Adult Embryonal Rhabdomyosarcoma of the Ethmoid: A Rare Entity

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ABSTRACT
Embryonal rhabdomyosarcoma is an exceedingly rare tumor in adult. We report an embryonal rhabdomyosarcoma of the ethmoid in a 59-year-old Iban lady who presented with proptosis and complete ptosis of her left eye for two months. Imaging investigations showed left ethmoidal and left orbital soft tissue mass with extradura and dura involvement. The patient was planned for chemotherapy. Unfortunately, in such an advanced disease, she succumbed before treatment.

KEY WORDS:
rhabdomyosarcoma, embryonal rhabdomyosarcoma, ethmoidal rhabdomyosarcoma

INTRODUCTION
Embryonal rhabdomyosarcoma is predominantly a childhood malignancy and is extremely rare in adult. It is a highly malignant neoplasm of pleuripotential embryonic mesoderm, which differentiates to form cells similar to rhabdomyoblasts of the foetus. Therefore in adult, this malignancy is uncommon since the tumor arises from fully mature muscle cell which is not prone to undergone malignant change. Although childhood embryonal rhabdomyosarcoma has a very good prognosis with 88% 5 year survival rate, the prognosis of head and neck adult onset embryonal rhabdomyosarcoma has poor outcome. Most of the articles report studies of case series from in a single centre and reports of single cases. This report illustrates an adult patient with embryonal rhabdomyosarcoma of the ethmoid sinus which locally aggressive without distant metastasis.

CASE REPORT
A 59-year-old housewife presented with left eye proptosis and complete ptosis within two months duration. It was associated with epistaxis, nasal blockage, left facial swelling and numbness, diminution of left eye vision and severe throbbing headache. Other significant history was unremarkable.

Clinical examination revealed left complete ptosis with proptosis which was associated with swelling of the left frontal area (Fig. 1). The eyeball was tender and deviated downwards and outwards. Nasoendoscopy revealed a friable mass occupying the entire left nasal cavity and the roof of left nose.

Fig. 1: Photograph of the patient showed proptosis and complete ptosis of the left eye. The underlying skin is inflammed.

Fig. 2: Contrast-enhanced (axial) CT scan showed a homogenous mass that originates from the left ethmoid sinus and extends to nasal cavity and the roof of left nose.

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Fig. 2: Contrast-enhanced (coronal) CT scan showed a homogenous mass in both ethmoid and destruction of surrounding bony structures.
Biopsy was sent for histopathological examination and immunohistochemistry which revealed small round blue cell tumor, which stained for desmin and vimentin, suggestive of moderately-differentiated embryonal rhabdomyosarcoma (Fig.3).

This case was discussed in combined otolaryngology, neurosurgery and oncology meeting and planned for chemotherapy due to her advanced disease and extreme age. She was admitted to Oncology ward, but her general condition deteriorated and was been bed bounded. Unfortunately, chemotherapy was not started because she succumbed to the disease.

REFERENCES