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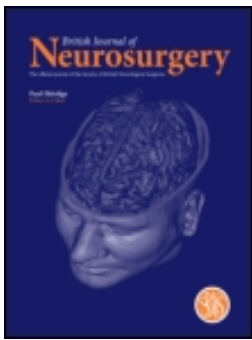


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Naim-ur-Rahman, ., Jamjoom, A., al-Rayess, M., Jamjoom, Z. A. (2000). Cerebellopontine angle medulloblastoma. *British Journal of Neurosurgery*, 14(3), 262-263.

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To cite this article: Naim-Ur-Rahman, A. Jamjoom, M. Al-Rayess, Z. A. Jamjoom (2000) Cerebellopontine angle medulloblastoma, *British Journal of Neurosurgery*, 14:3, 262-263, DOI: [10.1080/026886900408504](https://doi.org/10.1080/026886900408504)

To link to this article: <https://doi.org/10.1080/026886900408504>



Published online: 06 Jul 2009.



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NEUROSURGICAL IMAGE

Cerebellopontine angle medulloblastoma

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Introduction

Medulloblastoma, one of the most common paediatric brain tumours, occurs within the cerebellum, where the vermis is the most frequent site, especially in childhood cases,¹ but a minority of these tumours are more laterally located in the cerebellar hemispheres, especially in adults.^{1,2} A medulloblastoma with the main mass projecting into the cerebellopontine angle (CPA) is rare, and only 15 such cases were reported by 1993.¹ A medulloblastoma presenting as an entirely exophytic

CPA tumour with no apparent connection to the cerebellum or brainstem in an infant is a rarity, seldom reported in the literature.

A 3-year-old girl with a 1-year history of increasing headaches, vomiting, defective vision and right-sided deafness was admitted to the emergency department. On admission the child was irritable, drowsy and had neck-retraction. Neurological examination revealed marked bilateral papilloedema, deafness and cerebellar signs on the right side, and bilaterally upgoing

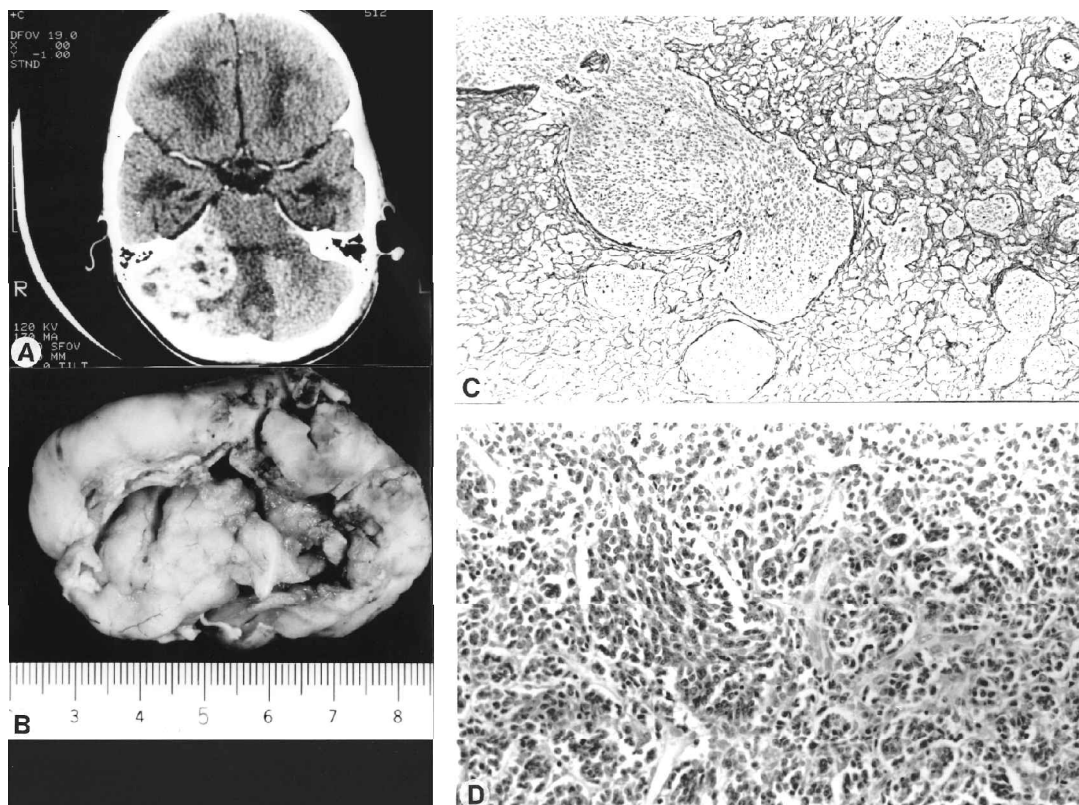


FIG. 1. (A) CT scan of the brain showing enhancing tumour in the left CPA. (B) Excised tumour specimen showing the smooth nodular surface facing the CPA. Absence of any connection or adherence to the cerebellum, as confirmed at operation, is notable. (C) Prominent reticulin fibrosis surrounding reticulin-free islands (reticulin stain, $\times 50$). (D) High power view showing the primitive nature of the tumour cells, a fibrotic background and lobular pattern.

plantars. Urgent CT revealed marked hydrocephalus with periventricular oedema and a posterior fossa tumour in the right CPA. Emergency VP-shunt resulted in marked improvement in child's condition. Post shunt CT (Fig. 1A) demonstrated an enhancing lesion (6 × 5 cm) in the right CPA. Suboccipital craniotomy and retraction of the right cerebellar hemisphere revealed a firm, well-demarcated tumour occupying the right CPA. The tumour was densely adherent to the back of the petrous pyramid especially in the region of the porus but there was no demonstrable connection with the cerebellum (Fig. 1B). The tumour was easily mobilized from the cerebellum. Complete tumour excision was achieved and confirmed by postoperative CT. Histology (Fig. 1C, D) showed a very cellular tumour with cells arranged in lobular pattern. Reticulin stain showed dense reticulin fibrous network between tumour nests. After detailed histopathological studies including immunostains, etc., a diagnosis of a desmoplastic variant of medulloblastoma was made. Postoperative course was marked by rapid resolution of symptoms except right-sided deafness and defective vision. Patient received radiotherapy postoperatively. At 1 year follow-up, there was no evidence of recurrence.

Discussion

All the reported cases of CPA medulloblastomas were in adults aged 19–46 years, were desmoplastic type and some sort of connection of the tumour to the cerebellum could be demonstrated.^{1,3} Our case is

unique and probably the first, where a desmoplastic medulloblastoma in an infant was entirely located in the CPA, having lost any demonstrable connection to the cerebellum as seen at operation. Differentiation of CPA medulloblastoma from the commoner CPA lesions, i.e. acoustic neurinomas, meningiomas, cholesteatomas and neurinomas of other cranial nerves in the posterior fossa is usually possible on clinical and radiological grounds.^{1,3,4} The rarer tumours of the CPA include exophytic astrocytoma, ependymoma, choroid plexus papilloma, medulloblastoma and paraganglioma.^{1,4,5} Differential diagnosis of CPA medulloblastoma from other rare CPA tumours requires a knowledge of their possible occurrence in this location, although a histological examination would usually be required for the final diagnosis.

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