Sinonasal Teratocarcinosarcoma: Is Minimally Invasive Resection followed by Adjuvant Histology-directed Chemoradiation a Better Alternative to Radical Excision?

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ABSTRACT

Sinonasal teratocarcinosarcoma (SNTCS) is a rare, highly malignant tumor arising from the primitive embryonic sinonasal tissue or immature pluripotent cells occurring almost exclusively in the sinonasal tract. It is an aggressive tumor with high propensity for locoregional recurrence and mortality. Local recurrence of SNTCS after excision has been reported as high as 45% with a mean recurrence time of 21.3 months. Even though distant metastasis is rare, local recurrence frequently leads to treatment failure and subsequent death. In view of its aggressive behavior, radical excision with or without chemoradiation is advocated as the optimum treatment. Here, we share our experience of SNTCS in a 23-year-old male managed with endoscope-assisted craniofacial resection followed by histocytology-directed chemotherapy with external beam radiation. He remains disease-free in last 3 years of follow-up.

Keywords: Nasal mass, Olfactory neuroblastoma, Sinonasal teratocarcinosarcoma, Teratoid carcinosarcoma.


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INTRODUCTION

Sinonasal teratocarcinosarcoma is a rare highly malignant tumor arising from the primitive embryonic sinonasal tissue or immature pluripotent cells occurring almost exclusively in the sinonasal tract. It is an aggressive tumor with high propensity for locoregional recurrence and mortality. Local recurrence of SNTCS after excision has been reported as high as 45% with a mean recurrence time of 21.3 months. Even though distant metastasis is rare, local recurrence frequently leads to treatment failure and subsequent death. In view of its aggressive behavior, radical excision with or without chemoradiation is advocated as the optimum treatment.

Here, we share our experience of SNTCS in a 23-year-old male managed with endoscope-assisted craniofacial resection followed by histocytology-directed chemotherapy with external beam radiation. He remains disease-free in last 3 years of follow-up.

CASE REPORT

A 23-year-old male presented with a 3-month history of gradually progressing nasal obstruction, anosmia, intermittent epistaxis, and fullness of the right cheek. Vision was normal and there was no cervical lymphadenopathy. Diagnostic nasal endoscopy revealed a smooth lobulated pale to pinkish mass completely filling up the right nostril. Contrast-enhanced computerized tomography showed a heterogeneously enhancing mass filling up the right nasal cavity, nasopharynx extending into maxillary, ethmoid, and sphenoid sinuses, with partial erosion of the lamina papyracea and the cribriform plate. On magnetic resonance imaging, the tumor was found closely abutting to dura at the cribriform area; however, no dural breach was noted (Fig. 1). Moderate vascularity of the mass, deriving its blood supply from both internal maxillary and anterior ethmoidal arteries, was confirmed in angiography. Histopathological examination of punch-biopsy specimen, submitted as multiple punched fragments, revealed heterogeneous admixture of epithelial, mesenchymal, and neuroepithelial elements, rendering a diagnosis of “sinonasal teratosarcarcinoma.”

With an informed consent from the patient, “endoscope-assisted craniofacial resection” was performed under general anesthesia. The tumor had variable consistency, generally firm and fibrous with few friable patches mostly near the cribriform area. It was found adherent to nasal septum and the ethmoid sinus area, confusing the actual site of its origin. The tumor was removed in toto; care was taken near the cribriform area to ensure complete removal of visible tumor avoiding dural injury. Estimated total intraoperative blood loss was around 300 mL.
Histopathological evaluation of the resected specimen showed a heterogeneous malignant tumor composed of three different elements in a necrotic background. An admixture predominantly of primitive neuroectodermal cells immunopositive for mic-2 and neuron-specific enolase, along with epithelial elements including ductal and glandular structures, glandular and sarcomatous stroma with osteoid differentiation was found. Numerous invasive epithelial islands composed of malignant squamous elements and characteristic hybrid squamoglandular units were noted. There was no evidence of a germinoma, embryonal carcinoma, yolk-sac tumor, or chorionicarcinoma in any of the sections (Fig. 2). Surgical margins were positive for tumor. According to the major components in the histocytological examination, he received six cycles of cisplatin (20 mg/m² day) and etoposide (100 mg/m² day). This was followed by 70 Gy of external beam radiation. An excellent clinical response was noted. The patient remains under our regular follow-up for last 3 years, with no apparent recurrence or distant metastasis.

**DISCUSSION**

Sinonasal teratocarcinosarcoma, a very rare tumor arising from the pluripotent cells of olfactory epithelium, occurs almost exclusively in the sinonasal cavity. However, rare occurrence in the nasopharynx and oral cavity is reported in literature.1,3 Sinonasal teratocarcinosarcoma generally presents with relatively benign complaints of nasal obstruction (62%) and recurrent epistaxis (53.52%) in its early stage. Other symptoms raising suspicions of malignancy, such as dysphagia, odynophagia, epiphora, vision loss, exophthalmos, anosmia, headache, and altered sensorium, appear when the tumor spreads into the orbit and intracranially.3 Our patient presented with complaints of progressive nasal obstruction, anosmia, intermittent epistaxis, and fullness of right cheek. The progressive nature of symptoms in our patient may be attributed to the aggressive rapidly growing behavior of SNTCS. Similar to earlier studies,4-6 histopathological evaluation of specimen in our patient showed variegated
components, i.e., epithelial elements including ductal and glandular structures, neuroectodermal elements, and mesenchymal components consisting of fibrous and myxomatous stroma with definite osteoid differentiation. Immunohistochemical studies clearly demonstrated characteristic cellular differentiation of each component. No evidence of a germinoma, embryonal carcinoma, yolk-sac tumor, or choriocarcinoma was seen in any of the sections. Surgical margin was found positive for tumor.

Sinonasal teratocarcinosarcoma usually occurs in elderly males with an average age at presentation of 51.75 years. 
It rarely occurs at a younger age. Only four cases below the age of 25 years have been reported till date, all of them coming from the Indian subcontinent. This is a fact possibly pointing toward a common environmental or genetic link predisposing to early onset of this tumor.

Of the available literature, most were focused on the complex histopathological aspect of the tumor with only few having a mention regarding the treatment protocol and its outcome in follow-up. Management and follow-up details were available in less than half of the reported cases, analysis of which revealed that nearly 85% of cases underwent aggressive radical excision. Sixty percent (60%) of the patients received adjuvant radiotherapy and 12% underwent adjuvant chemoradiation.

In view of its aggressive nature, an extensive radical excision of the tumor with or without chemoradiation is advocated as the optimum treatment. However, local recurrence of SNTCS after excision has been reported as high as 45% with a mean recurrence time of 21.3 months. Even though distant metastasis is rare, local recurrence frequently leads to treatment failure and subsequent death.

Contrary to this general consensus, our patient received a more conservative “endoscope-assisted craniofacial resection” ensuring no residual visible tumor. It was followed with six cycles of cisplatin (20 mg/m² day) and etoposide (100 mg/m² day) directed by the prominence of premature neuroectodermal and epithelial elements embedded in sarcomatous stroma, followed by 70 Gy of external beam radiation. Publications advocating neoadjuvant chemotherapy to downstage the tumor and promote maturation of the neuroectodermal component in SNTCS, and use of histology-specific chemotherapy for a better outcome are indicative of attempts to progress in similar direction.

Disease-free status of indicated case for the last 3 years in follow-up prepared us to propose a minimally invasive, less-deforming endoscope-assisted resection followed by customized “histology-directed chemotherpay” along with radiation as a better alternative to aggressive radical excision for managing SNTCS.

REFERENCES