Burkitt's neurolymphomatosis of the trigeminal nerve

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Accepted 29 July 2018

DESCRIPTION

A 48-year-old woman presented with 8 months of right-sided burning facial pain and 4 weeks of unsteadiness. She had reduced right facial sensation and corneal reflex. She had no pyramidal or cerebellar signs. Examination of the other cranial nerves was unremarkable. She had reduced sensation in all modalities along the left side of her body.

MRI showed a right-sided enhancing cerebello-pontine angle lesion indenting the mid-pons contiguous with a thickened trigeminal nerve extending towards Meckel's cave (figure 1A) with subtle enhancement of the vermis and posterior pons. There was also oedema of the pons (figure 1B). Whole-body CT and PET were unremarkable.

A benign pathology such as trigeminal schwannoma or meningioma of the cerebellopontine angle was considered. Therefore, subtotal resection was performed to decompress the pons for symptomatic relief.

Histopathology revealed a malignant infiltrate of large, mitotically active, atypical lymphoid cells, showing a starry-sky pattern (figure 2A). Ki-67 index was nearly 100%, an archetypal feature of Burkitt's lymphoma (figure 2B). Fluorescent

in situ hybridisation confirmed the presence of an IGH-MYC translocation.

Burkitt's lymphoma comprised 0.62% of primary central nervous system lymphoma (PCNSL), which itself comprises 1%–2% of all non-Hodgkin's lymphoma.¹ Cranial nerve lymphoma, other than optic nerve involvement, is extremely rare.² Adjuvant methotrexate and rituximab as per treatment for PCNSL was performed. Autologous bone marrow transplant will be considered if there is good clinical response. The prognosis for Burkitt's lymphoma of the CNS is unknown due to its rarity, but 5-year survival for PCNSL is 42%.¹

Learning points

- ▶ The differential diagnosis is a benign pathology such as trigeminal schwannoma or meningioma. However, in view of rapid progression of symptoms of crossed brainstem signs, consider a malignant process.
- ▶ In view of brainstem oedema and diffuse enhancement, consider a malignant process in the differential diagnosis.

Figure 1 MRI brain. (A) T1-weighted post-gadolinium. (B) T2-weighted image.

Figure 2 Histopathology. (A) H&E-stained section. (B) Ki-67 staining.

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To cite: Phang I, Craig-McQuaide A, Kinch K, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2018-225958

Contributors IP drafted the article and did a literature review. AC-M performed a literature review and helped draft the article. KK provided neuropathology input and images. WT approved the submission of the final article.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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