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Case report

Primary central nervous system lymphoma at the cerebellopontine angle mimicking a trigeminal schwannoma: A unique case report and literature review

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ABSTRACT

Primary central nervous system lymphoma (PCNSL) is an uncommon extranodal manifestation of non-Hodgkin's lymphoma with those presenting at the cerebellopontine angle (CPA) being rare presentations with limited reported cases in the literature. We report a 60-year old female presenting with PCNSL of the left CPA radiologically mimicking a trigeminal schwannoma with corresponding clinical signs. Imaging showed a left CPA lesion that was biopsied and confirmed as diffuse large B-cell lymphoma. Given its rarity, PCNSL should be considered in the differential diagnosis for all CPA tumours.

1. Introduction

Primary central nervous system lymphoma (PCNSL) is a rare extranodal form of non-Hogkin's lymphoma that arises in the craniospinal axis, accounting for ≤1% of all lymphomas, 4–6% of all extranodal lymphomas and ~1–3% of all primary CNS tumours [1–3]. Usually attributable to immunosuppressed patients, PCNSL can be diagnostically challenging for clinicians due to its shared radiological features with other more commonly seen pathologies. PCNSL of the cerebellopontine angle (CPA) has thus far been rarely reported within the medical literature worldwide [4–21]. To our knowledge this is the first case of a diffuse large B-cell PCNSL of the CPA mimicking radiologically as a trigeminal schwannoma.

2. Case report

A 60-year-old female presented with a 2-week history of left sided neck pain with left sided facial sensory disturbance, headache, ataxia, left sided hearing loss with associated dizziness and nausea. On admission the neurological examination revealed left sided facial hemianesthesia with loss of the left corneal reflex, left abducens palsy with left dysdiadochokinesia. The patient's past medical history included depression and migraines. The patient was HIV negative with all other hematological parameters normal with no history of immunosuppression although she did have a 40-pack year history of smoking.

MRI of the brain showed a left cerebellopontine angle mass measuring 24 mm × 15 mm (axial) × 21 mm (craniocaudal) extending into the cisternal portion of the left trigeminal nerve. The lesion was iso to hypointense on T1 and hypointense on T2 that avidly enhanced post contrast. The lesion appeared extraaxial with a cleft of CSF seemingly separating it from the adjacent pons, upon which it exerted mass effect. There was extensive vasogenic oedema throughout the pons, left cerebellar hemisphere and extending through the fibers towards the left mid brain. The fourth ventricle was distorted and there was mild dilation of the lateral and third ventricles. The cerebellar tonsils extended 5mm below the foramen magnum (Fig. 1).

On the basis of the imaging coupled with the clinical examination, a working differential (provisional) diagnosis of a left trigeminal schwannoma was considered despite an abducens palsy being an atypical sign. The patient's headache significantly improved on dexamethasone. A pre-operative MRI showed reduction in the lesion size to 23 mm × 14 mm (axial) × 16 mm (craniocaudal) as well as the vasogenic oedema. A stereotactic left retrosigmoid craniotomy was performed following insertion of an external ventricular drain for brain relaxation. The tumour, which was intra axial within the pons was accessed between trigeminal and VII/VIII complex. There was a venous loop adherent to the tumour. Multiple biopsies were taken. The frozen section returned a high-grade malignant tumour. There were no distinguishable planes between tumour and normal pons and thus further debulking was not

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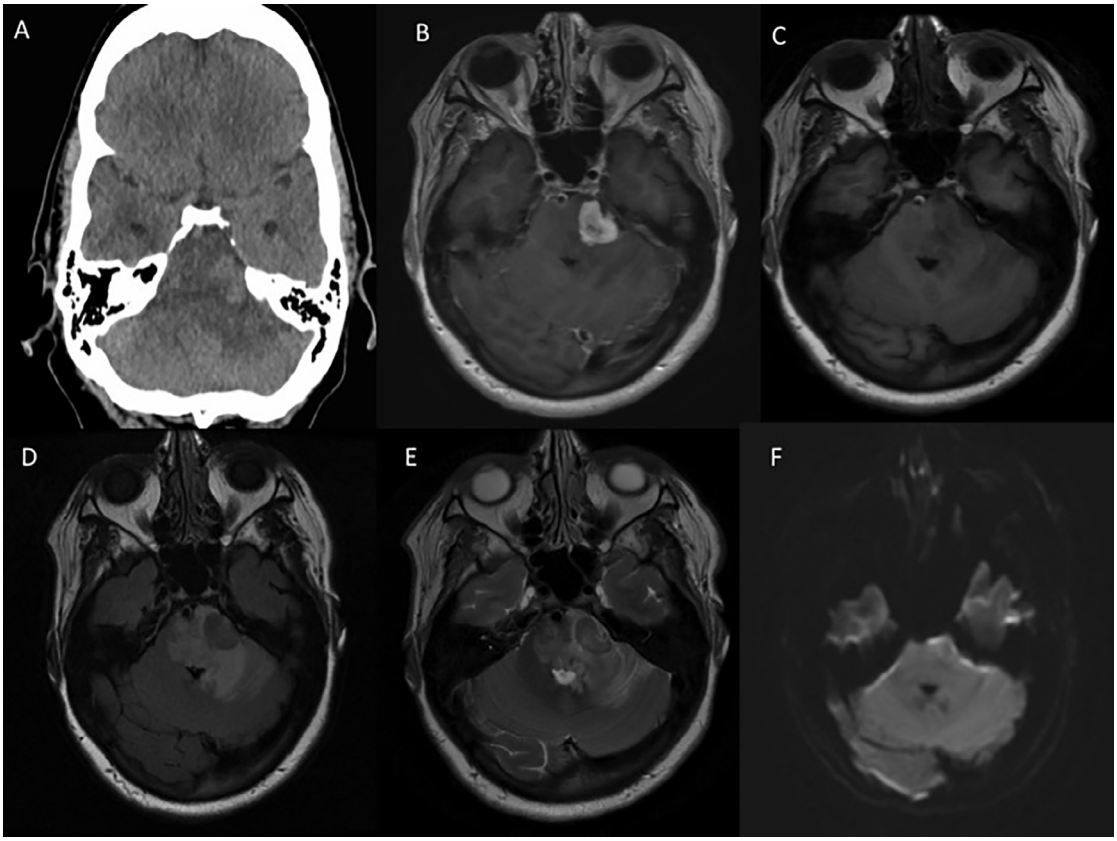

Fig. 1. A. Axial CT with hyperdense lesion at the left CPA, B. Axial T1 weighted magnetic resonance imaging with gadolinium contrast, showing involvement of the left trigeminal nerve C. Axial T1 weighted magnetic resonance imaging post contrast, D. Axial T2 weighted magnetic resonance imaging FLAIR showing extensive vasogenic oedema, E. Axial T2 magnetic resonance imaging seemingly showing a cleft of CSF around the lesion, F. Diffusion weighted imaging showing mild restriction.

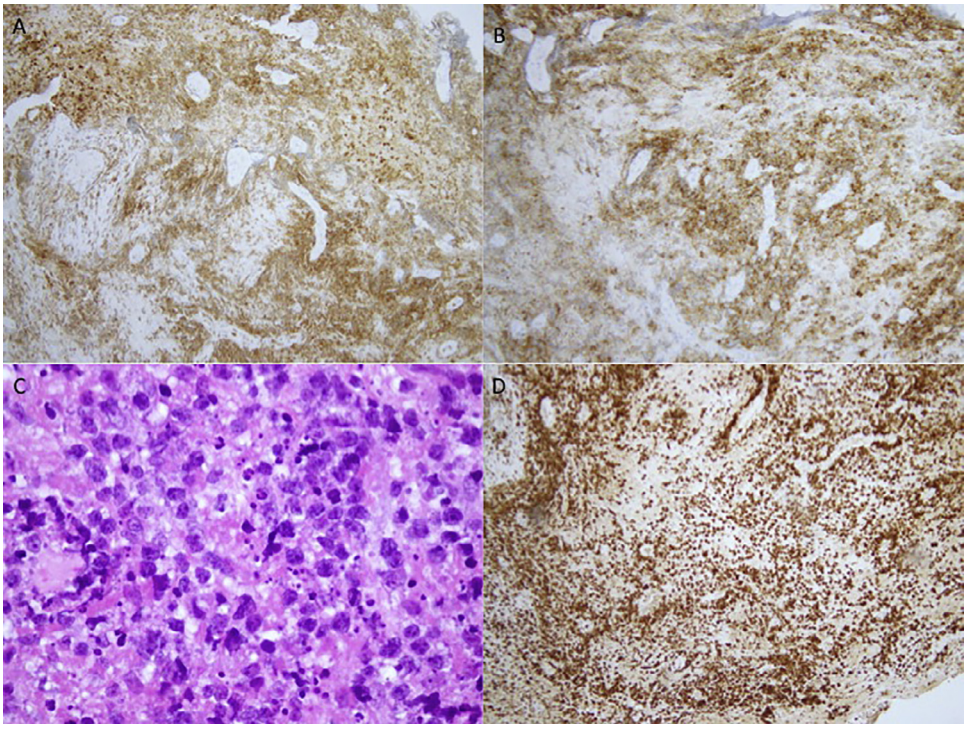

Fig. 2. A. Immunoperoxidase staining confirming tumour cells were positive for CD20 (×10 magnification) and B. positive for CD10 (×10 magnification), C. Haematoxylin and eosin staining reveals vascular nuclei, peripherally placed nucleoli and a moderate amount of cytoplasm (×60 magnification), D. Ki-67 index is greater than 95%.

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