

## CASE REPORT

## Alveolar ethmoidal rhabdomyosarcoma in a young adult male

Andrew Kelly,<sup>1</sup> Michael Moran,<sup>2</sup> William Primrose<sup>2</sup>

<sup>1</sup>Department of Otolaryngology, Portadown, Ulster, UK  
<sup>2</sup>Department of Otolaryngology, Belfast Trust, Belfast, County Antrim, UK

**Correspondence to**  
 Andrew Kelly,  
 andrewpaulkelly@doctors.org.uk

**SUMMARY**

We report a 22-year-old man who presented with a 3 week history of left-sided headache, orbital pain and epiphora progressing to diplopia. He was being treated by his general practitioner with a  $\beta$ -blocker and simple analgesia for possible diagnosis of cluster headaches. Initial examination revealed a reduction in visual acuity and poor abduction and upward gaze. Routine blood tests were normal. CT imaging was reported as a left ethmoid sinusitis with extension into a left orbit subperiosteal abscess. No pus was obtained on frontal sinus trephine and a biopsy was taken, which proved to be an alveolar-type rhabdomyosarcoma. The patient has completed a full course of chemoradiotherapy and has responded well to the treatment. His vision is back to normal and an MRI has shown complete regression of tumour.

**BACKGROUND**

Alveolar rhabdomyosarcoma (RMS) rarely presents in patients of this age. However, owing to its aggressive nature and tendency for local spread and distant metastases, it should be considered in the differential diagnosis of someone presenting with ophthalmic symptoms of this nature.

**CASE PRESENTATION**

A 22-year-old man presented with a 3 week history of worsening left-sided headache. Associated with this he had a left-sided epiphora and a left-orbital pain. He was referred by his general practitioner (GP) when he began to complain of diplopia. He was otherwise fit and well, having recently returned from travelling in Australia and south east Asia. He had been started on simple analgesia and a  $\beta$ -blocker by his GP for his symptoms but was on no other regular medications. He had no known drug allergies. He was a university graduate who did not smoke but drank alcohol occasionally. There was no family history of note.

Upon examination he was found to be stable and was not having fever. He had a left tender submandibular node. Visual acuity was 6/9 on the left side and there was a reduction in abduction and upward gaze on the left side.

**INVESTIGATIONS**

Blood tests including inflammatory markers were normal.

Initial CT imaging was reported as a left ethmoid sinusitis with extension into a left orbit subperiosteal abscess with probable intracranial extension

through a bony defect in the floor of the anterior cranial fossa (figure 1).

Attempted frontal trephine revealed no pus. Infection remained the primary differential diagnosis at this point and his history of swimming in Thailand had raised the suspicion of infection from a rare bacteria.

Repeat CT imaging in 2 days following loss of colour vision and development of a proptosis was reported as soft-tissue density abnormality on the posteromedial aspect of the left orbital wall, which had increased slightly in size compared to previous imaging (figure 2).

Medial orbital exploration via a Lynch-Howarth incision was performed and a biopsy was taken.

Histopathological diagnosis of the biopsy obtained was RMS of the alveolar subtype.

CT of the chest/abdomen/pelvis showed no distant metastases.

**TREATMENT**

The patient was initially treated with intravenous antibiotics and pain relief. When it became clear that it was a mass and not an abscess he was also started on oral dexamethasone. Following histopathological diagnosis he was discussed at a multidisciplinary team meeting and a course of chemoradiotherapy was decided upon.

Nine cycles of IVADo chemotherapy consisting of ifosfamide, vincristine, actinomycin D and doxorubicin was given as well as 30 fractions of radiotherapy.

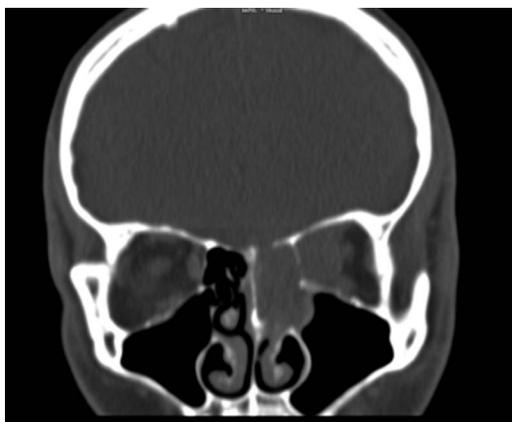
**OUTCOME AND FOLLOW-UP**

MRI of the sinuses 10 months from diagnosis showed complete regression of tumour (figure 3).



**Figure 1** Coronal view—initial CT of the brain.

**To cite:** Kelly A, Moran M, Primrose W. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-008737



**Figure 2** Coronal view—CT of the brain following loss of colour vision.

Severe inflammatory change within the maxillary sinuses was noted, which was secondary to radiation.

Left-sided vision returned to normal with only slight reduction in abduction and upward gaze.

The patient returned to his studies in January 2013 with planned follow-up in a number of months with a repeat MRI.

## DISCUSSION

RMS was first definitively described by Stout<sup>1</sup>. There are four histological subtypes of RMS that include pleomorphic, alveolar, botryoid and embryonal.<sup>2</sup> Alveolar RMS is the second most common subtype of the tumour. Embryonal RMSs are the most common of these tumours that are found in the head and neck with the alveolar subtype rarely being found in the orbit.<sup>3</sup>

All RMS tumours show a strong tendency for local invasion and local recurrence, as well as for haematogenous and lymphatic spread. They most commonly spread through the blood to the cervical lymph nodes, lungs and bone. It is generally deemed that the alveolar subtype has the worst prognosis with poorly differentiated cells and a propensity for distant metastases.



**Figure 3** Coronal view—MRI of the brain following completion of chemoradiotherapy.

CT imaging of the head and neck RMS have shown poorly defined, inhomogeneous masses with destruction of adjacent bones. In this case the CT imaging had initially queried a fluid component but repeat imaging was indicative of a solid mass and a typical adjacent destruction of bones. MRI appearances are those of an isointense or minimally hypointense mass relative to muscle on the T1-weighted images and hyperintense appearance in relation to fat and muscle on the T2-weighted images.<sup>4</sup>

The treatment approach is established following staging of the disease. The staging system commonly used is that of the intergroup RMS studies. Postsurgical grouping classification ranges from group I–IV.<sup>5</sup> Group III suggests gross residual disease locally following biopsy, as was the situation in this case. The treatment commonly involves a combination of surgery, radiotherapy and chemotherapy,<sup>6</sup> which is also the case in the clinical scenario presented. The prognosis for RMS has improved significantly over the last 30 years with the development and refinement of chemoradiotherapy regimes. Five-year survival for alveolar RMS is reported to be as high as 74% with the embryonal subtype having an observed 94% 5-year survival.<sup>7</sup>

In conclusion, RMS is a rare tumour; however, if diagnosed early can be associated with a good clinical outcome. With advances in imaging techniques and chemoradiotherapy regimes, the 5-year survival will hopefully continue to improve.

## Learning points

- ▶ Rhabdomyosarcoma (RMS) is a rare malignant tumour in young adults.
- ▶ It has a tendency for local spread and distant metastases.
- ▶ An MRI is preferred to a CT for diagnostic and follow-up investigation.
- ▶ Ethmoidal RMS should be considered in the differential diagnosis of orbital pathologies.

**Acknowledgements** Thanks to Dr B James FRCR.

**Contributors** All authors were involved in the drafting of the article and revising it critically. The conception/design and final approval were also shared.

**Competing interests** None.

**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

## REFERENCES

- 1 Stout A. Rhabdomyosarcoma of the skeletal muscles. *Ann Surg* 1946;123:447–72.
- 2 Moon H, Kwon S, Lee J. A case of alveolar rhabdomyosarcoma of the ethmoid sinus invading the orbit in an adult. *Korean J Ophthalmol* 2006;20:70–5.
- 3 Horn R Jr, Enterline HT. Rhabdomyosarcoma: a clinicopathological study and classification of 39 cases. *Cancer* 1958;11:181–99.
- 4 Lee J, Lee M, Choe D, *et al*. Rhabdomyosarcoma of the head and neck in adults: MR and CT findings. *AJNR Am J Neuroradiol* 1996;17:1923–8.
- 5 Crist WM, Anderson JR, Meza JL, *et al*. Intergroup rhabdomyosarcoma study-IV: results for patients with non metastatic disease. *J Clin Oncol* 2001;19:3091–102.
- 6 Shields C, Shields J, Honavar S, *et al*. Primary ophthalmic rhabdomyosarcoma in 33 patients. *Trans Am Ophthalmol Soc* 2001;99:133–42.
- 7 Rafsanjani K, Vossough P, Bashardoust A, *et al*. Survival rate of children with rhabdomyosarcoma and prognostic factors. *World J Pediatr* 2007;3:36–40.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.  
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact [consortiasales@bmjgroup.com](mailto:consortiasales@bmjgroup.com)

Visit [casereports.bmj.com](http://casereports.bmj.com) for more articles like this and to become a Fellow