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ALVEOLAR TYPE OF ADULT ORBITAL RHABDOMYOSARCOMA: A CASE REPORT.

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ABSTRACT

Rhabdomyosarcoma is a soft tissue tumour of undifferentiated mesenchymal cells that commonly occurs in the paediatric age group, particularly the embryonal subtype. Pleomorphic rhabdomyosarcoma usually occurs in an adult. On the other hand, alveolar rhabdomyosarcoma can affect all age groups. The common site of alveolar rhabdomyosarcoma is mainly the extremities. Embryonal rhabdomyosarcoma usually affects the head and neck region as well as the genitourinary tract. Only 10% of overall rhabdomyosarcoma involved the orbital region. Imaging features of rhabdomyosarcoma may not be specific; however, early suspicion based on radiological imaging may help in the patient's management and treatment course. We report a rare case of orbital rhabdomyosarcoma, an alveolar subtype in a 54 year-old-woman. The aim of this case report is to discuss the clinicopathological features of this type of tumour in an adult.

Keywords: Alveolar rhabdomyosarcoma, Adult, Orbital neoplasm, Chemo-radiotherapy, Radiological imaging.

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ABSTRACT

Rhabdomyosarcoma is a soft tissue tumour of undifferentiated mesenchymal cells that commonly occurs in the paediatric age group, particularly the embryonal subtype. Pleomorphic rhabdomyosarcoma usually occurs in an adult. On the other hand, alveolar rhabdomyosarcoma can affect all age groups. The common site of alveolar rhabdomyosarcoma is mainly the extremities. Embryonal rhabdomyosarcoma usually affects the head and neck region as well as the genitourinary tract. Only 10% of overall rhabdomyosarcoma involved the orbital region. Imaging features of rhabdomyosarcoma may not be specific; however, early suspicion based on radiological imaging may help in the patient's management and treatment course. We report a rare case of orbital rhabdomyosarcoma, an alveolar subtype in a 54 year-old-woman. The aim of this case report is to discuss the clinicopathological features of this type of tumour in an adult.

Keywords: Alveolar rhabdomyosarcoma, Adult, Orbital neoplasm, Chemo-radiotherapy, Radiological imaging.

INTRODUCTION

Rhabdomyosarcoma is a neoplasm of striated muscle or its precursor mesenchymal cells. It contributes 50% of all soft tissue sarcoma in paediatric age group, making it the most common soft tissue sarcoma.[1] In contrast, rhabdomyosarcoma contribute about 3% of adult soft tissue sarcoma, whereas soft tissue sarcoma accounts for only 1% of all adult malignancies.[1] The common site of rhabdomyosarcoma is head and neck (40%), genitourinary system (20%), extremities (20%), trunk (10%) and other (10%).[2] To our knowledge, there were less than five cases of adult alveolar orbital rhabdomyosarcoma reported since 2010.[3–5] Most of the reported alveolar rhabdomyosarcoma were from paranasal sinuses with orbital extension. With regards to the rarity of rhabdomyosarcoma in adults, we would like to share a case describing adult orbital rhabdomyosarcoma, which was suspected based on imaging and supported by histopathological confirmation. The patient's condition improved after chemo-radiotherapy.

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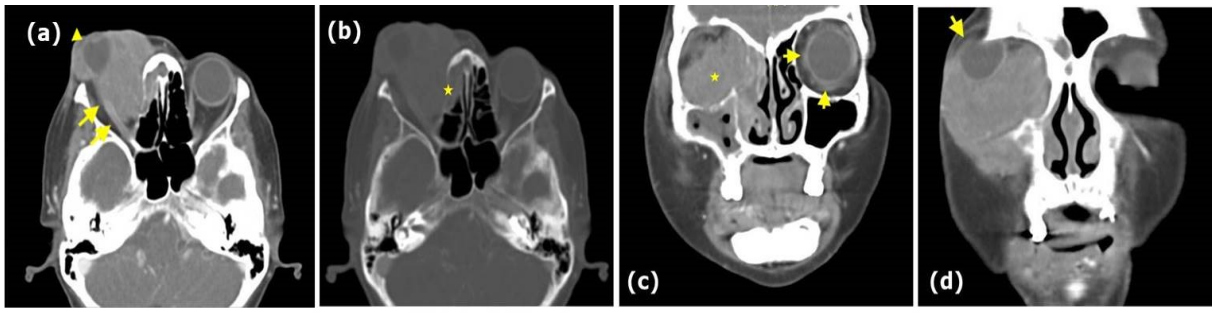

Figure 1: Contrasted CT images pre-treatment. (a) Axial image showed enhancing lesion medial to the right globe (yellow arrowhead) occupying the orbit causing proptosis. Extension seen to the anterior ethmoid air cells. No fat plane between the lesion and optic nerve (yellow double arrows). (b) Bone window in axial cut, no calcification within the lesion and the right lamina papyracea (yellow *) is thinned out. (c) Coronal image

CASE REPORT

A 54-year-old female who previously had no known medical illness presented with right lower eyelid swelling associated with eye discharge for 3 weeks duration. These symptoms worsened and were associated with right eye proptosis which prompted her to seek medical attention. No visual disturbances or infective symptoms were noted. The symptoms worsened during the presentation. Upon examination, no impairment in the visual acuity was seen.

CT scan was performed which showed a soft tissue lesion extending from the orbit and into the anterior ethmoid sinus along the lacrimal duct (Figure 1). Biopsy was taken from the soft tissue mass extension at the anterior ethmoidal sinus, sphenoid sinus, right extraconal tissue, lacrimal duct tissue and medial wall of the maxillary sinus.

Histopathological examination showed small round to oval cells arranged mainly in alveolar patterns. The tumour cells clinging to the fibrovascular core with central dyshesion (Figure 2a). The cells were monomorphic with round hyperchromatic nuclei and minimal cytoplasm. Eosinophilic rhabdomyoblast was also noted. The tumour cells were positive for desmin, myo-D (Figure 2b) immunostains, and negative for LCA, CKAE1&A3, synaptophysin and chromogranin. The final diagnosis of alveolar rhabdomyosarcoma was established. A CT for staging was done; which showed no evidence of distant metastases. She was offered right orbital exenteration and maxillectomy, however she refused. Chemotherapy was started two weeks after the histopathology result. Currently patient is on ongoing chemotherapy Ifosfamide, Vincristine, Actinomycin (IVA) regime. She had completed radiotherapy 30 cycles.

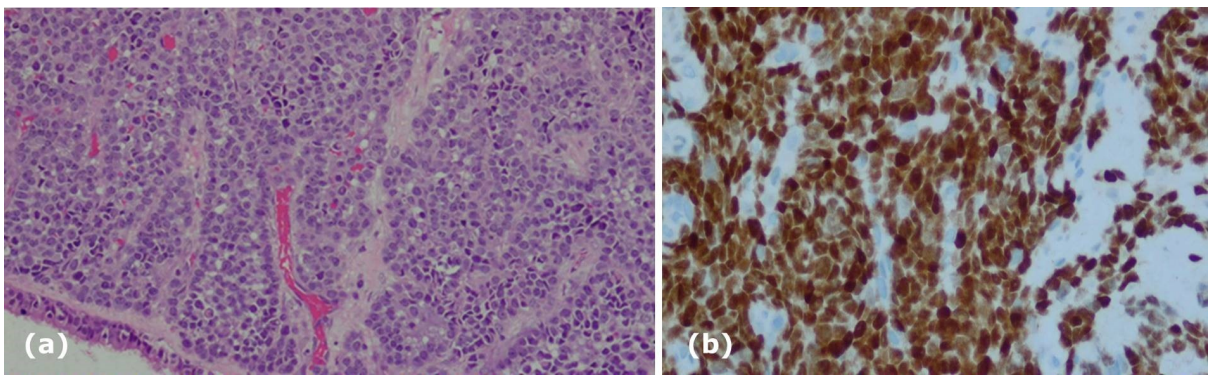

Figure 2: (a) Hematoxylin & Eosin stain exhibits small round blue cells tumour arrange in alveolar pattern with discohesive cells at the center (200x), (b) Exhibits MyoD nuclear positivity by immunohistochemistry stain (400x). (Click on image to enlarge).

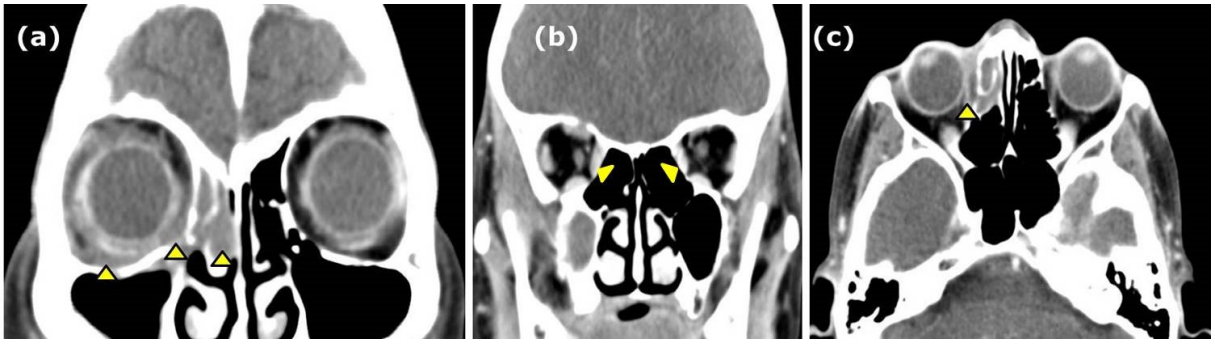

Figure 3: Contrast enhanced CT images during mid cycle chemoradiotherapy. (a) Coronal image at the level of mid globe showed reduced tumour size with residual tumour (yellow arrowheads) at the inferomedial of right orbit and extended to right ethmoid sinus. (b) Coronal image at the posterior part of the orbit showed the rectus muscles (arrowhead) are symmetrical bilaterally. (c) Axial images showed the lesion (yellow arrowhead) is smaller at the medial aspect of the right globe and extension to the ethmoid sinus. Her previous right proptosis had resolved in this study. (Click on image to enlarge).

A repeat CT mid-cycle of chemotherapy (Figure 3) shows reduced tumour size, which overall concluded as a partial response based on RECIST criteria. Currently, she is still on chemotherapy and has refused surgical intervention.

DISCUSSION

Rhabdomyosarcoma is the second common soft tissue sarcoma in childhood after osteosarcoma and is highly malignant with extensive local invasion.[4] Soft tissue sarcoma accounts for 1% of adult tumor and rhabdomyosarcoma accounts for 3% of the soft tissue sarcoma.[6] There are four subtypes of rhabdomyosarcoma: embryonal (49%), [6] alveolar (30%), [6] pleomorphic and botryoid. Although rare in adults, the most common primary site for an adult is extremities, head and neck, and abdominal wall.[7]

The embryonal subtype mostly arises in the head and neck. In contrast, the alveolar subtype is commonly an intramuscular tumour of the extremities, and both of these are common in paediatric age group. [6] It is the pleomorphic subtype that almost exclusively affects adults and is common in the skeletal muscle of thigh.[6] In comparison to our case, the histopathology revealed an alveolar subtype that is rare for head and neck and even rarer for adult-onset. Most alveolar rhabdomyosarcoma reported in literatures are from paranasal sinuses with extension to the orbit. Since 2010, we have only been able to find 3 cases of primary orbital alveolar rhabdomyosarcoma reported in literature, the summary of which is presented in table I.[3–5]

Orbital rhabdomyosarcoma may present insidiously, and the presentation is similar as other space-occupying lesion with characteristics of rapid onset of proptosis and globe displacement, however, it is uncommon to have invasion to the paranasal sinuses.[2] This case presentation is typical for orbital space-occupying lesions.

Imaging is useful in determining the extent of the tumour and for characterization of the tumour to reach a diagnosis. Cross-sectional imaging appearance for adult rhabdomyosarcoma is moderately well-circumscribed, homogenous, round to ovoid mass, isodense to muscle, and shows moderate to marked enhancement post-contrast.[2] It is common to have lymphadenopathy in adults (more marked with alveolar subtype) and uncommon to have calcification within.[6] These features were seen in our case, however there was no lymphadenopathy. If there is associated lymphadenopathy, differential diagnosis of lymphoma needs to be considered. Although there is no lymphadenopathy, the

Table I: Summary of cases of orbital alveolar rhabdomyosarcoma in adults previously reported in current literature.

No	Authors	Age (years)	Presenting symptoms	Radiological findings	Diagnosis
1	Lavaju P et al.[3]	45	Gradual protrusion of right eye for 3 years and worsened for 15 days.	homogenous orbital mass with erosion of the superior and lateral orbital walls and a deformed right globe with retinal detachment. The optic nerve sheath complex was normal.	Alveolar orbital rhabdomyosarcoma
2	Bagdonaite et al. [5]	52	Right eye and eyelid swelling and proptosis.	Extraconal soft tissue mass in orbital floor with cortical destruction of roof and lateral wall of the maxillary sinus.	Alveolar orbital rhabdomyosarcoma
3	Kimario et al.[4]	25	Right facial swelling, nasal blockage and right eye protrusion for 3 months	Not stated	Orbital alveolar rhabdomyosarcoma

overall feature of the lesion is aggressive locally by causing displacement of the orbit anteriorly and causing proptosis. The other uncommon imaging feature of our case of extension to the ethmoid sinus raised a possibility of mucocele of ethmoid sinus. For sinonasal mucocele, the CT appearance is well-defined, homogenous and sometimes hyperdense but no post-contrast enhancement can be appreciated.[8] Mucocele is often associated with bony expansion. For this case, the first differential diagnosis from imaging was orbital rhabdomyosarcoma because of the homogenous lesion epicenter is within the orbit, post-contrast enhancement, absence of sinus expansion and no evidence of lymphadenopathy.

The principal treatment of rhabdomyosarcoma is surgery followed by radiation therapy and chemotherapy. In comparison with pediatric rhabdomyosarcoma, prognosis of adult rhabdomyosarcoma is very poor with less than 50% of 5 years survival rates.[6] The patient in our case refused the surgical option. She completed radiotherapy with reduction in size of the tumour and was undergoing chemotherapy, 1 year after diagnosis. It has to be kept in mind that although with surgery, radiotherapy and chemotherapy, the adult rhabdomyosarcoma is less radiosensitive and chemosensitive than paediatrics rhabdomyosarcoma.[7] The tumour recurrence rate is high even after completion of treatment, hence it is important to follow up the patient and request prompt imaging if new symptoms arise during follow up.

CONCLUSION

Although rhabdomyosarcoma is rare in adults and shares common features with other soft tissue sarcomas, a high index of suspicion is essential when there is soft tissue lesion seen at the orbit, head and neck on imaging assessment. This may improve the lesion detection and tissue diagnosis is essential to differentiate between other lesions occurring in this region. Once confirmatory diagnosis is made, prompt treatment can be initiated to patient.

CONFLICT OF INTEREST DISCLOSURE

There is no conflict of interest to be disclosed by all authors.

CONSENT

Informed consent has been obtained from the patient with regards to the imaging pictures and details included in this report.

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