CASE REPORT

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.

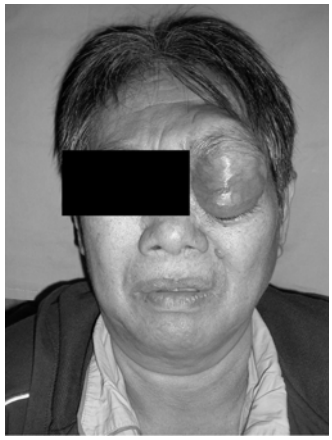

Fig. 1: Photograph of the patient showed proptosis and complete ptosis of the left eye. The underlying skin is inflammed.

Computed tomography showed huge tumor occupying the ethmoid and left orbit with extension to frontal sinus superiorly and erosion of its inner wall. The involvement of the ethmoid includes the cribriform plate, medial orbital wall and crista galli. There was infiltration of the anterior part of left optic nerve and extension to medial extraconal space of right orbit. Maxillary sinus, nasal cavity and erosion at the floor of right orbit were also seen. Magnetic resonance imaging showed tumour infiltrating the dura, without involvement of brain parenchyma (Fig.2).

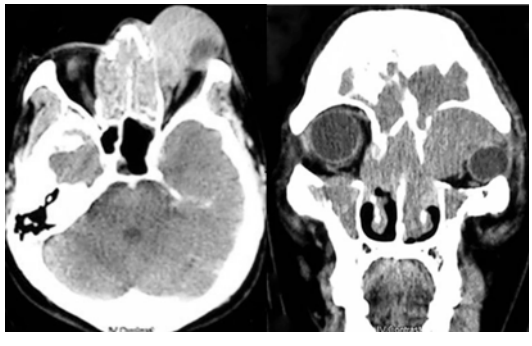

Fig. 2: Contrast-enhanced (axial) CT scan showed a homogenous mass that originates from the left ethmoid sinus and extends to nasal cavity and left orbit. Medial wall of maxillary sinus are eroded. Contrast-enhanced (coronal) CT scan showed a homogenous mass in both ethmoid and destruction of surrounding bony structures.

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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 rhabdomysarcoma (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.

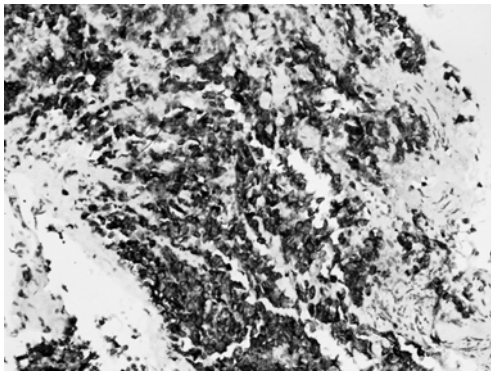

Fig. 3: High-power photomicrograph showed immunostaining with desmin on the cell (desmin, X200).

DISCUSSION

Adult rhabdomyosarcoma is a rare sarcoma occurring mainly in the extremities and it is exceedingly rare in head and neck region. Head and neck sarcoma accounted only about 10% of all sarcoma[1]. In 1996, Lee *et al* reported 22% incidence of head and neck adult rhabdomyosarcoma over five year period, compared to extremities[2]. Therefore, our initial provisional diagnosis was ethmoidal carcinoma based on the presentation and age of the patient.

Generally rhabdomyosarcoma is thought to arise from primitive mesenchymal cells that would develop into striated muscle. It can be found virtually anywhere in the body, including those sites where striated muscle are not normally found. Histologically rhabdomyosarcoma are classified into four type; embryonal, alveolar, pleomorphic and mixed[2]. Embryonal further divide as myxoid, round, botyroid and spindle cell variant[3]. The embryonal rhabdomyosarcoma accounts 60% of childhood rhabdomyosarcoma and occur most frequently in the head and neck region. Although it has been reported in adult, they are exceedingly rare especially after the age of 45 years[3].

The spectrum of presentation of rhabdomyosarcoma varies according to the primary site. Ethmoidal involvement is extremely dangerous since the extension of the tumor comes in contact with the cribriform plate, which subsequently goes intracranial. Orbital involvement clinically presents with ptosis and/or unilateral proptosis, dislocation of the lens or impairment of ocular mobility. It may immitates orbitocellulitis, chalazion, epibulbar papilloma or as nasolacrimal duct obstruction[4]. About 45% of patient eventually develops distant metastasis with lung is the most common site and this risk is largely dependent on the grade of the tumor[1]. However, this is not seen in our patient.

The radiological finding for adult and childhood rhabdomyosarcoma will produce similar imaging findings[2]. Imaging of the primary tumor is necessary to define the extent of tumor. The extent of bone erosion has been suggested to be an important prognostic factor in nonorbital rhabdomyosarcoma of head and neck[2]. Computed tomography studies will show poorly defined, relatively homogenous masses with destruction of adjacent bony structures with contrast enhancement generally same degree as muscle[2]. This findings mirror the description of our patient.

The histologic appearance of embryonal rhabdomyosarcoma is characterized by the presence of undifferentiated patternless spindled cells and small round blue cell[3]. Immunostaining are required for a diagnosis of embryonal rhabdomyosarcoma. The strongest desmin stain will be best seen in well-differentiated tumors while vimentin are more indicative of poorly differentiated cell[3]. In this patient, both desmin and vimentin were positively stained which consistent with moderately-differentiated embryonal rhabdomyosarcoma.

An approach of multimodal therapy has been practiced includes en bloc resection, followed by radiotherapy and chemotherapy[7]. When combined with surgery and radiation therapy, chemotherapy using doxorubicin, ifofosfamide and vincristine yielded 55% overall and 64% disease free survival at 2 years[5]. Adult type soft tissue sarcomas are considered to be relatively radioresistent. Adjuvant external beam RT following surgery is the usual treatment for intermediate and high grade sarcoma[1].

After multidisciplinary discussion, the patient was planned for chemotherapy due to the tumor invasiveness and the age of presentation. En bloc dissection of the tumour in this patient was not possible due to aggressive, extensive nature of the tumour with addition of the patient's poor medical status which renders patient to be unfit for surgery.

CONCLUSION

In conclusion, this case is illustrative of several points. The clinical presentation of our patient was unusual at her age. The location and rapidly progressive course of the tumor were not typical and finally, the imaging investigations and immunostaining pattern are consistent with previous studies.

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