Journal of Radiotherapy in Practice

*Journal of Radiotherapy in Practice* (2015)
**14**, 438–441 © Cambridge University Press 2015
doi:10.1017/S1460396915000412

## Case Study

# Sinonasal teratocarcinosarcoma (TCS): tackle aggressively

Siddanna Palled, Shekar Patil, Rekha Kumar, Shashidara Poppa Reddy

*Department of Radiation Oncology, Kidwai Memorial Institute of Oncology, Bengaluru, Karnataka, India*

(Received 21 June 2015; revised 7 August 2015; accepted 8 August 2015; first published online 14 September 2015)

## Abstract

*Background:* Sinonasal teratocarcinosarcoma (TCS) are highly aggressive and rare malignant tumours arising from the heterogeneous admixture of components of all the three germ cell layers. There are <60 cases that have been reported in the literature. In spite of aggressive therapy, the average survival is <3 years with multimodality therapy. In total, 70% of the patients who survived >1 year received regimen of combined surgery and adjuvant therapies and this suggests that aggressive therapeutic approaches may improve the treatment outcome.

*Materials and methods:* We are reporting a case study of sinonasal TCS treated with initial surgery followed with concurrent chemoradiotherapy using intensity-modulated radiotherapy (IMRT) technique. The concurrent chemotherapy and adjuvant chemotherapy consisted of carboplatin and etoposide.

*Results:* This aggressive treatment protocol of concurrent chemoradiation along with adjuvant chemotherapy is well tolerated and produced 5-year locoregional control and survival without any long-term morbidities.

*Conclusion:* Our treatment protocol was well tolerated and the outcome in this individual patient has been encouraging. This could justify a combined modality approach with post-operative simultaneous-integrated boost-IMRT and chemotherapy (concurrent and adjuvant) for future patient with sinonasal TCS.

*Keywords*: immunohistochemistry; radiation therapy chemotherapy; sinonasal teratocarcinosarcoma

## INTRODUCTION

Sinonasal teratocarcinosarcoma (TCS) are rare malignant tumours arising from heterogeneous admixture of components of all the three germ cell layers. The incidence is common in adults, with a varied age incidence with marked male predominance. Histologically, tumour exhibiting various degrees of maturation of three germ cell layers and immunohistochemistry supports the diagnosis. There are <60 cases reported in the literature with varied aggressiveness. Despite aggressive therapy, the average survival is <3 years. We are reporting a case study of sinonasal TCS wherein we tried post-surgery concurrent chemotherapy with simultaneous-integrated boost (SIB)-intensity-modulated radiotherapy (IMRT) and succeeded with adjuvant chemotherapy.

Correspondence to: Dr Siddanna Palled, Department of Radiation Oncology, Kidwai Memorial Institute of Oncology, Dr M H Marigowda Road, Bengaluru 560029, Karnataka, India. Tel: +91 99 80 666727. E-mail: siddannap@gmail.com

## MATERIALS AND METHODS

A 37-year-old patient presented with a history of nasal blockage for a period of 3 weeks, which was resultant of a mass in the right nostril. On computed tomography (CT), a diffuse soft tissue mass involving the right frontal, ethmoid, maxillary and sphenoid sinuses, with no evidence of bony erosion was observed. Patient underwent functional endoscopic sinus surgery polypectomy and endoscopic fronto-maxillo-ethmoidectomy to debulk the tumour.

Histopathological examination revealed a high-grade malignant tumour formed by variety of elements resembling immature neuroepithelial tissue, well-formed glands lined by atypical epithelium and sarcomatous areas with abnormal mitosis. Overall features were suggestive of TCS. Immunohistochemistry supported the diagnosis, with the glandular epithelial component expressing cytokeratin (CK) (Figure 1), the small round cell component expressing neuron-specific enolase (NSE) and the spindle cell component expressing vimentin (focal), desmin (focal) and smooth muscle actin (SMA). The tumour cells were negative for S100, Myf-4 and CD99, which effectively ruled out diagnosis such as melanoma and Ewing sarcoma.

An 18 Fr fluorodeoxyglucose positron emission tomography (PET)-CT, 1 month later, revealed (2·2 × 1·9 cm) metabolically active soft tissue in the right ethmoid air cells (which was suggestive of residual metabolically active tumour) (Figure 2). Chemoradiation was planned at this juncture. Patient received radiation therapy with a dose of 60 Gy/28 fractions (#) using IMRT technique. The gross tumour volume (GTV) received 60 Gy and the subclinical disease received 54 Gy in 28# with the use of SIB-IMRT technique along with concurrent three cycles of carboplatin 600 mg (AUC = 5), etoposide 500 mg/m$^2$ in 3 days(Day 1 to Day 3). Post-chemoradiation patient also received three cycles of adjuvant chemotherapy with same regimen of carboplatin and etoposide. At 3 months of post-treatment, PET-CT showed no active mass lesion. She remains disease free on CT imaging for the past 5 years with no long-term treatment-related sequelae.

## RESULTS

Sinonasal TCS are highly aggressive and rare malignant tumours taking origin from epithelial, mesenchymal and neuroepithelial elements. There are <60 cases that have been reported in

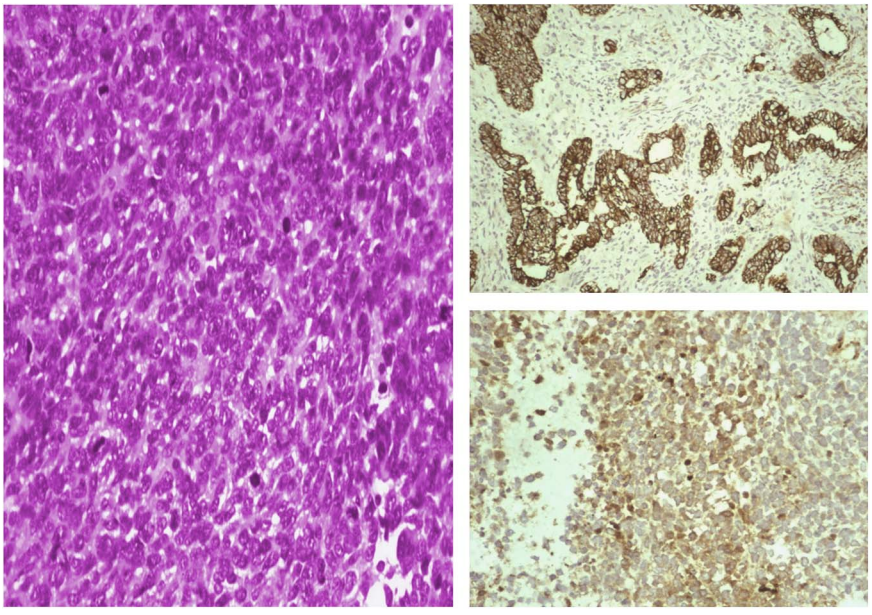

*Figure 1. Histopathological slide of teratocarcinosarcoma with immunohistochemistry study.*



https://doi.org/10.1017/S1460396915000412 Published online by Cambridge University Press

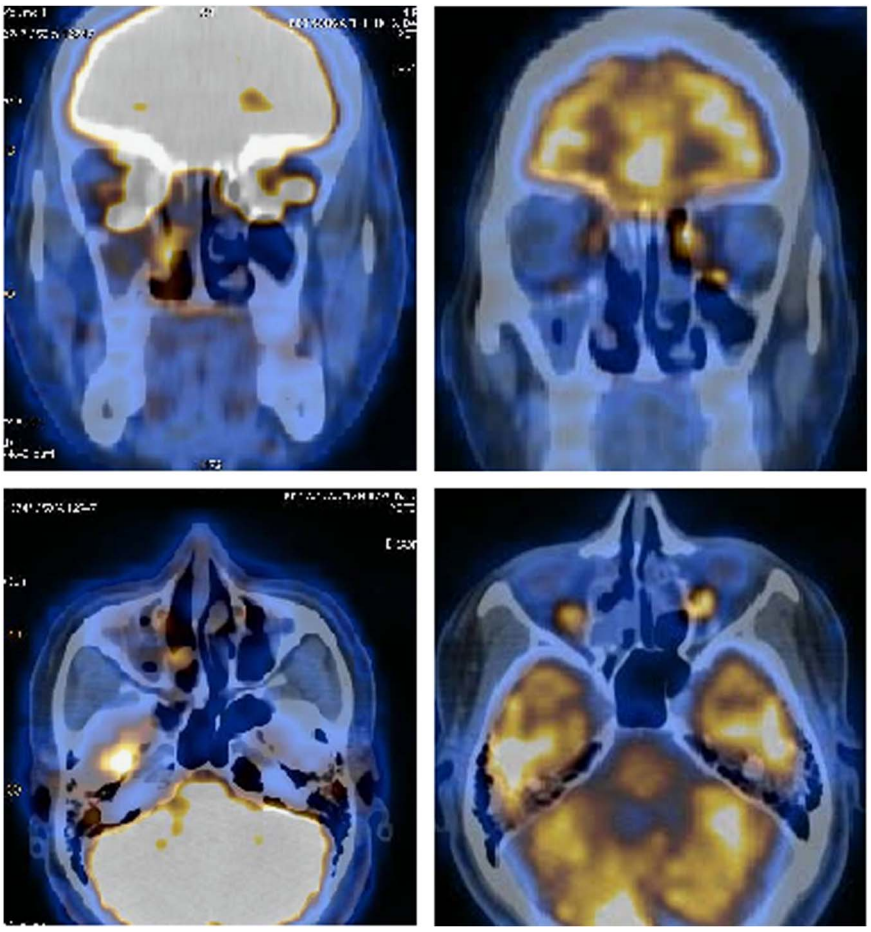

*Figure 2. Positron emission tomography scan pre-treatment and post-treatment.*

the indexed literature (search through PubMed). The average survival is <3 years in spite of aggressive therapy.[3] The patients are exclusively adults, with a varied age incidence in different case series, a mean age of 60 years and marked male predominance.[1] In one of the Chinese case series, the mean age incidence was 39 years.[2]

Histologically, the tumour shows a heterogeneous admixture of components from the three germ cell layers, exhibiting various degrees of maturation. Squamous epithelium, smooth muscle cells, chondro-osseous tissue, intestinal or respiratory type epithelium, 'foetal-type' clear cells and immature neuroepithelium are commonly seen. Immunohistochemical study demonstrates that the epithelial component expresses CK and epithelial membrane antigen, the mesenchymal component variably expresses vimentin, SMA and S-100 protein, the neuroepithelial component expresses NSE, synaptophysin and chromogranin, and the primitive component expresses CD99. In our case, the immunohistochemistry supported the diagnosis.

Among the 54 reported cases of sinonasal TCS, 67% (36 cases collectively from literature) of patients with initial single surgical resection and 80% of patients primarily treated with radiotherapy (RT) had recurrence, or metastasis, or unresponsiveness to treatment. The high rate of local recurrence and metastasis is indicative of the highly aggressive biologic behaviour of the tumour.[3] Almost half of the patients died of tumour within 3 years of diagnosis, inspite of aggressive therapy. In total, 70% (38 cases collectively from literature) of



https://doi.org/10.1017/S1460396915000412 Published online by Cambridge University Press

the patients who survived >1 year had the initial therapeutic regimens of combined surgery and adjuvant therapies, suggesting that aggressive therapeutic approaches may improve the treatment outcome.[3]

In the reported cases where combined modality therapy was used, local adjuvant RT to a dose of 60 Gy[4] was followed with adjuvant cisplatin, etoposide and ifosfamide therapy.[5] In the present case, we treated aggressively combining chemotherapy as well as RT, we delivered 60 Gy/28# to PET-defined GTV and the subclinical disease received 54 Gy/28# using SIB-IMRT along with three cycles of concurrent chemotherapy using carboplatin (AUC = 5) and etoposide succeeded with three cycles of adjuvant chemotherapy using the same regimen. The patient tolerated the treatment well, without any treatment interruption. During treatment, patient developed grade II acute mucositis. At follow-up, PET-CT scans obtained at 3 months and 1 year of treatment showed no evidence of active disease. Currently, at 5 years post-treatment, the patient remains free of disease without any long-term comorbidities of adjacent organs.

## CONCLUSION

Given the high incidence of recurrence in previously published cases, wherein either surgery alone, or RT alone was used, we utilised a rather aggressive approach using SIB-IMRT along with concurrent and adjuvant chemotherapy. Our treatment protocol was well tolerated and the outcome in this individual patient has been encouraging. This could justify a combined modality approach with post-operative SIB-IMRT and chemotherapy (concurrent and adjuvant) for future patient with sinonasal TCS.

## Acknowledgement

The authors acknowledge Dr Kallure K. G. for his contribution in imaging.

## References

1. Heffner D K, Hyams V J. Teratocarcinosarcoma (malignant teratoma?) of the nasal cavity and paranasal sinuses a clinicopathologic study of 20 cases. Cancer 1984; 53 (10): 2140–2154.
2. Wang S Y, Zhu L, Li S M, et al. Sinonasal teratocarcinosarcoma: a clinical, radiologic and pathologic study of 5 cases. Zhonghua Bing Li Xue Za Zhi 2007; 36 (8): 534–538 (in Chinese).
3. Wei S et al. Sinonasal teratocarcinosarcoma: report of a case with review of literature and treatment outcome. Ann Diagn Pathol 2008; 12 (6): 415–425.
4. Takasaki K, Sakihama N, Takasaki H. A case with sinonasal teratocarcinosarcoma in the nasal cavity and ethmoid sinus. Eur Arch Otorhinolaryngol 2006; 263 (6): 586–591.
5. Nitsche M, Hermann R M, Christiansen H, Berger J, Pradier O. Rationale for individualized therapy in Sinonasal Teratocarcinosarcoma (SNTC): case report. Onkologie 2005; 28 (12): 653–656.