

# A Rare Case of Bleeding in a Cerebellopontine Angle Epidermoid Cyst

I Zamzuri, MBBCh, J Abdullah, PhD, M Madhavan, MD (Path), A R Ariff, MMed (Radiology), Department of Neuroscience, School of Medical Science, Universiti Sains Malaysia, Health Campus 16150 Kubang Kerian, Kota Bharu, Kelantan

## Summary

Epidermoid cysts of the central nervous system are described as rare, benign, slow growing lesions with a history of high rate of recurrence even after surgical removal. This lesion is rarely located at the cerebellopontine angle and is found to be composed of solid and cystic components with close adherence to vital neurovascular structures that might complicate its removal. We present a rare case of a twenty-five year old housewife with signs and symptoms of increased intracranial pressure due to the above pathology after multiple episodes of intra-tumoral bleeding. Microneurosurgical techniques were used for tumour dissection and excision. The patient recovered well after a three-year follow-up. This is the fourth example in the index medicus of bleeding seen in a histopathologically proven cerebellopontine angle intraepidermoid cyst.

**Key Words:** Epidermoid cyst, Haemorrhage, Radiological imaging, Cerebellopontine angle

## Introduction

Epidermoid cyst or tumour is a rare benign lesion which occurs in the midline and occasionally around the cerebellopontine angle. Few such cases have been reported in the past with most of them being treated surgically<sup>1</sup>. The best diagnostic tools for these lesions are computed tomographic CT scan and magnetic resonance imaging (MRI). This case highlights features that are totally different from previously reported cases<sup>2</sup>.

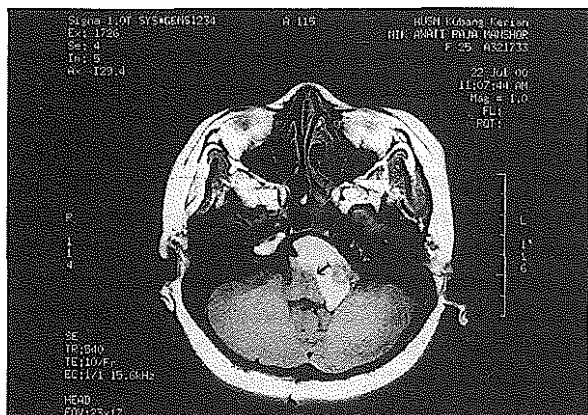
## Case Report

A twenty five year-old Malay housewife first presented with a history of generalized headache of eight-months' duration. It was throbbing in

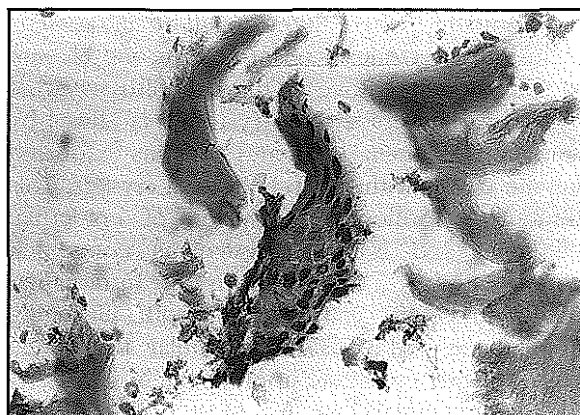
nature and associated with vomiting. There were also multiple episodes of giddiness leading to vertigo. There was no history of seizures or neurological deficits. On admission she was found to be conscious and alert. Her vital signs were normal. Neurological examination revealed bilateral horizontal nystagmus with the fast component to the left without ophthalmoplegia. Fundoscopy showed early changes of papilloedema bilaterally with slight reduction in visual acuity. CT scan of the brain revealed haemorrhage in the left cerebellum. The fourth ventricle was compressed producing a non-communicating hydrocephalus. Cerebral angiography showed no abnormality. She discharged herself against medical advice. At follow up three months later, she was found to

This article was accepted: 4 January 2002

Corresponding Author: Zamzuri Idris, Department of Neuroscience, School of Medical Sciences, Hospital University Sains Malaysia, 16150 Kubung Kerian, Kelantan



**Fig. 1:** Axial MRI T1WI showed hyperintense mass compressing the mid-brain. This lesion returns hypointense signal on T2WI.



**Fig. 2:** Histopathology of epidermoid cyst: A strip of stratified squamous epithelium surrounded by keratin flakes.

have had complete recovery of her eye signs. A year later, she was again admitted for headache, double vision and vomiting. Neurological assessment at this time revealed left third and sixth nerve palsies with nystagmus and bilateral non-reactive pupils. The Glasgow coma score on admission was 14/15. CT scan revealed an acute haemorrhage in the left cerebellar area with non-communicating hydrocephalus. MRI of the brain showed an extra-axial mass which was predominantly hyperintense on the T1WI and hypointense on T2WI consistent with an acute haemorrhage (Figure 1). The fourth ventricle was compressed causing a non-communicating hydrocephalus. A four vessel cerebral angiogram performed during this admission was again normal. Blood investigations were normal except for the total white blood cell count which was elevated to 15,600/mm<sup>3</sup>. The cerebrospinal fluid microscopy at that time revealed an increased white blood cell count of 2,420/mm<sup>3</sup> of which ninety-five percent were polymorphs. CSF cultures were negative. The patient was operated via a median suboccipital craniotomy with an infratentorial interhemispheric-cerebellar route approach. Intraoperative findings revealed a

heterogenous mixed cystic and solid lesion which was grey in colour with patchy cottage cheese appearance and was necrotic in nature. The lesion had a capsule which was stuck to the basilar artery anteriorly and the 9th and 10th cranial nerves laterally. A near total excision was made with some portions of the capsule attached on the basilar artery. Histopathological examination (Figure 2) revealed features of an epidermoid cyst consisting predominantly of flakes of keratin with a strip of stratified squamous epithelium. No skin adnexal structures were present. Post-operatively the patient developed hoarseness of voice due to vocal cord paralysis from a partial left vagus nerve palsy. She was referred to an ENT surgeon who treated her conservatively and was discharged home soon after. She was found to be healthy with complete resolution of her cranial nerve palsies noted on subsequent follow-up.

### Discussion

An epidermoid cyst is a benign, non-cancerous neoplasm. It is a congenital malformation formed by skin cells that are displaced during fetal

## CASE REPORT

development, where intracranial or intraosseous inclusion of ectoderm occurs during the closure of the neural groove at third to fifth week of gestation. The epidermoid cyst is a sac lined by squamous epithelial cells and contains exfoliated keratin debris with solid cholesterol crystals. This tumour is formed by inclusion of epidermal elements and is also known as congenital cholesteatomas or keratomas. Intracranial epidermoids are usually solid with glistening appearance and consistency similar to cottage cheese. They constitute approximately 0.2 to 1% of all intracranial tumours. Most occur laterally and are the third most common cerebellopontine angle tumours. The lesion also occur at the parasellar as well as, suprasellar regions, cerebral and cerebellar hemispheres, ventricular system and pineal region.

Vertigo, nystagmus, decreased visual acuity, gait ataxia, dizziness, trigeminal neuralgia, diplopia, tinnitus and seizures are other symptoms and in this case, the patient complained of dizziness, nystagmus, decreased visual acuity and vertigo. This could have been caused by the lesion itself which compressed the adjacent vital brainstem nervous tissue or nerve roots, since it was located at the cerebellopontine angle (CPA). The cranial nerves commonly involved are fifth, sixth, seventh and eighth nerves. In cases of epidermoid cyst, unilateral hearing loss and involvement of facial nerve occur earlier than acoustic neuroma due to its tendency to strangle the seventh nerve and reduce its blood supply<sup>1-3</sup>. The lower cranial nerve involvement as in this case, happened normally post-operatively secondary to radical resection of the tumour.

The radiological investigations of choice in epidermoid tumour are CT scan and MRI. The CT scan characteristically reveals hypodense non-enhancing lesions. The hypodensity most commonly observed on plain CT scan is due to the high lipid content deduced embryologically. However, these lesions are occasionally hyperdense on plain CT scan. This atypical feature is highlighted in this case, the reason for this could

be due to a prior bleeding into the cyst based upon hemosiderin containing macrophages in the cyst wall or an abundance of polymorphonuclear leucocytes and protein that may have also contributed to the hyperdensity<sup>2</sup>. Enhancement with contrast is uncommon and often indicates malignant transformation. Hyperdensity of CPA masses on CT scan gives rise to preoperative histological variant confusion. Meningiomas, acoustic neuromas and choroid plexus papillomas are some other important CPA tumours with hyperdense features on plain CT scan. The definitive diagnosis is not always possible on the basis of CT findings alone because of the overlap in the absorption values of above mentioned lesions and hyperdense type of epidermoid cyst which is demonstrated in this case.

MRI plays an extremely useful role in the diagnosis of epidermoid tumour with accurate delineation of the extent of tumour using its multiplanar capability and its certain characteristics. The MRI signal characteristics of epidermoids are well described with the mixture of elements within the tumours causing two types of MRI signal intensity. "Black" epidermoid-appear as low intensity on T1-weighted images, with heterogenous high signal intensity on T2-Weighted images because of their solid cholesterol content intermixed with CSF. "White" epidermoid which are less common, contains liquid triglyceride but no cholesterol and like fat appears as high signal intensity on T1-weighted images<sup>3</sup>.

In our patient, the high signal intensity on T1-weighted images was partly due to bleeding. This was evidenced by the presence of methaemoglobin within the lesion of our patient. If it was due to the high lipid or cholesterol content in the lesion, this should have manifested as hypodensity on CT which was not seen in our case. MRI with gadolinium contrast agent gives no enhancement of epidermoid lesions probably due to their minimal vascularity. This minimal vascularity was seen in two initial preoperative cerebral angiographies. MRI is now the best modality available for

monitoring the progress of tumour post-operatively. Intracranial epidermoid cyst should be treated by radical surgical resection. Literature review showed 50 - 80% of this lesion could be removed completely<sup>1</sup>. An incomplete removal has meant leaving only tiny remnants of cyst lining behind when removal of those fragments

would jeopardize underlying neurovascular structures. In this case, the basilar artery which lies close to the tumour made the excision incomplete despite the radical resection attempted by the surgeon as signified by temporary left vagus and glossopharyngeal nerve palsies.

---

### References

1. A Talacchi, F Sala, F Alessandrini *et al.* Assessment and surgical management of posterior fossa epidermoid tumours: report of 28 cases. *Neurosurgery* 1998; 42(2): 242-51.
2. Ochi M, Hayashi K, Hayashi T *et al.* Unusual CT and MR appearance of an epidermoid tumor of the cerebellopontine angle. *AJNR. Am J Neuroradiol.* 1998; 19: 113-5.
3. Dee RH, Kishore PR and Young HF. Radiological evaluation of cerebellopontine angle epidermoid tumour. *Surg Neurol* 1980; 13: 293-9.