggestive of both carcinoma and sarcoma (Fig. 3), and so immunohistochemistry was done. The tumor cells expressed readily Desmin' myogenin, cytokeratin and SMA and were immunonegative for Mic 2 and synaptophysin.

Inspite of a complete clearance surgically the patient was given a full course of radiotherapy with combination of chemotherapy (Cisplatin + 5FU) for 6 weeks and was on regular follow-up. 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 this 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 to 40 cases are documented in world literature and was first described by Heffner and Hyams.[1] It is more common in males with male: female ratio of 4:1. It is seen in age groups between 18 and 79 years.[2] It is commonly seen in 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 different tissue types, i.e. malignant epithelial and stromal elements in varying proportions.[5] The epithelial components include glandular or ductal structures. The stromal components may be formed by benign or malignant fibro-myxoid, chondroid, osteoid myoid and primitive neural component with or without fibrillary background.[6] 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. Aggressive management of such an entity is the order of the day. Treatment options include radical excision followed by radiotherapy or a combination of radiotherapy with chemotherapy.[7] This is followed-up with a very meticulous extended follow-up to monitor any recurrences and treat them accordingly.

CONCLUSION

- Sinonasal teratocarcinosarcoma is a rare sinonasal tumor.
- Total excision of tumor is the preferred mode for HPE examination.
- HPE and immunohistochemical correlation is a must for clinching diagnosis.
- Radiotherapy with or without chemotherapy following surgery is mandatory for good clinical outcome.
- Very aggressive follow-up and management is required to detect early recurrences.
- Early diagnosis and management before regional spread of the tumor can give a better prognosis.

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