

# An Aggressive Sinonasal Mass

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## ABSTRACT

**INTRODUCTION:** Sinonasal teratocarcinoma (SNTCS) is an extremely rare aggressive malignant tumor arising in the sinonasal tract, often confused with other sinonasal tumors. It is more common in adult males. These tumors are often difficult to diagnose because of their diverse histology.

**CASE REPORT:** We are presenting a case of right sided SNTCS in a 38-year-old male patient. The patient had a history of right sided nasal obstruction associated with recurrent episodes of epistaxis for last 3 months. On examination, a solid mass was noted in the right nasal cavity that was greyish black in colour, irregularly shaped, brittle, exhibits localized surface necrosis and bleeds easily on touch. Microscopic examination revealed an admixture of carcinomatous (glands, non-keratinized squamous epithelia), sarcomatous (atypical spindle cells, smooth muscle), primitive neuroectodermal and teratoid elements (immature squamous and clear cell nests).

**CONCLUSION:** SNTCSs are highly aggressive tumors with poor prognosis. Majority of the patients do not survive beyond 3 years. Complete excision and extensive sampling are necessary for their diagnosis.

However, because of their aggressive nature, aggressive treatment approach of surgery with adjuvant chemo-radiation is more beneficial than surgical excision alone.

**Key words:** adeno-squamous carcinoma, epistaxis, immunohistochemistry, olfactory neuroblastoma, paranasal sinus, sinonasal mass, teratocarcinoma

## INTRODUCTION

Sinonasal teratocarcinomas (SNTCSs) are very aggressive and rapidly progressing tumors associated with poor prognosis <sup>[1]</sup>. They are very uncommon, and typically arises in the nasal cavity and paranasal sinuses (ethmoid and maxillary sinus), but can also extend into the anterior skull base, oral and orbital cavity, nasopharynx <sup>[2]</sup>. Intracranial extensions found in 20% cases <sup>[3,4]</sup>.

It is a tumor of uncertain histogenesis <sup>[4,5]</sup>, that probably arises from pluripotent progenitor cell <sup>[5]</sup>. They usually occur in adult males with M:F ratio of 7:1 to 8:1<sup>[3]</sup> Most common clinical features of SNTCSs are nasal obstruction, epistaxis and headache, when associated with intracranial extensions these tumors may present with neurological symptoms like seizures, facial or ocular pain,

loss of vision, anosmia, epiphora, odynophagia, dysphagia [6,7].

Microscopic examination reveals presence of benign and malignant epithelial, malignant mesenchymal tissue and neural elements including immature tissue with blastema like features. However, choriocarcinoma, embryonal carcinoma or seminoma is typically absent [8,9]. SNTCSs are commonly treated by radical surgical resection followed by radiotherapy and chemotherapy. The recurrence rate of these tumors is 38%, while metastasis occurs in 10.9% cases and both recurrence and metastasis occur in 8.7% of cases [10].

### **CASE REPORT**

A 38-year-old male patient presented with right sided nasal obstruction associated with recurrent episodes of epistaxis for 3 months duration. On examination, an irregular shaped greyish-black coloured solid mass was seen in the right nasal cavity, that bled easily on touch. CT scan was advised, that revealed soft tissue attenuation of a portion of maxillary sinus that was extending to the right nasal cavity.

The mass was resected and sent to the Department of Pathology in 10% neutral buffered formalin. We received a single irregular shaped greyish-black coloured solid soft tissue mass with localised surface necrosis measuring 5cm x 4cm x 2.5cm (Fig.1). The sections were made from paraffin embedded tissue blocks and followed by hematoxylin and eosin staining. Microscopic examination of the sections revealed an admixture of carcinomatous, sarcomatous, primitive neuroectodermal and teratoid elements (Fig.2,3,4,5). Carcinomatous component included non keratinizing squamous epithelia, malignant glandular structures indicating areas of moderately differentiating adenocarcinoma (Fig.6). Sarcomatous component predominantly composed of atypical spindle

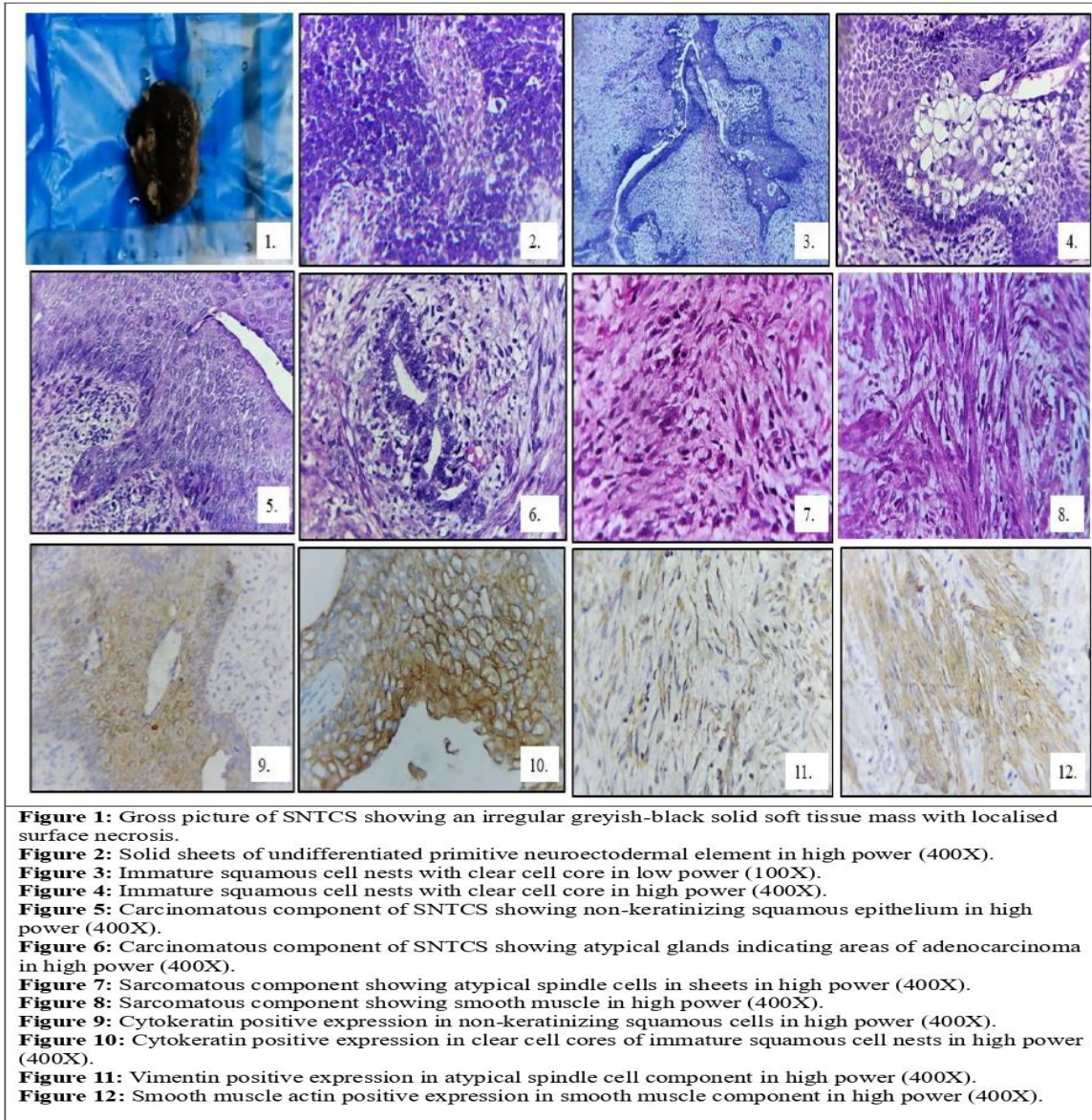
cells in fascicles, sheets encircling the glandular structures and smooth muscle (Fig. 7,8). There were areas of myxoid differentiation and solid sheets of undifferentiated primitive neuroectodermal elements. Immature squamous cell nests with clear cell core indicated the teratoid element of the tumor. Immunohistochemical study for Cytokeratin was positive (Fig. 9,10). Vimentin (Fig.11) and Smooth muscle actin (Fig.12) were positive.

The histological features are consistent with the SNTCS. After receiving the histopathology report wide local excision of the tumor was done. Then the patient was undergone radiotherapy. Now six months have been passed. The patient does not have any recurrence till now. He is doing well.

### **DISCUSSION**

SNTCS is a highly aggressive malignant tumor. The sinonasal SNTCS was diagnosed in 1966 for the first time. Then it was diagnosed as malignant Teratoma of Ethmoidal sinus. Heffner and Hyams first named the tumor "Teratocarcinosarcoma". depending on a clinicopathological study on 20 cases of sinonasal tract tumors. These tumors were previously termed as malignant teratoma, teratocarcinoma, carcinosarcoma, teratoid carcinosarcoma, malignant mixed tumor. SNTCS term was introduced by Shanmugaratnum and Heffner in 1983 and 1984, respectively [9] Only 127 cases have been reported in English literature till date. They are commonly seen in adults, with a male predominance [9].

Our patient was also an adult male. The most common site is nasal cavity and then ethmoid sinus and maxillary sinus [4,5,1]. In this case the tumor was located in right nasal cavity. Grossly these tumors are friable to firm, bulky reddish-brown mass [4,5]. Here the mass was friable and also bled to touch. It was greyish black in color.



Microscopically, SNTCS is composed of a combination of epithelial, mesenchymal, primitive neuroectodermal and teratoid tissue components [9]. The epithelial components included glandular structures lined by columnar epithelium, areas of non keratinizing squamous epithelium, along with areas of squamous cell carcinoma and adenocarcinoma. Mesenchymal components include benign and malignant fibroblasts or myofibroblasts, chondroid or osteogenic tissue. The teratoid components include fetal appearing clear cell, squamous epithelium,

neural tissue in the form of neural rosettes and neurofibrillary matrix. Foci of embryonal carcinoma, choriocarcinoma, seminoma have not been found associated with these tumors [8,9,10,11]. SNTCSs usually lacks specific CT and MRI findings, the findings are similar to those of common malignant sinus tumors [9]. In our case the microscopical examination of the sections revealed an admixture of carcinomatous, sarcomatous, primitive neuroectodermal and teratoid elements. Carcinomatous component included non keratinizing squamous epithelia, malignant



glandular structures. Atypical spindle cells were arranged in fascicles, sheets.

Rarity of SNTCSs and inadequate sampling often leads to misdiagnosis of these tumors as Olfactory neuroblastoma, squamous cell carcinoma, undifferentiated carcinoma, adenocarcinoma, adeno-squamous carcinoma, malignant salivary gland type tumors [10,12].

Immunohistochemical examination helps in the diagnosis of these tumors. Epithelial components are cytokeratin (CK) and Epithelial membrane antigen (EMA) positive, mesenchymal components are vimentin positive, and depending on the cell type may be positive for myogenic markers or smooth muscle actin (SMA), neuroepithelial components are positive for Neuron specific enolase (NSE), CD99. Chromogranin, synaptophysin, Glial fibrillary acidic protein (GFAP), S100<sup>[11,13]</sup>. However, in our case CK vimentin and SMA immunohistochemistry were performed. Here Squamous cells showed positive CK expression, spindle cells showed positive expression for vimentin. Positive SMA expression was seen in the smooth muscle.

SNTCSs are locally aggressive tumors that can invade bone and soft tissue, orbit and cranial cavities. They can also metastasize to the regional lymph nodes and distant sites. Average survival of the patients for 5 years is about 45%. These tumors have high recurrence rate and that usually occurs within 3 years [10,15,16].

SNTCSs require multimodality treatment consisting of surgical resection of the tumor followed by adjuvant radiotherapy and/or chemotherapy depending on the disease extent and performance status of the patient [18]. Our patient underwent surgical excision followed by radiotherapy. Six months have been passed. The patient is doing well and no recurrence till now.

## CONCLUSION

SNTCSs are morphologically and histologically heterogenous malignant tumor with a poor prognosis. These are rare tumors, highly aggressive and often misdiagnosed due to their heterogenous nature. Extensive sampling is necessary for proper histopathological diagnosis. Early diagnosis and surgery combined with radiotherapy and chemotherapy improve the prognosis.

### *Declaration by Authors*

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