The Journal of Laryngology and Otology
October 1993, Vol. **107**, pp. 937–939

Primary cerebral lymphoma presenting with bilateral cerebellopontine angle lesions

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Abstract
A case is described of a primary cerebral lymphoma which presented with bilateral cerebellopontine angle lesions. No similar case has been reported in the English language in the last 25 years. The literature is reviewed and discussed.

Key words: Cerebellopontine angle; Lymphoma

Case report

A 68-year-old lady presented with a six-week history of mild diplopia and hearing loss, most noticeable in the left ear. She had also been unsteady on her feet, but had experienced no rotatory vertigo. On examination, she had slight dysarthria, a positive Rombergs test and Bruns nystagmus with the fine component to the right and coarse to the left. Both corneal reflexes were reduced but there were no other neurological findings of note and no stigmata of von Recklinghausen's disease. Pure tone audiometry showed a bilateral sensorineural deafness which was worse on the left side (Figure 1) and speech audiometry demonstrated marked impairment with a PB-max of 43 per cent in the right ear and only three per cent in the left (Figure 2), suggesting retrocochlear pathology. A CT scan was performed which showed fairly symmetrical lesions of both cerebellopontine angles but no expansion of the internal auditory meatus (Figure 3). MRI confirmed these lesions which were embedded slightly more into the pons and cerebellum than usually found with acoustic neuromata (Figure 4).

After referral to the neurosurgeons, angiography and CSF cytology were performed but proved unhelpful in reaching a diagnosis. A left craniotomy was performed and the tumour removed from the cerebellopontine angle in an attempt to decompress the vestibulocochlear nerve. Post-operatively she suffered from various fluid and electrolyte problems and required a tracheostomy. Histology showed sheets of cells, many with vesicular nuclei and prominent nucleoli, and large numbers of mitotic figures (Figure 5). Immunohistochemistry (Figure 6) showed strong positive staining with L26, a B cell marker, in keeping with a B cell lymphoma. Dexamethasone was given and a course of radiotherapy started but her condition gradually deteriorated and she died of bronchopneumonia after four weeks. At postmortem examination it was found that her lymphoma had responded well to the radiotherapy with no residual tumour. There were no other deposits of lymphoma found elsewhere in the body.

Discussion

In addition to acoustic neuroma, there are a variety of uncommon tumours that may occur in the cerebellopontine angle,

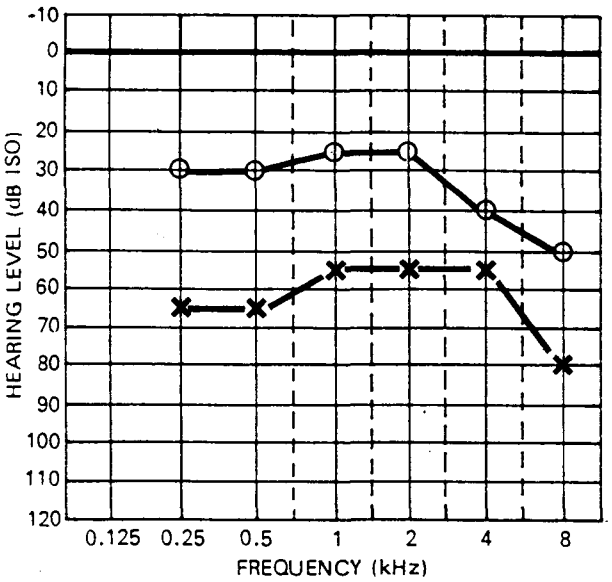

FIG. 1
Pure tone audiogram.

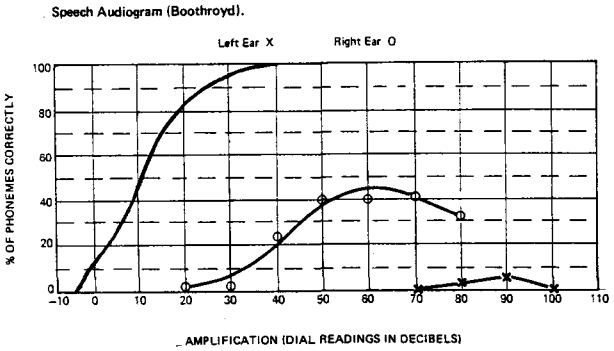

FIG. 2
Speech audiogram.

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Accepted for publication: 24 April 1993.

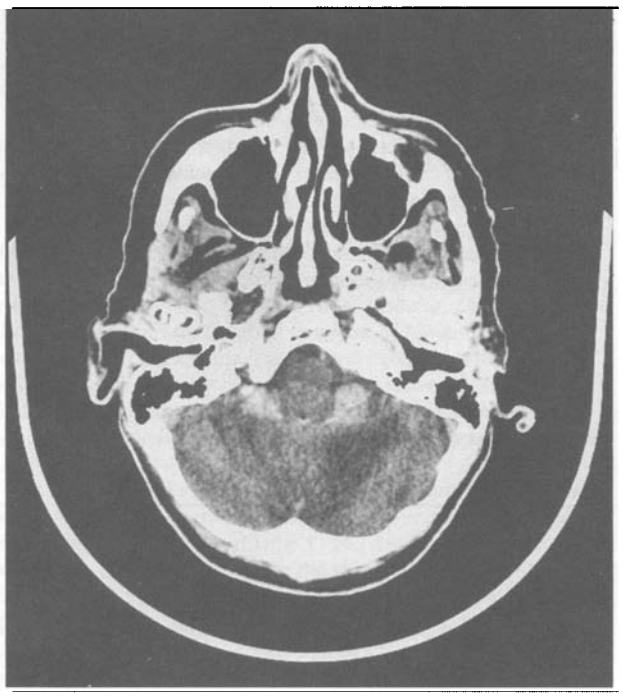

FIG. 3
CT scan demonstrating bilateral CPA lesions.

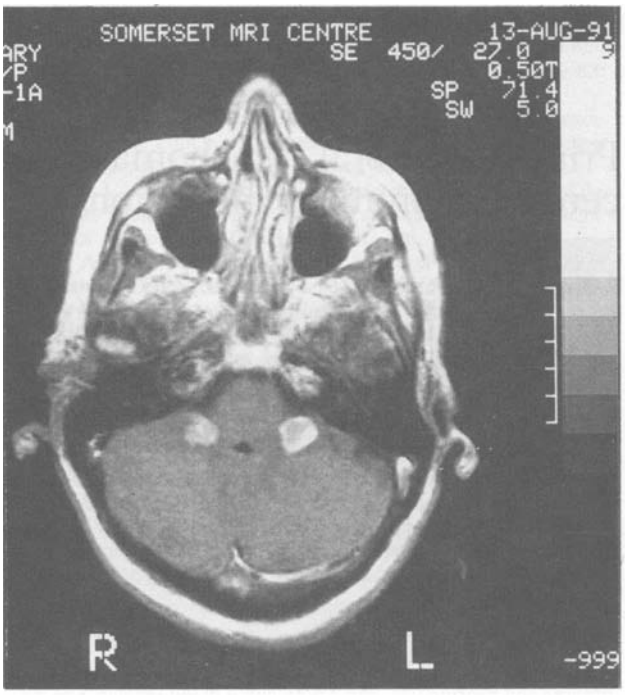

FIG. 4
MRI demonstrating bilateral CPA lesions.

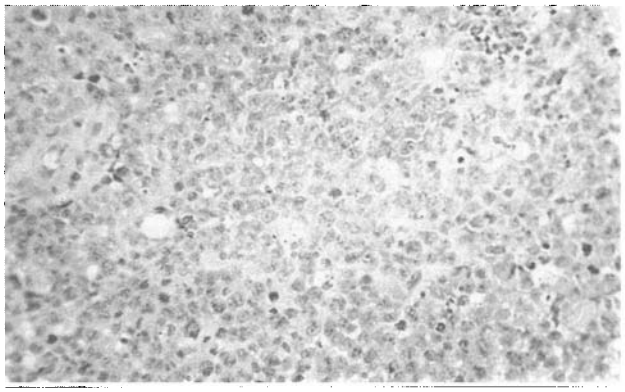

FIG. 5
Section of the left CPA lymphoma (H & E).

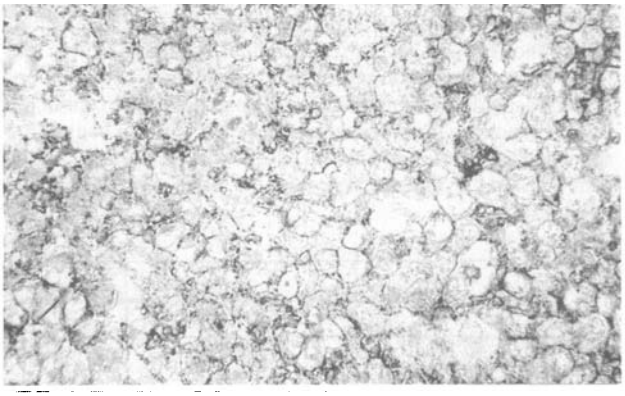

FIG. 6
Strong positive staining with B cell marker L26.

accounting for 10 per cent of lesions at this site. In a series of 1354 cerebellopontine angle tumours (Brackmann and Bartels, 1980), after exclusion of acoustic neuromas (91.3 per cent), meningiomas (3.1 per cent), cholesteatomas (2.4 per cent) and cranial nerve neurinomas (1.4 per cent), there were 25 'rare tumours'. These rare tumours included arachnoid cysts, haemangiomas, haemangioblastomas, gliomas, metastatic tumours, dermoids, lipomas and a teratoma. In a series of 32 non acoustic cerebellopontine angle tumours (Hitselberger and Gardner, 1968) haemangiosarcomas were also recorded.

Primary cerebral lymphomas are rare tumours, accounting for 0.7–0.9 per cent of lymphomas at all sites and 0.3–1.5 per cent of all intracranial tumours (Henry *et al.*, 1974; Jellinger *et al.*, 1979). Presentation has been reported between the ages of 16 days and 90 years, but this condition is predominantly one of the sixth and seventh decades, with a male to female ratio of 1.5:1 (Helle *et al.*, 1984).

Primary cerebral lymphoma may present with a variety of focal or nonfocal neurological signs and symptoms, depending on the site of the lesion. Most tumours are supratentorial, but the majority of infratentorial tumours are found in the cerebellum (Helle *et al.*, 1984). Usually patients have single lesions though 44 per cent of patients have multiple CNS lesions (Henry *et al.*, 1974). This contrasts with secondary lymphoma in which chronic leptomeningeal infiltration and multiple cranial nerve palsies are common (Mackintosh *et al.*, 1982).

In all reported cases of these tumours in the cerebellopontine angle, the diagnosis has been made on excision or biopsy. Imaging with CT or MRI are essential in the investigation and management of these patients, angiography and CSF cytology also have an important role. Valavanis *et al.* (1981) have suggested criteria for the diagnosis on CT, but the exceptional rarity of these tumours makes it unlikely that many will be diagnosed pre-operatively.

Treatment regimens have usually centred around excision followed by radiotherapy. Although few have favoured chemotherapy, there have been occasional reports of its success in patients after surgery and radiotherapy have failed (Ervin and Canellos, 1980). Helle *et al.* (1984) did not find any significant prognostic factors in a literature review of 400 reported cases, but figures from his own series suggested that diffuse mixed lymphoma and lymphomas situated below the tentorium had a

poorer prognosis. Prognosis is usually poor, with untreated patients surviving three to six months. Radiotherapy may prolong life, giving a survival of 15 to 45 months, with occasional cases surviving over 10 years (Merchut *et al.*, 1985).

An extensive literature search using the Dialog network found only four cases of primary cerebral lymphoma in the cerebellopontine angle (Valavanis *et al.*, 1981; Ierokomos and Goin, 1985; Yang *et al.*, 1987) in the last 25 years. Secondary lymphoma is more common and has also been reported at this site (Rosen, 1979; Nakada *et al.*, 1983; Yang *et al.*, 1987). No reported cases of primary cerebral lymphoma presenting with bilateral cerebellopontine angle lesions have been found.

Acknowledgements

We would like to thank Dr C. Gardner-Thorpe, Consultant Neurologist, and Mr W. E. Strachan, Consultant Neurosurgeon, who were also involved in the management of this case.

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