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Oncology in Focus

An unusual cause of acute labyrinthine failure

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Abstract
We report a case of a 40-year-old man presenting with acute vertigo and deafness. Computed tomography (CT) scanning at initial presentation was normal. However, one year later he developed numbness on the right side of his face and examination revealed fifth, seventh and eighth cranial nerve palsies as well as cerebellar dysfunction. Magnetic resonance imaging (MRI) demonstrated a cerebellopontine angle lesion. He underwent near total excision followed by neuro-axis irradiation. The main mass of the tumour projected into the cerebellopontine angle. Histology showed this to be a medulloblastoma. All features of this case are unusual; hence we discuss the pathogenesis and management of this very rare tumour.

Key words: Hearing loss, sensorineural; Vertigo; Cerebellopontine angle; Medulloblastoma

Introduction

Vertigo may be of peripheral (vestibular) or central origin. Vestibular vertigo may be associated with nausea, vomiting, deafness and nystagmus and commonly an acute history indicates a infective, iatrogenic or vascular origin. However, cerebellopontine angle tumours may present in this way and sudden deafness is often a first symptom of an acoustic neuroma lying in the cerebellopontine angle. Other diagnoses include meningioma, angle cholesteatoma and epidermal cysts (Gálvez *et al.*, 1994).

In this report we present a case of an adult cerebellar medulloblastoma presenting as acute labyrinthitis with sudden onset of deafness. Medulloblastomas mimicking a cerebellopontine angle lesion are very rare with only 15 cases in the literature. However, only one other case presented with sudden deafness (Yamada *et al.*, 1993). We analyse the importance of thorough diagnostic evaluation as well as management of this very uncommon pathological lesion.

Case report

A 40-year-old man presented with an acute episode of apparent labyrinthitis, during which he was in bed for two days due to severe vertigo and ataxia; subsequently he developed hearing loss in the right ear. Over four weeks the dizziness had settled but the deafness continued. However, the latter fluctuated and was associated with some tinnitus. Examination revealed a dull right ear drum, and pure tone audiometry showed an 80 dB sensorineural hearing loss at 4000 Hz, on the right. CT scanning, 2 mm through the posterior fossa and 1 cm through the rest of the brain, pre- and post-contrast, revealed no abnormality in the cerebellopontine angle or ventricles. He was reviewed over the following year with no deterioration in hearing on serial audiometry, but had occasional episodes of vertigo if he rushed around; he was otherwise stable.

He subsequently developed a headache and numbness on the right side of his face. He also noticed that his balance had become worse, appetite had deteriorated and he had lost twenty-eight pounds in weight since the previous winter. Examination confirmed sensorineural deafness and reduced sensation to pinprick in the maxillary and mandibular divisions of the right trigeminal nerve; fundoscopy was normal. However, there was a nonfatiguable, positional nystagmus to the left and right and also on upward gaze. He had an absent right corneal reflex and mild facial weakness on the right. His gait was mildly unsteady and heel to toe walking was difficult.

Routine radiographs of the skull and chest, as well as haematology/biochemical analysis, were all normal. An MRI scan revealed a 4 cm tumour in the right cerebellopontine angle causing distortion of the fourth ventricle, which radiologically appeared suggestive of a meningioma or an acoustic/trigeminal schwannoma (Figure 1).

He underwent a near total resection of the tumour which seemed to be sprouting from the trigeminal root/pons region, hence the root was sacrificed but the seventh/eighth neural bundle and lower cranial nerves were preserved. However, there was evidence of some infiltration in the cerebellar-brainstem junction. Histology showed this to be a desmoplastic cerebellopontine angle medulloblastoma (Figure 2).

The patient recovered slowly with a degree of residual ataxia, as well as a dense right facial sensory abnormality and mild facial palsy. He was referred for adjuvant radiotherapy. Nine months later the patient remains well with improving facial sensation, resolution of facial palsy and a mild ataxic gait. Hearing loss was unchanged.

Discussion

Cerebellar medulloblastoma is an uncommon, small cell, malignant tumour with 90 per cent of cases being reported

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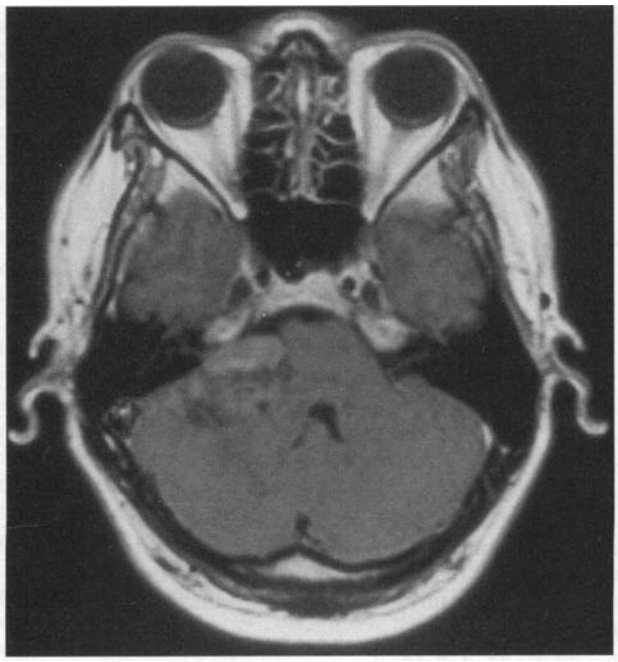

FIG. 1

MRI scan demonstrating the laterally-placed cerebellopontine angle tumour.

in children under the age of 15 and representing < one per cent of adult central nervous tumours (Smith *et al.*, 1973). Bailey and Cushing (1925) introduced the term medulloblastoma to describe a dense cellular tumour located in the vermis or the roof of the fourth ventricle and they believed the medulloblast to be a bipotential, primitive neuroglial cell able to differentiate into neuronal, glial or muscle cells (Frost *et al.*, 1995). Histologically, cerebellar medulloblastoma is a malignant growth originating from the external granular layer of the cerebellar parenchyma (Iaconetta *et al.*, 1994). This neuro-ectodermal remnant can be demonstrated as an external granular cell layer in the vermis in normal brains up to the age of 16 months (Leo *et al.*, 1997). In children the tumour usually arises as a soft, friable, midline lesion but in adults there is a tendency to originate laterally and be firm, tough and lobular (Iaconetta *et al.*, 1994). Histologically identical in adults and children, the desmoplastic variant is commoner in adults, unlike in children where the classic type is usually found, and the former varient is also associated with a better prognosis due to the increased levels of reticulin and fibrous tissue and homogenous DNA distribution (ie. stable diploid pattern) (Giangaspero *et al*, 1991).

The mean length of the clinical history in adults is seven months, and intracranial hypertension is present in 80–100 per cent of cases, while cerebellar signs are found in 50 per cent (Muller *et al.*, 1982).

Positional nystagmus appears to be any early sign of medulloblastoma as opposed to acoustic neuroma, cholesteatoma, meningioma or arachnoid cysts, which are the other main contenders in this region (Grand, 1971). While acoustic neuromas are the commonest extra-axial posterior fossa tumours, metastases and haemangioblastomas (solid/cystic) are the most common intracerebellar tumours (Koci *et al.*, 1993). The intracerebellar tumours located peripherally may be distinguished by selective angiography from medulloblastomas (Becker *et al.*, 1995).

The laterally-situated medulloblastomas are usually clearly demarcated masses with a smooth or slightly lobulated outline, unlike the infiltrating paramedian tumours, while vermian tumors are similar to childhood medulloblastomas and hence create no major diagnostic problem (Becker *et al.*, 1995). Typical CT scan findings that are found in children (homogenous enhancement after injection of contrast) are rarely seen in adults. Hyperdensity (on unenhanced scans) and diffuse contrast enhancement may be found on CT scanning; however, the primary tumour may not enhance in 10 per cent of cases (Zee *et al.*, 1982). On MRI scanning isointensity/hypointensity in T1-weighted images and hyperintensity in T2-weighted sequences are present in 60–80 per cent of cases (Hubbard *et al.*, 1989) but there is no pathognomic MRI scan appearance (Koci *et al.*, 1993). Hubbard *et al.* (1989), recognized that the laterally-placed cerebellar medulloblastoma may mimic meningiomas on CT scanning.

Uncommon appearances including cystic, necrotic, haemorrhagic and calcified multifocal cerebellar medulloblastomas are known (Zee *et al.*, 1982). The CT/MRI scan features of the tumour in adults are more variable than in children. Hence the diagnosis should be included in the differential diagnosis of a posterior fossa mass.

The treatment of choice is considered to be surgical resection (to establish adequate outflow through the fourth ventricle) and post-operative irradiation to the entire neuraxis (to prevent seeding in the cerebrospinal space) (Iaconetta *et al.*, 1994). This combination offers a 30 per

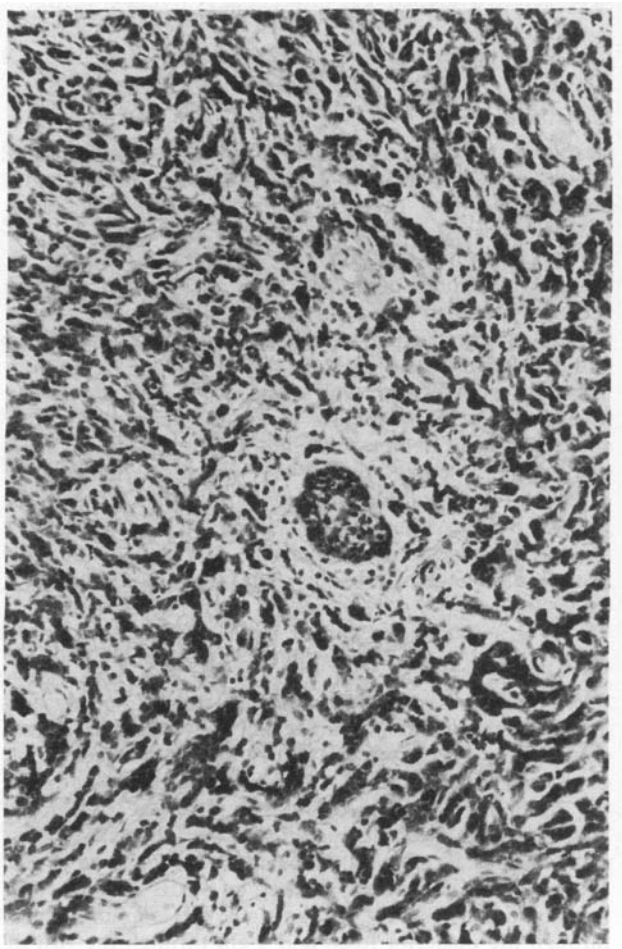

FIG. 2

Desmoplastic medulloblastoma composed of small cells with scant cytoplasm and oval/round nuclei with abundant reticulin fibres intermingled with reticulin-free pale islands. Presence of areas of necrosis, glomeruloid endothelial proliferations, Homer-Wright rosettes, neoplastic ganglion cells, nuclear polymorphism and differentiation may be evaluated and related to survival (Giordana *et al.*, 1995). (H & E; × 150).

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cent survival at five years. The role of adjuvant chemotherapy is controversial, but there are cases of improvement when used in conjunction with irradiation (Bloom and Bessel, 1990). Commonly vincristine, CCNU (1-(2-chloroethyl)-3-cyclohexyl-1-nitrosourea) and intrathecal methotrexate are used; however, others found no difference in survival (Leo *et al.*, 1997). Metastases are common via the cerebrospinal fluid and less commonly systemic metastases may occur; sites include bone, lymph nodes, lung and liver. These usually occur within two years from initial diagnosis (Becker *et al.*, 1995).

Retrospective studies have shown that five and 10-year survival rates for adult medulloblastoma are 62 and 41 per cent respectively (a wide range exists in the literature, 20–77 per cent 5 year, 10–48 per cent 10 year) (Frost *et al.*, 1995). This is no better than that for children (Leo *et al.*, 1997) but improving survival rates for more recently treated patients are seen (Bloom and Bessel, 1990). This is attributed to improved surgical skill, better imaging studies and improved radiotherapeutic techniques (Hartsell *et al.*, 1992).

Factors that predict a favourable prognosis include: lateral hemispheric location, total removal, small tumour size (<2.5 cm), good general and neurological status on discharge, radiotherapy (higher doses associated with fewer local relapses) (Ildan *et al.*, 1994) and DNA diploidy (Frost *et al.*, 1995). However, studies have refuted factors such as the age and sex of the patient affecting the prognosis (Hartsell *et al.*, 1992).

Local recurrence, central nervous system and systemic metastases make the prognosis worse and treatment is limited to repeated irradiation (conventional fractionation or stereotactically) (Frost *et al.*, 1995) and chemotherapy. Remission of 120 months is reported in a patient with bone metastases after multiple chemotherapeutic regimes (Leo *et al.*, 1997). The most common site for relapse is the posterior fossa and the best pathological correlate of recurrence is the mitotic index of the tumour (higher index, higher rate of recurrence) (Hartsell *et al.*, 1992). Belza *et al.*, (1991) felt that freedom from relapse beyond eight years can be considered as cure.

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