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Orbital Rhabdomyosarcoma: A Case Series

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ABSTRACT

In this paper, we report three cases of orbital rhabdomyosarcoma in different age groups with different histopathologic types. Case 1 is a 10-year old Malay boy who presented with painless proptosis of the left eye. Magnetic resonance imaging (MRI) showed soft tissue mass arising from the lateral rectus muscle. Tissue biopsy revealed rhabdomyosarcoma of embryonal type. Case 2 is a 32-year old Malay man who presented with a progressive left eye proptosis associated with pain and redness. Computed Tomography (CT) scan showed a retrobulbar mass extending medially in the left orbit. Meanwhile, biopsy showed rhabdomyosarcoma of pleomorphic type. Case 3 is a 67-year old Malay woman who presented with proptosis and fungating growth of left orbital region, epistaxis and nasal blockage. CT scan revealed an ill-defined mass filling up the left orbital cavity. Biopsy showed rhabdomyosarcoma of alveolar type. Rhabdomyosarcoma is the most common in childhood but it should be considered as a differential diagnosis of orbital tumours irrespective of age.

Keywords: Orbital rhabdomyosarcoma, proptosis

INTRODUCTION

Rhabdomyosarcoma is the most common orbital malignancy of childhood. The primary sites of rhabdomyosarcoma are head and neck (45%), trunk (40%), and extremities (15%)[1]. Orbital rhabdomyosarcoma accounts for about 25% to 35% of the head and neck rhabdomyosarcoma[1]. The average age at diagnosis is eight years old[2]. There is predilection of males. Main presenting complaint is proptosis or displacement of the eyeball[3]. Meanwhile, the presenting symptoms are generally similar in different age groups.

CASE REPORTS

Case 1:

A 10-year old Malay boy presented with painless left eye proptosis for five days. It was associated with left eye redness, diplopia and blurring of vision. An examination of the left eye revealed a vision of 1/60 with proptosis of eight millimetres. The left fundus was normal. Relative afferent pupillary defect (RAPD) was positive in the left eye. The right eye was unremarkable. Magnetic Resonance Imaging (MRI) scan of the orbit and brain showed a well defined soft tissue mass arising from the left lateral rectus muscle extending into the nasal cavity, adjacent sinuses, left temporal and frontal lobe. Patient underwent craniotomy and debulking of the tumour. Meanwhile, histopathological examination (HPE) revealed rhabdomyosarcoma of embryonal type. His stage of disease was Group III based on the Intergroup Rhabdomyosarcoma Study Group (IRSG) staging classification. The child underwent chemotherapy. Unfortunately, he developed left temporal lobe abscess and succumbed.

Case 2:

A 32-year old Malay man presented with progressive left eye proptosis associated with pain, lid swelling and conjunctival congestion for one month. An examination of the left eye revealed a vision of 6/12 with proptosis of five millimetres and the presence of conjunctival injection with mild periorbital swelling. There was no RAPD. Fundus examination was unremarkable. The right eye was normal. Computed Tomography (CT) scan showed a homogenous mass of soft tissue density in the left retrobulbar region extending medially in the orbit with left proptosis. There was no bony deformity, while the rectus muscles and optic nerve were normal. Biopsy result is consistent with rhabdomyosarcoma of pleomorphic type. He was further managed in another centre.

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Case 3:

A 67-year old Malay lady presented with progressive left eye proptosis for six months. It was associated with eyelid swelling, reduced vision, nose block and epistaxis from the left nostril. An examination revealed a huge fungating growth from the left orbital region extending to the nasal region and upper cheek. The left eye had no perception to light. There was multiple cervical lymphadenopathy. Anterior rhinoscopy showed that the left nostril was filled with mass. A CT scan showed a heterogenous ill-defined mass in the left orbital cavity pushing the left globe outside the orbit. The lesion infiltrated the left maxillary sinus, both nasal cavities, subcutaneous tissue of the nasal bridge and left cheek (Figure 1). The CT scan of the thorax and abdomen showed lesions in the lungs and liver. Biopsy from the left nostril is consistent with rhabdomyosarcoma of alveolar type (Figure 2A-D). She was in Group IV based on the IRSG staging classification. She also underwent chemotherapy but subsequently defaulted follow-up.

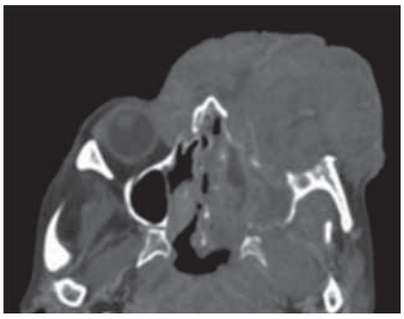

Figure 1. The axial image of CT scan showed a huge, ill-defined mass in the left orbital cavity. Left globe is pushed outside from its cavity and distorted in shape. It infiltrated into the subcutaneous tissue of nasal bridge.

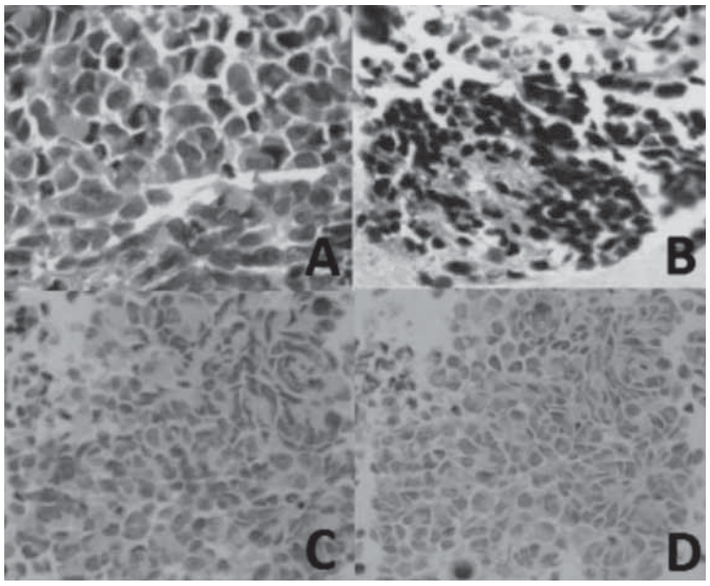

Figure 2.

A: The cells are small to medium in size, with hyperchromatic nuclei and scanty eosinophilic cytoplasm. Some cells show prominent nucleolus (× 400).
B: The tumour cells are positive for anti-vimentin antibody (× 200).
C: The tumour cells are negative for anti-cytokeratin antibody (× 200).
D: The tumour cells show scattered positivity to anti-desmin antibody (× 200).

DISCUSSION

Rhabdomyosarcoma is the most common soft tissue sarcoma in children. Orbital rhabdomyosarcoma is the occurrence of this tumour in the area of the eye[1]. Nonetheless, rhabdomyosarcoma is very uncommon in adults. Shields *et al*., in their study of primary ophthalmic rhabdomyosarcoma, reported that 24% were older than 10 years and 12% were older than 20 years at the age of presentation[3]. In this paper, we presented a case series which covers the typical age of presentation and also in older age group. Though uncommon, rhabdomyosarcoma should not be excluded as a differential diagnosis in the older age group.

The common ocular findings of orbital rhabdomyosarcoma include proptosis (79%), globe displacement (79%), conjunctival congestion (61%), blepharoptosis (55%), dilated episcleral vessels (42%), and ocular motility restriction (42%)[3]. Proptosis was the main complaint present in all the three cases reported. In Case 3, the patient presented late with advanced rhabdomyosarcoma with nasal symptoms. Presenting symptoms of orbital rhabdomyosarcoma depend on the site of lesion. Tumours in the anterior and inferior parts of the orbit tend to cause eyelid swelling and chemosis, whereas posterior tumours often cause optic disc swelling, choroidal folds and ophthalmoplegia[2].

Rhabdomyosarcoma is composed of malignant cells with characteristics of striated muscle in different stages of embryogenesis[1]. It is now known that rhabdomyosarcoma develops from pleuripotential mesenchymal cells that have the ability to turn into striated muscle although it was once believed to arise from extraocular muscle[2]. Horn-Enterline classification of histopathological subtypes of rhabdomyosarcoma includes pleomorphic, embryonal, alveolar and botryoid[4]. Other entities of rhabdomyosarcoma include anaplastic, spindle cell, sclerosing, and lipid-rich rhabdomyosarcoma[4].

Children are likely to have embryonal rhabdomyosarcoma, whereas young adults tend to have alveolar rhabdomyosarcoma[4]. Pleomorphic rhabdomyosarcoma is more common in older adults. Embryonal subtype is the most common in orbital rhabdomyosarcoma[1, 2, 5]. The International Classification of Rhabdomyosarcoma is proposed following modifications to the classic Horn-Enterline classification[4]. It reveals that botryoid and spindle cell rhabdomyosarcoma have superior prognosis, whereas embryonal rhabdomyosarcoma has intermediate prognosis. Meanwhile, alveolar rhabdomyosarcoma and undifferentiated sarcoma have poor prognosis[4].

Rhabdomyosarcoma is staged according to the IRSG staging classification[1, 3]. Group I denotes completely resected localized disease that was confirmed microscopically with the absence of regional lymph node involvement. Group II consists of those with residual disease on microscopic examination and/or regional lymph node involvement. Group III indicates an incomplete resection with biopsy or gross residual disease at the site of origin or in the regional lymph nodes. Group IV indicates distant metastasis present at onset.

IRSG has proposed a treatment for Groups I to III[1, 3]. Group I is advised for chemotherapy without radiotherapy. Groups II and III are recommended for both chemotherapy and radiotherapy. Management of Group IV generally consists of chemotherapy combined with radiotherapy that is delivered to the involved sites of tumour[3]. Recurrent tumours are usually managed with orbital exenteration with subsequent chemotherapy and radiotherapy[3]. Advancement in radiotherapy and chemotherapy has increased the survival rates of patients with orbital rhabdomyosarcoma[2].

CONCLUSION

In conclusion, rhabdomyosarcoma can present in any age group although it is the most common primary orbital malignancy of childhood. It should be considered as differential diagnosis of orbital tumours irrespective of age. Majority of orbital rhabdomyosarcoma patients present with proptosis. Imaging studies and tissue diagnosis are necessary. Treatment plan should be individualized according to the stage of the disease.

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