Diagnostic Images

Bilateral 'acoustic' neuromas

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The patient

An 18 year old youth known to have neurofibromatosis presented with progressive loss of hearing for 3 years and lower limb unsteadiness for 6 months.

Figure 1 Towne's view of skull. There is marked widening of the internal auditory meatus diagnostic of bilateral 'acoustic' neuromas (arrows). Marked calcification of the choroid plexuses is also present, the left being higher than the right (white arrows).

Figure 2 Lateral skull view shows the marked choroidal plexus calcification (arrows).

Figure 3 (a) Axial computed tomography (CT) at bone window settings showing the markedly enlarged internal auditory meatuses; (b) enlarged view.

Figure 4 Non-enhanced view of the posterior fossa (CT) with bilateral large soft tissue masses visible in the cerebello-pontine angles (arrows).

Figure 6 Following intravenous contrast there is marked enhancement of the tumours centred on the enlarged internal auditory meatuses, characteristic of acoustic neuromas.

Figure 7 An enhancing lobulated mass is also shown in the foramen magnum, found to be a meningioma on histology.

Figure 5 (a), (b), (c) CT sections through the ventricular system showing the heavily calcified choroidal plexus in the lateral ventricles as well as in the 3rd ventricle (arrow).

Comment

Neurofibromatosis or von Recklinghausen's disease is autosomal dominant, being classified as one of the neuroectodermal disorders or phacomatoses but any tissue in the body may be affected. The typical skin changes are present in more than 90% of cases, being twice as common in males, and affects 1 in 3,000 births.

The finding of bilateral 'acoustic' neuromas is pathognomonic of neurofibromatosis and associated marked calcification in the choroidal plexuses due to 'meningiomas' is uncommon but characteristic. Neuromas may occur on other cranial nerves and plexiform neuromas on the optic nerve enlarge the orbit. Aplasia of the sphenoid wings shown as a 'bare' orbit is another characteristic feature. Other tumours such as optic nerve or chiasmal gliomas and meningiomas also occur in association with neurofibromatosis. In this case densely calcified choroid plexuses, bilateral acoustic neuromas and a meningioma in the foramen magnum were present.

Incidentally, the well established designation of acoustic neuroma is a misnomer as the tumour arises on the vestibular nerve.

Reference

1. Brismar, I., Lane, B., Moselay, I.F. & Théron, J. Cranial pathology (3). In: Grainger, R.C. & Allison, D.J. (eds) *Diagnostic Radiology*, vol. 3. Churchill Livingstone, Edinburgh, London, 1986, pp 1777–1778.