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Case Report

Sinonasal teratocarcinosarcoma: is minimally invasive resection followed by adjuvant histology directed chemo-radiation a better alternative to radical excision? Case report

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ABSTRACT

Sinonasal teratocarcinosarcoma (SNTCS) is a rare higly malignant tumour occuring almost exclusively in the sinonasal tract. This aggressive tumour arising from the primitive embryonic or immature pluripotential cells of olfactory epithelium has high propencity for loco-regional recurrence and mortality. SNTCS generally presents with relatively benign symptoms like nasal obstruction and recurrent epistaxis in its early stage. In advance stage with orbital and intracranial spread other symptoms raising suspicion of malignancy, such as dysphagia, odynophagia, epiphora, vision loss, exophthalmos, anosmia, headache and altered sensorium appears. In view of its aggressive behaviour, radical excision with or without chemoradiation is advocated as the optimum treatment. 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. Here we share our experience of SNTCS in a 23 year old male managed with endoscopic craniofacial resection followed by histocytology directed chemotherapy with external beam radiation. He remains disease free in last 3 years of follow up.

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

INTRODUCTION

Sinonasal teratocarcinosarcoma (SNTCS) is a rare higly malignant tumour arising from primitive embryonic sinonasal tissue or immature pluripotential cells occuring almost exclusively in the sinonasal tract. It is an aggressive tumour with high propencity for loco-regional recurrence and mortality.[1] 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.[2] In view of its aggressive behaviour, radical excision with or without chemoradiation is advocated as the optimum treatment.

CASE REPORT

A 23 years old male presented with 3 months history of gradually progressing nasal obstruction, anosmia, intermittent epistaxis and fullness of right cheek. Vision was normal and there was no cervical lymphadenopathy.

Diagnostic nasal endoscopy (DNE) revealed a smooth lobulated pale to pinkish mass completely filling up the right nostril. Contrast enhanced computerized tomography (CECT) showed a heterogenously enhancing mass filling up the right nasal cavity, nasopharynx extending into maxillary, ethmoid and sphenoid sinuses, with partial erosion of the lamina paparacea and the cibriform plate. On magnetic resonance imaging (MRI)

the tumour was found closely abutting to dura at the cribriform area; however no dural breach was noted (Figure 1). Moderate vascularity of the mass, deriving its blood supply from both internal maxillary and anterior ethmoidal arteries was confirmed in angiography. Histo-pathological examination of punch-biopsy specimen, submitted as multiple punched-fragments, revealed heterogeneous admixture of epithelial, mesenchymal and neuroepithelial elements, renderring a diagnosis of 'sinonasal teratosarcocarcinoma'.

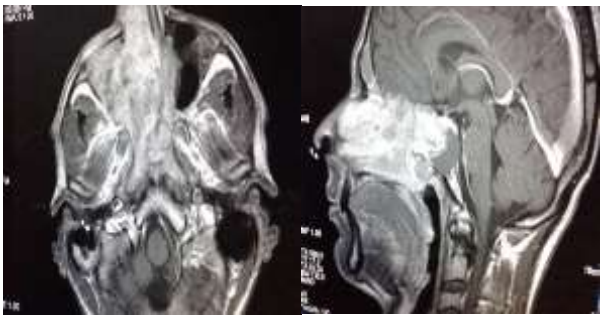

Figure 1: MRI axial and saggital sections showing the mass involving right nasal cavity, maxillary and ethmoid air cells and closely abutting to dura at the cribriform area.

'Endoscopic craniofacial resection' was performed under general anaesthesia. The tumour 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 tumour was removed in toto, care was taken near the cribriform area to ensure complete removal of visible tumour avoiding dural injury. Estimated total intra-operative blood loss was around 300 ml.

Histopathological evaluation of the resected specimen showed a heterogeneous malignant tumour composed of three different elements in a necrotic background. An admixture predominently of primitive neuroectodermal cells immunopositive for mic-2 and NSE (neuron-specific enolase), along with epithelial elements including ductal and glandular structures, glandular and sarcomatous stroma with ostoid differentiation was found. Numerous invasive epithelial islands composed of malignant squamous elements and characteristic hybrid squamo-glandular units were noted. There was no evidence of a germinoma, embryonal carcinoma, yolk-sac tumour or choriocarcinoma in any of the sections (Figure 2). Surgical margins were positive for tumour. According to the major components in histocytological examination, he received 6 cycles of Cisplatin (20 mg/m^2 day) and Etoposide (100 mg/m^2 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.

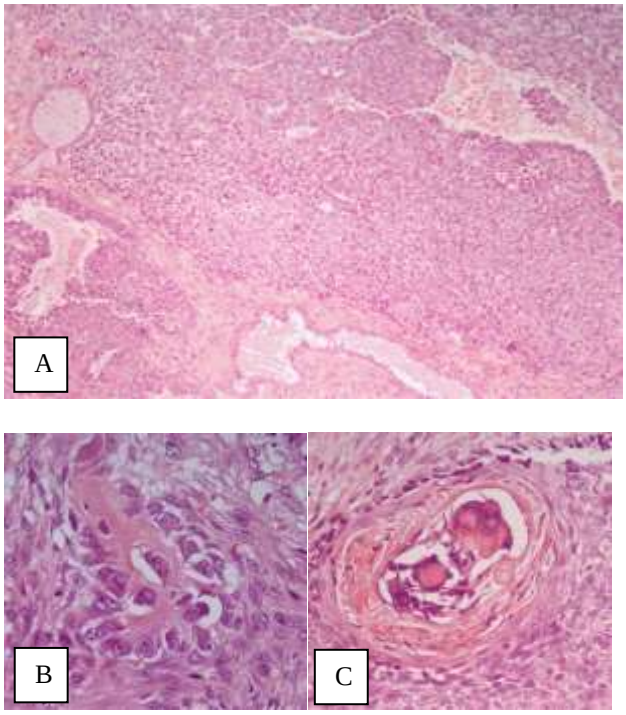

Figure 2: Photomicrographs of resected tumour specimen. (A) showing variegated components of the tumour in the form of heterogeneous admixture of primitive neuroectodermal cells, epithelial elements including ductal and glandular structures, glandular and sarcomatous stroma [H&E 10X] with (B) Ostoid [H&E 40X] and (C) Squamous epithelial differentiation [H&E 20X].

DISCUSSION

SNTCS, a very rare tumour arising from the pluripotent cells of olfactory epithelium, occurs almost exclusively in the sinonasal cavity. Although rare occurrence in the nasopharynx and oral cavity are reported in literature.[1,3]

SNTCS 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 appears when the tumour spreads into the 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 behaviour of SNTCS.

Similar to earlier studies histopathological evaluation of specimen in our patient showed variegated components i.e epithelial elements including ductal and glandular structures, neuro-ectodermal elements and mesenchymal components consisting of fibrous and myxomatous stroma with definite osteoid differentiation.[4-6] Immunohistochemical studies clearly demonstrated

characteristic cellular differentiation of each component. No evidence of a germinoma, embryonal carcinoma, yolk-sac tumour or choriocarcinoma was seen in any of the sections. Surgical margin was found positive for tumour.

SNTCS usually occur in elderly males with an average age at presentation of 51.75 years.[4,7] It rarely occurs at a younger age. Only 4 cases below the age of 25 years reported till date, all of them coming from Indian subcontinent; a fact possibly pointing towards a common environmental or genetic link predisposing to early onset of this tumour.[6]

Of the available literature most were focussed on the complex histopathological aspect of the tumour 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 chemo-radiation.[2,5,7]

In view of its aggressive nature, an extensive radical excision of the tumour 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.[6,8]

In contrary to this general consensus, our patient received a more conservative 'Endoscope assisted Craniofacial Resection ensurring no residual visible tumour. R esection was followed with 6 cycles of Cisplatin (20 mg/m^2 day) and Etoposide (100 mg/m^2 day) directed by the prominence of premature neuroectodermal and epithelial elements embeded in sarcomatous stroma, followed by 70 Gy of external beam radiation. Publications advocating neoadjuvant chemotherapy to downstage the tumour 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.[4,9]

Disease free status of index case for last 3 years in follow-up prepared us to propose a minimally invasive less deforming endoscope-assisted resection followed by customised 'Histology-directed Chemotherapy' along with radiation as a better alternative to aggressive radical excision for managing SNTCS.

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Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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