Cerebellopontine Angle Medulloblastoma

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SUMMARY

An extremely rare case of a right cerebellopontine angle medulloblastoma in a five year old Malay girl which had eroded into the petrous bone and extended into the temporal fossa is reported. Combined subtemporal and retromastoid approach to achieve gross total surgical resection was achieved followed by radiotherapy and chemotherapy.

KEY WORDS:
Primitive neuroectodermal tumour (PNET)/medulloblastoma, cerebellopontine angle (CPA)

CASE REPORT

A five year old Malay girl presented in November 2005 with a two month history of progressive right facial asymmetry and hearing loss. She had no other symptoms. General examination was normal. Physical examination revealed right lower motor neuron VII and VIII nerve palsy. Neurological examination was otherwise unremarkable. CT scan brain (Fig. 1) and MRI showed a heterogeneously enhancing lesion at the right temporal base and cerebellopontine angle. The size of the lesion measured 3.5 X 4cm. The right internal acoustic meatus was eroded and expanded. On the MRI, the lesion was isointense in T1, T2 and not suppressed in FLAIR and enhances homogenously with contrast.

The patient underwent craniotomy a week later with a combination of subtemporal and retromastoid approach. The intraoperative finding was a greyish white soft to firm tumour, which has eroded the petrous bone and extending from internal acoustic meatus into the subtemporal area. The right VII and VIII nerves were compressed. Gross total excision was done. The histological feature was a highly cellular tumour composed of malignant tumour cells with pleomorphism and mitosis. It was positive for MIC2 and the histopathological diagnosis was a medulloblastoma (classical type). The patient recuperated well after the operation but had persistent right lower motor neuron VII and VIII nerves palsy. Repeat MRI brain (Fig. 2) and spine done a month later showed a small residual tumour (1 X 0.5cm) in the brain. She underwent radiotherapy of the craniospinal axis and chemotheraphy two months later. A routine repeat MRI of the craniospinal axis done three months later showed a drop metastasis in the spine and slightly increased size of the residual tumour. Currently she is still undergoing chemotheraphy (4th cycle of PACKER regime) and she is otherwise well and able to perform activity of daily living except for urinary incontinence.

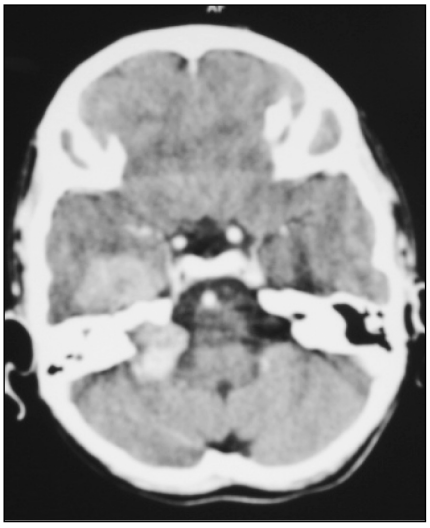

Fig. 1: CT scan brain axial with contrast: showing a Cerebello-Pontive Angle and subtemporal region lesion that enhances well with contrast eroding the internal auditory meatus (see black arrow).

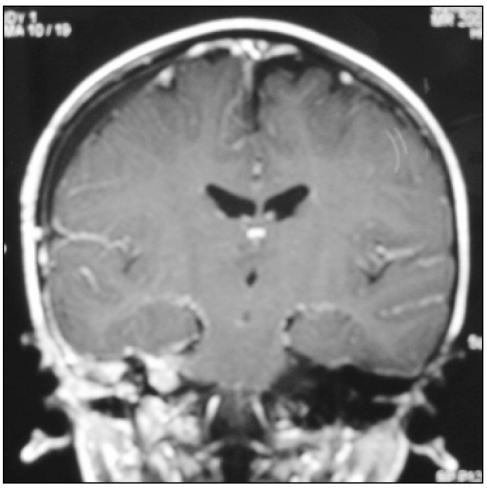

Fig. 2: MRI brain coronal with contrast: showing an enhancing residual lesion cerebellopontine angle. (see black arrow)

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DISCUSSION

Meduloblastomas arise from the roof of the fourth ventricle and superior medullary velum but the origin of the CPA region medulloblastomas is not certain and there have been several hypotheses. Among the suggestions are lateral extension of tumour from the fourth ventricle through the foramen of Lushka, or direct exophytic growth from the site of origin at the surface of the cerebellum or pons [2]. It has also been speculated that it may arise from the flocculus, which faces the CPA[4]. A possible origin for this case is the right cerebellar hemisphere with exophytic invasion of CPA, internal auditory canal and extension into the temporal fossa.

Medulloblastoma predominately affects the pediatrics, with a median age of nine years old. Medulloblastomas can have a very variable clinicoradiological pattern and biological behaviour with occurrence at unusual sites. Only 15 cases of CPA medulloblastomas have been reported in the world literature[2].

In the clinical presentation, the duration of the symptoms is usually short and progressive[2]. The patients commonly presents with cerebellar symptoms or symptoms of raised intracranial pressure (ICP) due to hydrocephalus. Diminution of hearing or facial asymmetriness is uncommon. In the literature review, only two cases had seventh and eight nerve palsies as the presenting symptoms as in our case [4].

The computed tomography (CT) features of the lesion in our patient was not consistent with appearance of medulloblastoma. Usually it enhances homogenously and intensely with contrast whereas in our case it was heterogenous. The MR imaging appearance of medulloblastoma is variable and non-specific. But it is generally hypointense on T1- weighted images and heterogenous hypo- to hyperintensity on T2 weighted images and enhances with contrasts [2]. In our case, it was difficult to ascertain correctly the preoperative diagnosis due to its variable iimaging features.

Histologically, medulloblastomas are classified by WHO as grade IV malignant tumour. There are four subtypes, classical, desmoplastic, advance neuronal differentiation and large cell medulloblastomas. In gross examination, the tumour appears as a pinkish-gray mass with cysts, areas of necrosis, or calcification may be present. Microscopically it is poorly differentiated, small cells with scanty cytoplasm and have a high mitotic index. The desmoplastic variant was considered by Kumar *et al* to have better prognosis than others in his series[4]. Immunohistochemical (MIC2 stain) has CD99 antigen which is specific for medulloblastomas but lacks sensitivity.

The goals of surgical treatment for medulloblastomas are gross total excision, and restoration of cerebrospinal fluid pathways. To achieve maximum cytoreduction, further aggressive removal of tumour from the brainstem is not advised. A high postoperative complication rate related to cranial nerves and brainstem functions of upto 26% has been reported following tumour resection involving the brainstem[5]. In our case, the tumour was not infiltrating the brain stem but compressing it, so it was possible for gross total excision without any complications. The VII and VIII cranial nerve complexes was unable to be identified and preserved.

After surgery, reassessment and Chang's Staging for this patient done at 1 and 3 months was T4/M0 and T4/M3 respectively which indicates poor prognosis. Currently, the best treatment modalities for medulloblastomas is surgery followed closely by combination of radiotherapy and chemotherapy. Many studies conclude that combination therapy has a better outcome than single treatment. Our case, was given radiotherapy with 40 Gy in 22 fractions and 8 cycles of chemotherapy (PACKER) regime (vincristine, CCNU and cisplatin). The five year survival rate is 25 - 70%[2] but is 30% for CPA tumour[4].

CONCLUSION

CPA medulloblastomas are malignant tumours with a poor long term survival. Usually these tumours occur more commonly in adults than children. This is the fourth reported case of this in a child in the English literature and at the time of submission of this paper the child was still alive.

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REFERENCES

1. Maleci A. Medulloblastoma in children and adults: A comparative study. Acta Neurochir 1992; 119: 62-67.
2. Kamil MA, Ersin E. Medulloblastoma of the cerebellopontine angle. Neurol Med Chir 2003; 43: 555-58.
3. Rorke LB. The cerebellar medulloblastoma and its relationship to primitive neuroectodermal tumours. J Neuropathol Exp Neurol 1983; 42: 1-15.
4. Kumar R, Achari G. Medulloblastomas of the cerebellopontine angle. Neurol India 2001; 49: 380-83.
5. Albright AL, Wisoff JH. Current neurosurgical treatment of medulloblastomas in children: A report from Children's Cancer Study Group. Paediatr Neurosci 1989; 15: 276-82.