Sinonasal Teratocarcinosarcoma – Case Report of an Unusual Neoplasm

Tengchin Wang MD
Department of otolaryngology, Tainan municipal hospital (Managed by Show Chwan Medical Care Corporation) , Tainan City, Taiwan

Abstract

Background : Sinonasal teratocarcinosarcoma (SNTCS) is an extremely rare malignant tumor arising in the sinonasal tract. Diagnosis can be challenging because of its infrequency and phenotypic diversity.
Methods: We experience on a 72-year-old female having nasal obstruction and epistaxis. A friable, easy touch bleeding mass locates at left nasal cavity. Computed tomography shows bony destruction. The mass is surgically resected.
Results: Histologically, this tumor comprises of mature and immature components of epithelial, mesenchymal and neuroectodermal origin. Germ cell components are absent. Teratocarcinosarcoma is confirmed.
Conclusions: Because of the rarity of this tumor, the clinical characteristics and optimal treatment strategy are not well characterized.

Key Words: teratocarcinosarcoma; mesenchyma; neuroectoderma; synaptophysin; desmin; myogen

.

INTRODUCTION

Sinonasal teratocarcinosarcoma (SNTCS) is an extremely rare malignant tumor arising in the sinonasal tract. The tumor can also extend intracranially, further complicating the treatment plan for this disease. Diagnosis can be challenging because of its infrequency and phenotypic diversity. Histologically, this tumor comprises of mature and immature components of epithelial, mesenchymal and neuroectodermal origin, which may be present in varying proportions. Germ cell components are absent in teratocarcinosarcoma . Because of the rarity of this tumor, the clinical characteristics and optimal treatment strategy are not well characterized.

CASE REPORT

We present a 72-year-old female patient suffered from a 3-weeks history of worsening, primarily left nasal obstruction and congestion followed by abrupt epistaxis. On examination, a friable, easy touch bleeding mass locates at left middle turbinate and meatus (figure 1A). Computed tomography shows the lesion making opacities of left maxillary sinus with bony destruction of posterior antral wall (figure 1B). The mass was surgically resected via lateral rhinotomy. During operation, two bony defects were found at orbital floor and posterior antrum wall, and frozen section favored squamous cell carcinoma. Histopathological examination reveals a heterogeneous tumor, which comprised of islands and nests of small round blue cells in a myxoid stroma (figure 2A). Under high power field, there are focal angulated blue cells with hyperchromatic nuclei, abundant eosinophilic and refractile cytoplasm(figure 2B). The epithelium component comprises squamous cells showing dyskeratosis, nuclear hyperchromasia and pleomorphism, and carcinoma is confirmed (figure 3A). Rhabdomyoblasts (primitive skeletal muscle cells) are seen within polygonal or spindle-shaped cells in the mesenchymal component (figure 3B)
Immunohistochemical study revealed positive for cytokeratin in epithelial islands (figure 4A); stain positively for neuroendocrine markers, such as synaptophysin is shown in primitive neuroectodermal or blastemal components (figure 4B); the stromal cells were focally immunopositive for desmin and myogen (figure 4C and D). There was no evidence of germ cell tumor in any sections. Based on the histomorphology and immunohistochemistry, a diagnosis of SNTCS was rendered.

DISCUSSION

Sinonasal teratocarcinosarcoma (SNTCS) is an aggressive malignant neoplasm. The middle-aged male are preponderance. The most common presenting symptoms are nasal obstruction and epistaxis. The differential diagnosis of this entity is varied and includes poorly differentiated squamous cell carcinoma, sarcoma, olfactory neuroblastoma, small cell carcinoma, malignant mixed tumor of salivary gland and extragonadal malignant germ cell tumor [1].
Histologically, SNTCS has a variable mixture of immature neural or blastemal-appearing components and various benign, atypical, or overtly malignant epithelial and mesenchymal elements without features of specific germ cell tumors [2].
The associated neuroectodermal and neuroendocrine differentiation can be staining positively for neuroendocrine markers, such as synaptophysin, chromogranin A, and CD99. Staining of the epithelial and mesenchymal elements depends on the

particular components of a given tumor. SNTCS is commonly misdiagnosed, especially in small biopsy, with a misdiagnosing rate of roughly 50% [1].

The most common treatment regimen is complete excision with adjuvant radiation

therapy. But the prognosis is poor, with reported over all survival rates ranging from 40-46% at the end of two years [3]. Recurrence and metastasis rates are high,

occurring in 67% of patients treated with surgery alone and 45.7% of patients treated with surgery and radiation [1].

In conclusion, STNCS is rare and often poses a diagnostic challenge. Even under an aggressive treatment, the prognosis is still poor.

.

REFERENCE

*[1]*Misra P, Husain Q, Svider PF, Sanghvi S, Liu JK, Eloy JA: Management of sinonasal teratocarcinosarcoma: A systematic review. *Am J Otolaryngol 35:5-11, 2014*

*[2]*Endo H, Hirose T, Kuwamura KI, Sano T. Case report: sinonasal teratocarcinosarcoma. *Pathol Int. 2001;51(2):107-112.*

*[3]*Smith SL, Hessel AC, Luna MA, Malpica A, Rosenthal DI, El-Naggar AK: Sinonasal teratocarcinosarcoma of the head and neck: A report of 10 patients treated at a single institution and comparison with reported series. *Arch Otolaryngol Head Neck Surg 134:592-595,2008*

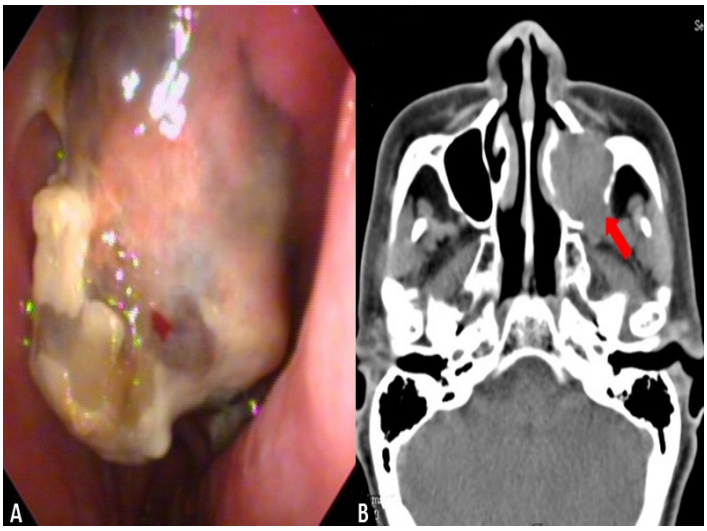

Figure 1. A: Friable, easy touch bleeding mass locates at left middle meatus. B: Computed tomography exhibits the lesion making opacities of left maxillary sinus with bony destruction of posterior antral w all (red arrow)

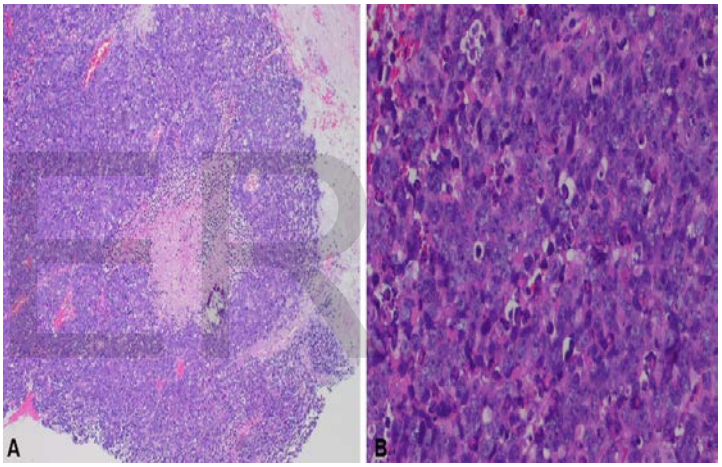

Figure 2. A: Heterogeneous tumor comprises of nests of small round blue cells in a myxoid stroma. B: The high power field exhibits blue cells with hyperchromatic nuclei, abundant eosinophilic and refractile cytoplasm.

- **Corresponding author: Tengchin Wang, Department of otolaryngology, Tainan municipal hospital (Managed by Show Chwan Medical Care Corporation), No.670, Chongde Rd., East Dist., Tainan City 701, Taiwan E-mail:tengchin27@hotmail.com Telephone Number：+886-6-2609926*