CASE REPORT

Orbital apex tumour caused by chronic lymphocytic leukaemia: an unlikely suspect

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SUMMARY

An 88-year-old woman with a background of chronic lymphocytic leukaemia (CLL) and presented with unilateral ptosis and dull facial pains for 1 month. Examination revealed a complete right-sided ptosis and pupillary dilation. Vision in her right eye was limited to light perception. She had total external ophthalmoplegia. Her corneal reflex was not present in her right eye and she had lost sensation on the right side of her forehead. MRI revealed abnormal enhancement in the right orbital apex extending posteriorly to the sphenoid sinus. The mass invaded the superior orbital fissure, optic canal and cavernous sinus. The lumbar puncture was normal. Owing to the proximity of the mass to the cavernous sinus, it was deemed that surgical excision of the tumour was unsafe; however, it was amenable to biopsy. Histology of the biopsies was consistent with CLL.

The patient declined to undergo single high-dose radiotherapy followed by dexamethasone.

lesion. The MRI performed revealed an abnormal enhancement in the right orbital apex extending posteriorly to the sphenoid sinus (figure 1E). The mass partially eroded the orbital plate and invaded the superior orbital fissure, optic canal and cavernous sinus.

A lumbar puncture was performed to identify a cause for the mass; however, the cerebrospinal fluid was clear and had no white cells, and had normal glucose and protein. The differentials for the mass at this point ranged from fungal infection to lymphoma. Owing to the proximity of the mass to the cavernous sinus, it was deemed that surgical excision of the tumour was unsafe; however, it was amenable to biopsy. ENT surgeons performed an endoscopic maxilloethmoidosphenoidotomy. The histology of the biopsies eventually revealed lymphocytic infiltrates in the mucosal tissue. The morphological appearance and the immunohistochemistry profile of the tissue were consistent with CLL. Interestingly, there was no evidence to suggest high grade transformation.

BACKGROUND

This case demonstrates the unpredictable nature of chronic lymphocytic leukaemia (CLL). CLL has never been known to cause orbital apex masses. Furthermore, the patient had very good signs which have been photographed, with the patient's permission. Finally, the case demonstrates the correct methodology to work-up a patient with an orbital apex mass.

CASE PRESENTATION

An 88-year-old woman with a background of stage II CLL and trigeminal neuralgia admitted to the acute medical unit with unilateral ptosis and dull facial pains present for 1 month. There were no symptoms of raised intracranial pressure. General examination revealed lymphadenopathy, the largest of which was a 1.2 cm axillary node and splenomegaly.

A focused examination of face and eyes revealed a complete right-sided ptosis. When her eye was held open, the pupil was dilated. Vision in her right eye was limited to light perception. She also had total external ophthalmoplegia. Furthermore, her corneal reflex was not present in her right eye and she had lost sensation to the right side of her forehead (figure 1A–D). There were minimal retinal changes.

INVESTIGATIONS

Following the examination, it was assumed that the patient was suffering from orbital apex syndrome with the involvement of the optic nerve. An MRI was performed to identify if there was a causative

OUTCOME AND FOLLOW-UP

The patient declined to undergo chemotherapy and it was eventually agreed that she would undergo single high-dose radiotherapy followed by dexamethasone with the hope of preventing further growth of the mass. She never regained sight in the eye.

DISCUSSION

The authors report this to be the first example of CLL leading to an orbital apex mass. CLL is known to have neurological sequelae albeit rarely; however, in all of these cases, the CLL is leptomeningeal or there was the presence of lymphocytes in the cerebrospinal fluid, both of which are not present in this case.^{1–2} Cases have been reported where CLL has presented with symptomatic central nervous system (CNS) involvement.³ There are previously reported incidents of isolated facial, abducens and vestibulocochlear palsy secondary to infiltration of CLL.^{1–5} This is in marked contrast to this case as the symptoms were secondary to a mass lesion which revealed itself as a deposit of lymphocytes, rather than direct infiltration of the CNS by lymphocytes. Finally, there are reported cases of invasive, destructive fungal disease arising in patients with CLL. This was a differential considered by the authors, but it was ultimately refuted by tissue diagnosis.

This case report highlights the unpredictable nature of haematological malignancy and its ability to cause

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Figure 1 Photographs of the patient's eyes demonstrating: (1) Native appearance of the patient. (2) The patient's eyes held open. (3) The patient looking to the left. (4) The patient looking to the right. The patient's right pupil is dilated and there is complete ophthalmoplegia of all extraocular muscles. (5) MRI images demonstrating the orbital apex mass and extension to the sinuses.

a variety of neurological sequelae. It is essential to consider leukaemia or lymphoma as a differential for patients presenting with neurological symptomology with a history of haematological malignancy or with an abnormal full blood count.

Competing interests None.

Patient consent Obtained.

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Learning points

- ▶ The peculiar and unpredictable nature of haematological malignancies.
- ▶ In patients suffering from neurological problems with a history of haematological malignancy, infiltration of the tumour cells or mass effect can be suspected.
- ▶ A negative lumbar puncture does not necessarily rule out the presence of intracranial haematological malignancy.

REFERENCES

- 1 Lange C, Brouwer R, Brooimans R, *et al.* Leptomeningeal disease in chronic lymphocytic leukemia. *Clin Neurol Neurosurg* 2007;109:896–901.
- 2 Cramer SC, Glaspy JA, Efird JT, *et al.* Chronic lymphocytic leukemia and the central nervous system: a clinical and pathological study. *Neurology* 1996;46:19–25.
- 3 Akintola-Ogunremi O, Whitney C, Mathur S, *et al.* Chronic lymphocytic leukemia presenting with symptomatic central nervous system involvement. *Ann Hematol* 2002;81:402–4.
- 4 Zappia JJ, Bunge FA, Koopmann CF, *et al.* Facial nerve paresis as the presenting symptom of leukemia. *Int J Pediatr Otorhinolaryngol* 1990;19:259–64.
- 5 Nageris B, Or R, Hardan I, *et al.* Sudden onset deafness as a presenting manifestation of chronic lymphocytic leukemia. *Leuk Lymphoma* 1993;9:269–71.

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