CASE REPORT

Orbital multiple myeloma: a diagnostic challenge

Evelyn Tai,¹ See Khim Sim,^{2,3} Juhara Haron,³ Wan-Hazabbah Wan Hitam¹¹Department of Ophthalmology, University Sains Malaysia - Health Campus, Kubang Kerian, Malaysia²Department of Radiology, Hospital Sultanah Bahiyah, Alor Setar, Malaysia³Department of Radiology, University Sains Malaysia - Health Campus, Kubang Kerian, Malaysia**Correspondence to**Dr Evelyn Tai,
daileid@yahoo.com

Accepted 28 July 2017

SUMMARY

Orbital involvement in multiple myeloma is unusual. We describe the case of an 85-year-old woman who presented with right eye proptosis, reduced visual acuity and diplopia. Computed tomography showed a lobulated, enhancing soft tissue mass arising from the right greater wing of the sphenoid with intraconal, lacrimal gland and ocular muscle involvement. Histopathology revealed predominantly atypical plasma cells in a background of reactive lymphocytes, with monoclonality towards kappa light chain protein, suggestive of multiple myeloma. This case illustrates the diagnostic imaging challenge of orbital multiple myeloma.

BACKGROUND

Multiple myeloma is a malignant disease characterised by monoclonal proliferation of plasma cells, causing destructive bone lesions and other systemic manifestations. It accounts for 10% of haematological malignancies.¹ Orbital involvement in multiple myeloma is uncommon and usually reflects recurrence of disease in a patient with established multiple myeloma.² The rarity of orbital presentation of multiple myeloma may result in a delayed diagnosis of this potentially treatable malignancy.^{3–5} We present this case to illustrate the diagnostic imaging challenge of orbital multiple myeloma.

CASE PRESENTATION

An 85-year-old woman who was bedridden for a year due to chronic back pain presented with right eye painless, progressive proptosis, associated with diplopia for one week. Her family members observed that her right eye movement had been limited for a week. There was no blurring of vision, eye redness or pain on eye movement. She had no

Figure 2 Coronal view of the CT of the orbit showed a mass invading the right lacrimal gland, lateral rectus and superior rectus muscles.

history of ocular trauma. There were no symptoms to suggest a thyroid disorder, such as palpitations, heat intolerance, excessive sweating or altered bowel habits. She also denied constitutional symptoms such as fever, loss of appetite or loss of weight. There was no family history of malignancy. The remainder of the systemic review was unremarkable.

Figure 1 Axial CT of the orbits showed (A) a lobulated, avidly enhanced soft tissue mass arising from the right greater wing of the sphenoid, displacing the globe anteriorly (B) with erosion of the greater wing of the sphenoid bone.

Figure 3 Axial CT of the brain showed multiple well-defined lytic lesions of the calvarium.

CrossMark

To cite: Tai E, Sim SK, Haron J, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-220895

Figure 4 (A) Tumour tissue composed of mainly atypical plasma cells in a background of reactive lymphocytes (H&E, $\times 100$ magnification). (B) Plasma cells showed moderate nuclear pleomorphism and binucleation, with occasional mitotic figures (yellow arrow) (H&E, $\times 400$ magnification).

On examination, the best corrected visual acuity was 6/24 in the right eye and 6/9 in the left eye. There was non-axial proptosis of the right eye, with conjunctival chemosis and injection. The right periorbital area had multiple lobulated masses, with engorged superficial veins. There was complete ophthalmoplegia of the right eye. The left eye extraocular movement was full. The pupils were equal and reactive bilaterally, with no relative afferent pupillary defect. The intraocular pressure in the right eye was elevated. The right eye anterior segment was normal. Examination of the right eye fundus revealed choroidal folds but no optic disc swelling. The anterior segment and fundus examination of the left eye were normal.

Systemic examination revealed bibasal lung crepitations. Cardiovascular examination was normal, as was the breast and abdominal examination. She had no palpable lymph nodes. Neurological examination was normal.

INVESTIGATIONS

Blood investigations for infective and inflammatory causes were unremarkable, except for an elevated erythrocyte sedimentation rate of 96 mm per hour. Computed tomography (CT) of the orbits showed a lobulated, avidly enhanced soft tissue mass arising from the right greater wing of the sphenoid, measuring 3.9 cm (anteroposterior (AP)) $\times$ 3.7 cm (W; width) $\times$ 3.1 cm (craniocaudal (CC)) (figure 1). There was intraconal and extraconal extension, involving the right lacrimal gland, lateral rectus and superior rectus muscles (figure 2). It was associated with multiple well-defined lytic lesions of the calvarium (figure 3). Thoracolumbosacral radiograph showed generalised osteopaenic bones, with degenerative spine changes. Chest radiograph showed generalised osteopaenic bones with a pathological fracture of the right clavicle. In addition, there was also an opacity obscuring the left retrocardiac region, suggestive of a mass in the lower lobe of the left lung. CT of the thorax was suggested for further work up, but the patient declined further imaging.

OUTCOME AND FOLLOW-UP

Patient underwent uncomplicated tru-cut excision biopsy of the right orbital mass under local anaesthesia. Intraoperatively, the mass was spongy in consistency and occupied the temporal aspect of the right globe. Histopathological examination revealed predominantly atypical plasma cells in a background of reactive lymphocytes; the cells showed monoclonality towards kappa light chain protein, suggestive of multiple myeloma (figure 4). Unfortunately, the patient passed away a few weeks after the biopsy.

DISCUSSION

Multiple myeloma is a malignant proliferation of monoclonal plasma cells, commonly in red marrow-bearing bones such as the skull, spine, ribs, pelvis and proximal ends of long bones.⁴ It rarely involves the orbit, where the most common malignancy is lymphoma, followed by metastatic tumours.⁶ Orbital involvement in multiple myeloma may be the first manifestation of systemic disease.⁴ The proliferation of neoplastic plasma cells is termed osseous plasmacytoma when it arises from the bone and extramedullary plasmacytoma when it arises from the soft tissue.⁷ Our patient had an osseous plasmacytoma arising from the right greater wing of the sphenoid, involving the superior and lateral extraocular muscles, as well as the right lacrimal gland. This resulted in proptosis and restriction of the extraocular movement, causing diplopia. Both of these presenting clinical features have been reported in the literature.⁸ The presence of choroidal folds on funduscopy was further evidence of a compressive retrobulbar lesion, which was supported by CT findings of a soft tissue mass causing anterior globe displacement (figure 1).

The diagnosis of multiple myeloma is usually based on clinical suspicion, supported by relevant investigations. As these patients tend to present with non-specific symptoms like back pain and fatigue,⁹ radio-imaging can provide clues to the diagnosis. In patients with pathological fractures, radiography may show two diffuse patterns of bony involvement: well-circumscribed lytic lesions and, less commonly, generalised osteopaenia. Our patient demonstrated both of these variants; she had generalised osteopaenia and lytic lesions of the calvarium, with a healed pathological fracture of the right clavicle.

CT scan is the imaging modality of choice.¹⁰ Although a definitive diagnosis is not possible without histological confirmation, the site of tumour origin and its radiological features can be clearly visualised with CT scan. CT scan provides a rapid, non-invasive and accurate method of identifying the origin and extent of lesion involvement, especially in cases of retro-orbital lesions. CT may demonstrate thinning of the involved bone,¹¹ or soft tissues masses arising from the bone, with resultant bony expansion and destruction.¹² These findings in CT may mimic metastases, especially in older patients. Approximately 10% of orbital tumours in adults are metastases; orbital metastases usually originate from lung and breast cancers.¹³ Other primary malignancies that may metastasise to the orbit include kidney, pancreas, prostate and gastric cancers.¹⁴ Magnetic resonance

Learning points

- An orbital mass may be the 'tip of the iceberg'; as orbital malignancies are usually secondary to systemic malignancy, a thorough systemic workup should always be performed.
- The diagnosis of multiple myeloma requires a high index of suspicion as its presentation is insidious and its clinical features are non-specific.
- In patients with pathological fractures, radiography may show two diffuse patterns of bony involvement: well-circumscribed lytic lesions and, less commonly, generalised osteopaenia.
- CT scan is the imaging modality of choice in orbital multiple myeloma; it may demonstrate proliferative soft tissue lesions arising from the bone, with resultant bony destruction.
- Biopsy provides a definitive diagnosis and guides further management.

imaging is superior to CT in the evaluation of soft tissue and tumour extension; however, it is not readily accessible and is more time-consuming. In multiple myeloma, lesions may demonstrate low signal intensity on T1-weighted images and a high signal intensity on T2-weighted images.¹⁵

Biopsy provides a definitive diagnosis and guides further management. Discovery of a plasmacytoma should always prompt investigation for systemic involvement, as the treatment and prognosis differs between the two. In our case, although proptosis secondary to the orbital plasmacytoma was the presenting complaint, further investigation revealed end-organ damage in the form of multiple bone lesions. Treatment options for solitary plasmacytomas are radiotherapy and surgical resection, while patients with active multiple myeloma characterised by end-organ damage require systemic chemotherapy.^{16 17} As progression to systemic multiple myeloma occurs in approximately 50% of solitary plasmacytomas, long-term follow-up is necessary.¹⁶

Contributors ET and SKS wrote the manuscript. JH and W-HWH reviewed the articles.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Castro-Rebollo M, Cañones-Zafra R, Vleming-Pinilla EN, *et al.* Unilateral exophthalmos as the debut of a non-secretory multiple myeloma. *Arch Soc Esp Oftalmol* 2009;84:631–4.
- 2 Liao J, Greenberg A, Shinder R. Relapsed multiple myeloma presenting as an orbital plasmacytoma. *Ophthal Plast Reconstr Surg* 2011;27:461.
- 3 Burkat CN, Van Buren JJ, Lucarelli MJ. Characteristics of orbital multiple myeloma: a case report and literature review. *Surv Ophthalmol* 2009;54:697–704.
- 4 Knapp AJ, Gartner S, Henkind P. Multiple myeloma and its ocular manifestations. *Surv Ophthalmol* 1987;31:343–51.
- 5 Coupland SE, Krause L, Delecluse HJ, *et al.* Lymphoproliferative lesions of the ocular adnexa. Analysis of 112 cases. *Ophthalmology* 1998;105:1430–41.
- 6 Shields JA, Shields CL, Scartozzi R. Survey of 1264 patients with orbital tumors and simulating lesions: The 2002 Montgomery Lecture, part 1. *Ophthalmology* 2004;111:997–1008.
- 7 Swerdlow SH, Campo E, Harris NL, *et al.* *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*. 4th ed. Lyon: International Agency For Research On Cancer, 2008.
- 8 Rodman HI, Font RL. Orbital involvement in multiple myeloma. Review of the literature and report of three cases. *Arch Ophthalmol* 1972;87:30–5.
- 9 International Myeloma Working Group. Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Working Group. *Br J Haematol* 2003;121:749–57.
- 10 Howling SJ, Tighe J, Patterson K, *et al.* Case report: the CT features of orbital multiple myeloma. *Clin Radiol* 1998;53:304–5.
- 11 Levin SR, Spaulding AG, Wirman JA. Multiple myeloma. Orbital involvement in a youth. *Arch Ophthalmol* 1977;95:642–4.
- 12 Ajithkumar TV, Sivasankar C, Ramachandran K. Orbital multiple myeloma: case report and review of computed tomography features. *Australas Radiol* 2002;46:119–20.
- 13 Johansen S, Heegaard S, Bøgeskov L, *et al.* Orbital space-occupying lesions in Denmark 1974-1997. *Acta Ophthalmol Scand* 2000;78:547–52.
- 14 Günel I, Gündüz K. Metastatic orbital tumors. *Jpn J Ophthalmol* 1995;39:65–70.
- 15 Tuncbilek N, Karakas HM, Oken OO, *et al.* MRI in multiple myeloma with orbital and dural invasion. *Tumori* 2005;91:270–2.
- 16 Röllig C, Knop S, Bornhäuser M. Multiple myeloma. *The Lancet* 2015;385:2197–208.
- 17 Hu K, Yahalom J. Radiotherapy in the management of plasma cell tumors. *Oncology* 2000;14:101–8.

Copyright 2017 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

Visit casereports.bmj.com for more articles like this and to become a Fellow