

## A Newborn with Ptosis Secondary to a Cavernous Haemangioma

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**Abstract** Intracranial cavernous haemangioma is rare in neonates. Its natural course is still unclear. Surgical resection is recommended in symptomatic cases. A neonate suffered from intracranial cavernous haemangioma causing unilateral third nerve palsy was presented. The cavernous haemangioma resolved spontaneously in a few months time. The presentations, imaging findings and progress were reported.

**Key words** Cavernous haemangioma; Intracranial

### Introduction

Intracranial cavernous haemangioma is uncommon in neonates. Surgery is recommended in most cases. A newborn presenting with unilateral ptosis was diagnosed to have an intracranial cavernous haemangioma. The lesion regressed spontaneously over a few months time.

### Case Report

HHY was a term baby born by normal vaginal delivery with normal newborn physical examination. Her left eye was noted being smaller on day 5. Physical examination on day 18 showed complete ptosis of the left eye and bilateral retinal haemorrhage. Pupils were equal and reactive to light. No other cranial neuropathy was found. There were no cranial bruit or neurocutaneous stigmata. The other neurological examinations were normal.

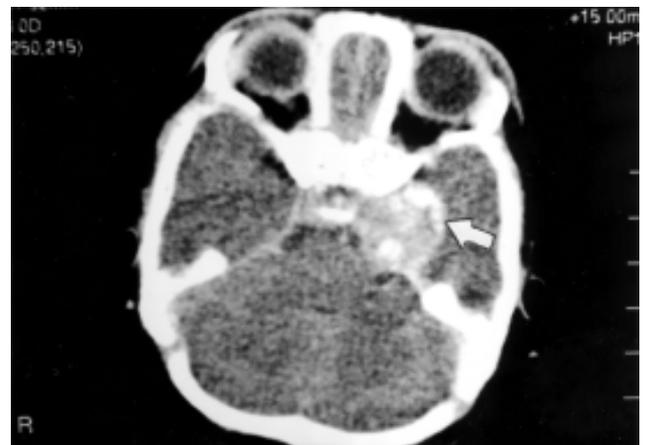
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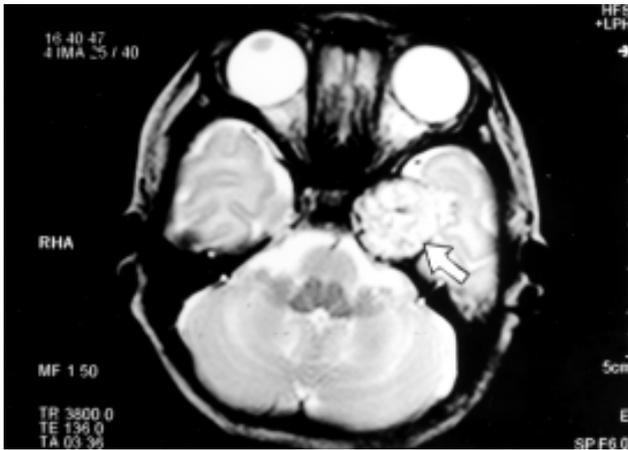
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Cranial ultrasonography showed an echogenic mass over the left temporal fossa. CT (Figure 1) and MRI brain (Figure 2) revealed an extra-axial cavernous haemangioma sized 2.2 cm in diameter over the left temporal fossa compressing the left cavernous sinus. Conservative management was adopted in view of the young age of the patient and the location of the lesion. She developed signs suggestive of progressive left third nerve palsy on day 21 when her left eyeball was noted to be displaced laterally and downward. Urgent CT brain showed similar size of the cavernous haemangioma. No acute haemorrhage was detected.



**Figure 1** CT brain axial image (post-contrast) showing a cavernous haemangioma (arrowed) at the left temporal fossa with contrast enhancement.



**Figure 2** MRI brain axial image (T2-weighted image) showing "salt and pepper" appearance of the cavernous haemangioma (arrowed) at the left temporal fossa.

She had a stable clinical course afterwards. There was no sign of raised intracranial pressure. The left eye ptosis and the extraocular eye movements improved with time. A 3-mm sized superficial capillary haemangioma was first noted over the right side of the chest wall at one month of age. At two months of age, her left eye ptosis and the ophthalmoplegia resolved totally and the size of the cavernous haemangioma started to decrease on CT scan. At eight months old, complete resolution of the cavernous haemangioma was seen on CT scan.

## Discussion

An intracranial mass lesion is always a challenge to neurologists and neurosurgeons. In recent days, intracranial cavernous haemangioma has been increasingly recognised due to the advent and easy accessibility of modern neuroimaging including CT and MRI scans. Cavernous haemangioma contributes to 5-13% of CNS vascular anomalies.<sup>1</sup> Such lesion can be found in all age groups, but it is rare in the first year of life. Some of these cases were found at autopsy in neonates, whereas the others were diagnosed during life and treated surgically.

The clinical presentations of cavernous haemangioma are mainly related to effect of space occupying lesion in the brain. The common presenting features in infancy include seizures and abnormal head enlargement.<sup>2,3</sup> Seizures are caused by a combination of mass effect and deposition of haemosiderin, which has epileptogenic properties. Supratentorial lesions are more common (85.7%).<sup>2</sup>

Complications also include lesional bleeding and infection. The risk of bleeding is usually mild and subclinical, though it increases in patients with prior haemorrhage.<sup>4</sup>

Majority shows the typical CT scan finding of a well-circumscribed hyperdense mass with low contrast enhancement and absence of adjacent oedema or significant mass effect. Calcifications are seen as many as 30% of lesions. Occasionally, cavernous haemangioma may appear to be hypodense on CT scan, which is believed to be related to old thrombosis or cystic degeneration. By MRI imaging, cavernous haemangioma shows a "salt and pepper" appearance.<sup>5</sup> A peripheral rim of very low signal density is present in all sequences and represents haemosiderin deposits within macrophages. Variable enhancement may be seen after gadolinium administration.

In our case, the cavernous haemangioma compressed the left third nerve and the left cavernous sinus, resulting in left third nerve palsy and raised intracranial venous pressure respectively. The retinal haemorrhage was likely caused by high back pressure via the retinal veins. CT brain and MRI brain confirmed the diagnosis.

The natural course of such cavernous haemangioma in infants, treated or untreated, is still unclear. The rarity of these cavernous haemangiomas in surgical series reported until recently and the inability to image them satisfactorily prior the advent of modern neuroimaging like MRI make it difficult to state with certainty the long-term prognosis of either symptomatic or asymptomatic lesions. Most case reports on infants with intracranial cavernous haemangioma were from neurosurgical case series where histopathological confirmations were available.<sup>2</sup> For years, surgical excision is the mainstay of treatment. The goals of surgery include control of progressive neurological deficits, elimination of mass effect, treatment of medically intractable seizures and prevention of haemorrhage.<sup>6</sup> Radiosurgery or gamma knife can also be used in selected cases where the cavernous haemangioma is located in inaccessible locations.<sup>7</sup> However, most lesions treated by radiosurgery have not been pathologically verified and may have included low-flow arteriovenous malformations and venous malformations as well as cavernous haemangiomas. It has not been possible to draw meaningful conclusion for the best treatment from series reported so far. Secondary haemorrhage of the cavernous haemangioma can be a complication of radiosurgery. Besides, the side effects of radiation injury are also significant for young infants.

Data concerning conservative management in these patients are limited. Regression of cavernous haemangiomas (extracranial) is common during infancy and

early childhood, and this regression has been enhanced with the use of steroids and alpha interferon.<sup>8,9</sup> The mechanism of steroids on cavernous haemangioma is not clearly understood. Steroid tends to sensitize the vascular bed to vasoconstricting agent. It seems likely that rapidly proliferating blood vessels such as those seen in cavernous haemangiomas may be highly susceptible to the mechanisms of steroid action. Alpha interferon can suppress the growth and even shrink these haemangiomas by its antiangiogenic property, as well as cytotoxic and cytostatic effects on the tumour cells. However, examples of intracranial cavernous haemangioma with spontaneous regression were seldom reported in children.<sup>10</sup>

In our case, after careful consideration and discussion among different specialists including neurosurgeons, neurologists, paediatric oncologists and neuroradiologists, conservative treatment was adopted. Systemic steroid was not given, as we really did not know how this intracranial cavernous haemangioma would behave, though steroid would be rather safe to patients provided that the patients are under regular clinical monitoring. In retrospect, we had proven conservative management is a probable treatment option instead of putting the patient into a more risky neurosurgical procedure. Indeed, our case is one of the first cases reported in the literature showing a spontaneous and complete regression of an intracranial cavernous haemangioma in a neonate with conservative treatment.

It is interesting to note that the child also has another superficial capillary haemangioma over the chest wall.

This suggested that she was prone to haemangioma malformations.

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