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CASE REPORT

Multidisciplinary Management of Adult Orbital Rhabdomyosarcoma*

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ABSTRACT

We report the case of a 52-year-old man who presented with a 10-day history of right eye and eyelid inflammation and intermittent diplopia following blunt trauma to the right eyebrow.

The CT and MRI scans revealed an extraconal soft tissue mass on the orbital floor with maxillary and ethmoid sinus wall destruction, which on orbital biopsy was proven to be an Alveolar Rhabdomyosarcoma. The patient had a central retinal vein occlusion due to mass effect that resulted in total visual loss at 2 months. He was referred to oncologists who treated him according to the paediatric RMS protocol and is still in remission at 2-year follow-up. Rhabdomyosarcoma is a rare tumour in adults which requires multi-disciplinary management. This highlights the necessity of considering rhabdomyosarcoma in the differential diagnosis of orbital tumours in any age group.

Keywords: Multidisciplinary treatment, orbital tumour, rhabdomyosarcoma

INTRODUCTION

Rhabdomyosarcoma (RMS) is uncommon in adults, but it encompasses over 50% of all soft tissue sarcomas in children.[1] It is an aggressive tumour, with strong tendency for local invasion, local recurrence, hematogenous and lymphatic metastasis.[2,4] In adults (>16 y) RMS usually involves the extremities (34%), genitourinary tract (31%) and only 18% arises in the head and neck region.[3] We describe a middle-aged patient with an unexpectedly discovered tumour, which was successfully treated with the paediatric RMS protocol, who had no tumour recurrence or any metastases during a two-year follow-up.

CASE REPORT

A 52-year-old man had a right eyebrow blunt trauma with subsequent bruising that subsided within few days. A week later, he developed periorbital swelling, a 3-mm proptosis followed by conjunctival chemosis and intermittent diplopia. He was treated by his physician with oral antibiotics, antihistaminic agents and corticosteroids. His symptoms deteriorated and he was referred to our department.

On examination his right BCVA was 6/9 and 6/6 on the left with restricted eye movements in all directions, swelling of his right eyelids and marked conjunctival chemosis. Optic nerve function was normal.

CT and MRI scans demonstrated $27 \times 21 \times 16$ mm extraconal soft tissue mass in the orbital floor, with cortical destruction and erosion of the roof and lateral aspect of the maxillary sinus, involvement of ethmoid sinus and inferotemporal fossa, with changes in leptomeninges (Figure 1A).

Anterior orbitotomy and incisional orbital biopsy was performed and histopathology revealed an alveolar rhabdomyosarcoma. Microscopically the tumour consisted of small spindle shaped cells to round rhabdoblast, eosinophillic cytoplasm and eccentric

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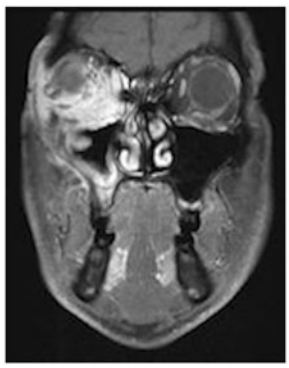

FIGURE 1. MRI head scan demonstrates extraconal soft tissue mass in the orbital floor, with cortical destruction and erosion of roof and lateral aspect of maxillary sinus, involvement of ethmoid sinus and inferotemporal fossa.

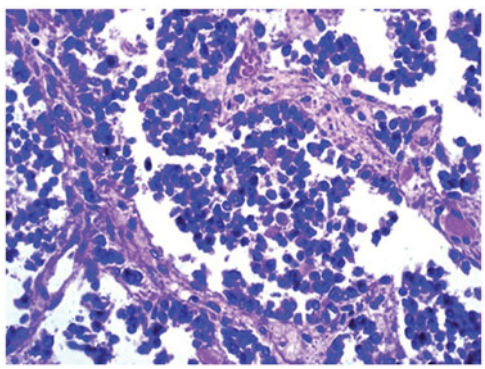

FIGURE 2. Biopsy of tumour- Alveolar RMS: small round blue cells with central cohesion, some malignant cells have septa resembling pulmonary alveoli.

nuclei (Figure 2). Cells were positive for muscle marker Desmin and transcription factor MyoD1. Due to mass effect with raised intraorbital pressure caused a central retinal vein occlusion, that resulted in significant visual loss of perception of light in the right eye.

Patient was referred to the oncology department that followed the EpSSG RMS 2005 protocol (Paediatric RMS regiment) and treated with Ifosfamide, Vincristine, Doxorubicin and Dactinomycin. He was treated every 3 weeks for up to 9 cycles −4 IVADo + 5 IVA. Good response was noted in a few weeks' time with reduction in lid swelling and conjunctival redness clinically and in the mass size radiologically (Figure 1B). External beam adjuvant radiotherapy, followed by chemotherapy, was given to orbit/maxilla and craniospinal axis, which consisted of 35 Gy into each area divided into fractions. Radical radiotherapy to orbit/maxilla given was 16.2 Gy, divided into fractions.

As part of the regiment in the current trial for very high-risk patients without metastases to commence adjuvant chemotherapy, this patient was given Cyclophosphamide orally every day and Vinorelbine

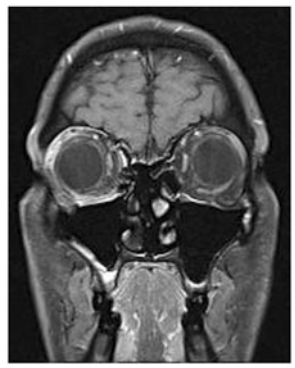

FIGURE 3. Head and orbit MRI after chemotherapy: reduction of mass size.

i/v on 1, 8, 15 day of a 28-day cycle up to 6 cycles. The patient is being followed-up with no evidence of tumour recurrence 2 years following commencement of treatment. Repeat body CT scans and MRI imaging excluded any distant metastases.

DISCUSSION

There are four major histologic subtypes of adult RMS: Embryonal, Alveolar, Pleomorphic, and Botryoid.[8] The embryonal subtype is the most common.[1] The alveolar subtype is the second most common subtype with a poor prognosis and is seen in approximately 20–30% of tumours.[5]

In this patient, there were no features to suggest the presence of this tumour prior to the injury. There are reports of RMS diagnosed in the lip and orbit after few months of injury.[7,9] Skeletal muscle (SM) regeneration that follows injury to SM has been reported to simulate histologically various malignancies such as squamous cell carcinoma and rhabdomyosarcoma.[7] Four international collaborative groups reported, in 2001, a 77% survival rate for orbital RMS in 306 children at 10 years' follow-up.[10] Less than 20 cases of adult orbital RMS are reported from 1965–2012 in the literature. Due to limited numbers of orbital RMS in adults, it is difficult to evaluate survival prognosis. The prognosis depends on anatomical localisation, tumour morphology and patient age. Despite the fact that there is no treatment protocol for adult RMS, our patient responded extremely well to chemotherapy and radiotherapy, based on children RMS protocol, which have helped tumour shrinkage and prevented any metastases so far.

CONCLUSION

Rhabdomyosarcoma is a rapidly progressive tumour which, although typically occurring in children, can rarely occur in adults. Furthermore, this case report illustrates that the current paediatric treatment

protocol can also be employed in the older patient. This case highlights the need for considering Rhabdomyosarcoma in the differential diagnosis of an orbital mass in any age group, especially where progression is rapid.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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