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Case Report

# Teratocarcinosarcoma: A rare malignant tumor of sinonasal tract

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ABSTRACT

**Background:** Sinonasal teratocarcinosarcoma is an extremely rare malignant tumor arising in the sinonasal tract, having combined histological features of teratoma and carcinosarcoma. Here, we are presenting a case of sinonasal teratocarcinosarcoma in a 44 year-old male patient.

**Case Report:** 44 year old male presented to us with complains of nasal obstruction and intermittent epistaxis since 1 month. On examination polypoidal mass was noted in left nasal cavity which bleeds on touch. On MRI, well defined heterogeneously enhancing lesion completely filling the left nasal cavity was noted. Histopathological examination of resected specimen revealed, malignant epithelial components being adenocarcinoma and squamous cell carcinoma, mesenchymal component composed of spindle cells, along with primitive mesenchymal and blastemal components. Immunohistochemistry showed epithelial and stromal markers positive for Cytokeratin and Vimentin respectively.

**Discussion:** Sinonasal teratocarcinosarcoma (TCS) is a rare and aggressive malignant neoplasm characterized by the combination of malignant teratoma and carcinosarcoma. Most common presentation being nasal obstruction and epistaxis with male preponderance. The tumor shows heterogeneous morphology with varying proportions of benign and malignant epithelial, mesenchymal, and blastemal components. Less than 100 cases have been reported in literature.

**Conclusion:** Teratocarcinosarcoma is a rare aggressive sinonasal tumor. Total excision of the tumor and aggressive sampling for histopathological examination is necessary to avoid misdiagnosis of olfactory neuroblastoma and carcinosarcoma.



## 1. Introduction

Teratocarcinosarcoma is a rare and unusual malignant neoplasm of sinonasal tract with aggressive behaviour.[1] It originates almost exclusively in sinonasal tract.[1] The most common presentation is nasal obstruction and epistaxis. Less than 100 cases have been documented[2] with male predominance with a male to female ratio of 7:1.[3] Overall survival has been reported to be at 1.7 years with 60% mortality within 3 years.[4]

## 2. Case Report

A 44-year-old, male patient presented with complains of nasal obstruction and intermittent epistaxis and headache since 1 month. On examination a polypoidal mass in the left nasal cavity was identified and resection of the same was performed. Patient was also taken up for CT scan to look for metastasis to brain. No evidence of metastasis was noted on CT scan. Gross examination: Received multiple grey black to grey brown friable mass of tissue, largest measuring 2x1x0.5cm and smallest measuring 0.5x0.5x0.1cm. Cut surface appears grey black to grey brown. Entire tissue was processed and subjected to histopathological examination and immunohistochemistry.

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### 2.1. Microscopy

Sections studied from the nasal mass revealed malignant epithelial components composed of glands which are highly pleomorphic with marked cytological atypia suggestive of adenocarcinoma. Also noted are polygonal cells with abundant eosinophilic cytoplasm, nucleus showing irregular nuclear border, vesicular nucleus and prominent nucleoli suggestive of squamous cell carcinoma, mesenchymal component composed of spindle cells with mild to moderate pleomorphism, along with primitive mesenchymal and blastemal components. The blastemal component is composed of small round blue cells suggestive of primitive component.

### 2.2. IHC revealed

EMA and VIMENTIN positive. EMA was positive in the epithelial component, Vimentin was positive for spindle cells suggestive of mesenchymal component. Ki 67 index of more than 50% and suggestive of highly aggressive nature of the tumor.

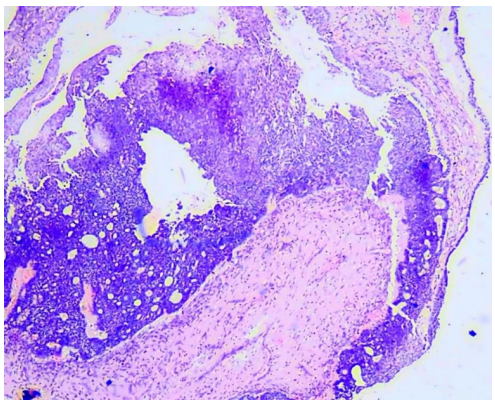

**Figure 1:** Low power view of adenocarcinoma.

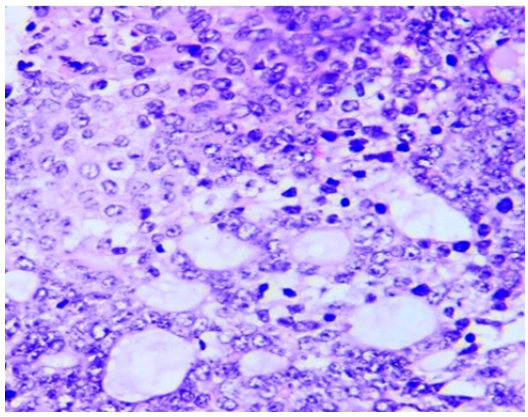

**Figure 2:** High power view of adenocarcinoma

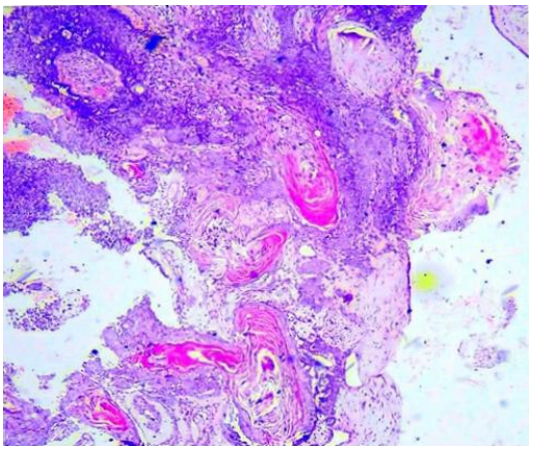

**Figure 3:** Low power view of squamous cell carcinoma

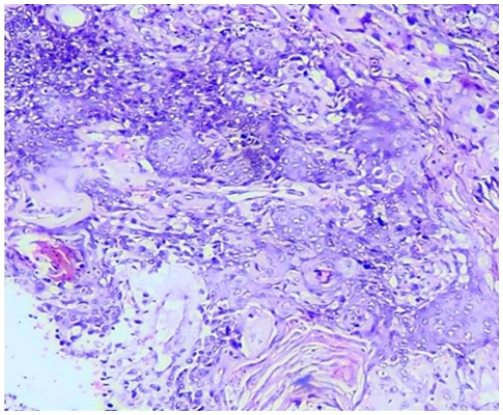

**Figure 4:** High power view of squamous cell carcinoma

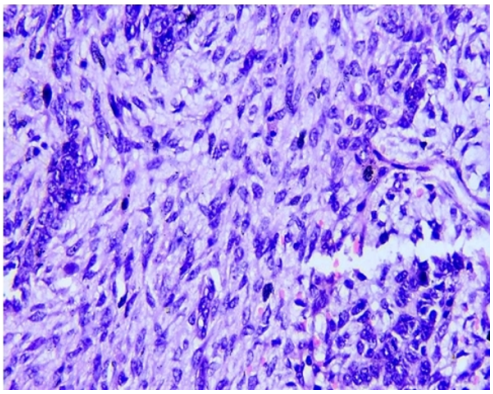

**Figure 5:** Mesenchymal component. High power showing spindle cells

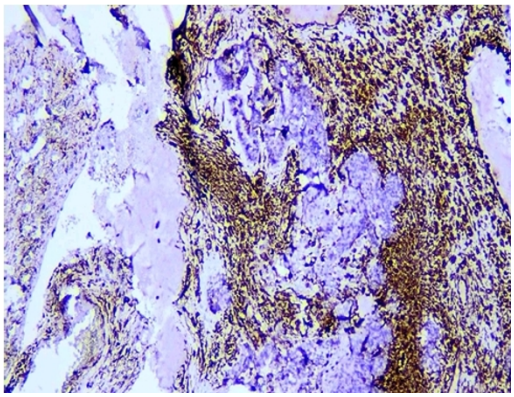

**Figure 6:** Vimentin positive in mesenchymal component.

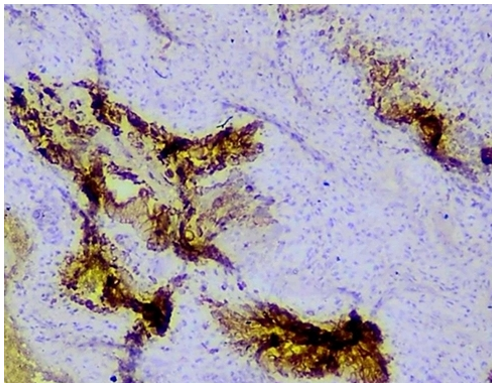

**Figure 7:** EMA positive in epithelial component

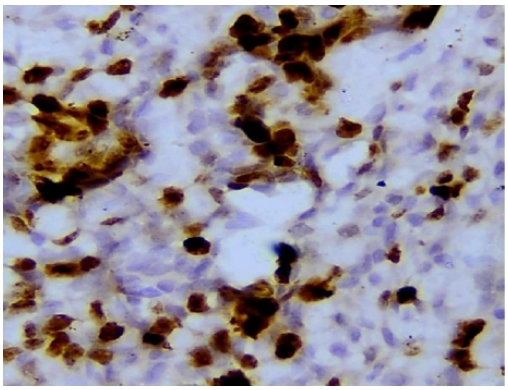

**Figure 8:** Ki67> 50%.

## 3. Discussion

Sinonasal teratocarcinosarcoma (TCS) is an unusual tumor, which arises from paranasal sinuses and nasal cavity. Patients present with epistaxis, nasal congestion and headaches.[5] Tumors arising from paranasal sinuses have diverse histological spectrum. Hence adequate biopsy has to be taken from nasal cavity and paranasal sinuses. Usually local tissue biopsy is inadequate for diagnosis. Sinonasal teratocarinoma have propensity to undergo early metastasis and is associated with poor prognosis.[6]It is rare and aggressive malignant neoplasm characterized by the combination of malignant teratoma and carcinosarcoma.[7] Most common presentation being nasal obstruction and epistaxis with male preponderance. Less than 100 cases have been reported in literature.[2] The tumor shows heterogeneous morphology with varying proportions of benign and malignant epithelial, mesenchymal, and blastemal components.[8]

The epithelial components are highly variable which includes both benign and malignant squamous and glandular components. The mesenchymal components appear as spindle shaped cells. The blastemal components are small round blue cells which are primitive neuroepithelial elements with well formed rosettes. In a few other studies, pathological findings revealed ciliated columnar and non keratinized squamous epithelia on the tumor surface and immature squamous cell nests containing clear cells in core. Mature and cancerous glands and poorly differentiated adenocarcinomic structures with nested cords were also evident. Further more multifocal, poorly differentiated sarcoma like areas around the adenocarcinoma and tumor like hyperplastic fibrous tissue were observed in some areas. Immunohistochemistry can be used in adjunct with histopathological examination to come to a diagnosis. Markers include EMA and Cytokeratin positivity for epithelial component, undifferentiated sarcoma positive for vimentin and primary tumor component positive for CD99 and NSE. CT and MRI features are similar to common malignant sinus tumors, a mass breaching the lamina papyracea and involving the medial rectus muscle causing proptosis. Radiological examination is necessary to rule out metastasis.[9] The treatment consists of surgical resection of tumor and adjuvant radiotherapy[10] Overall survival rate for the tumor is 46%.[11]

## 4. Conclusion

Teratocarcinosarcoma is a rare and aggressive sinonasal tumor. It has wide range of presentation from epistaxis, nasal obstruction, headache to metastasis to lung, presenting as pleural effusion. Total excision of the tumor and aggressive sampling for histopathological examination is necessary. Due to its rarity and overlapping morphologic characteristics, this neoplasm represents a diagnostic challenge. The treatment is a multimodality approach with wide excision of tumor and administration of adjuvant radiotherapy. The overall survival rate is 46% with overall mortality being as high as 60% within 3 years of diagnosis of tumor.

## 5. Conflicts of Interest

None.

## 6. Source of Funding

None.

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