



A case of posterior fossa dermoid

SURESH BABU THANGAVEL

Department of Neuro Surgery,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract :

Intracranial dermoid cysts are uncommon lesions. A 35 years old female presented with complaints of swaying and falling towards the left side while walking. MRI brain revealed a posterior fossa space occupying lesion. Suboccipital craniectomy and total excision of the tumour was done. Bunch of hairs admixed with pultaceous material were found within the cyst. Histopathological examination revealed it to be a dermoid cyst. The clinical, radiological features and therapeutic strategy in the management of dermoid cysts are discussed.

Keyword : Dermoid cyst, suboccipital craniectomy

Introduction

Dermoid cysts are benign congenital subcutaneous lesions classified in a group named "Inclusion tumours"¹. Dermoid cyst is a tumour arises due to defect in embryogenesis. Dermoid cysts are the least common among the similar intracranial cystic lesions. Various studies

propose its prevalence as 0.3 per cent^{1,4,5,6,8}.

Case report:

A 35 years old female patient was admitted with headache for 2 months and swaying and falling towards left side while walking for 20 days duration. The headache was holocranial, intermittent, with no diurnal variation, not associated with vomiting, nausea or visual obscuration, partly relieved after drugs and rest. No history of loss of consciousness and seizure, no history of weakness of limbs. No history suggestive of cranial nerve and sensory system involvement. No history suggestive of bowel and bladder incontinence.

On examination her higher mental functions were normal, gaze evoked nystagmus on looking towards the left side, No papilledema. With features of left cerebellar signs - *Rapid alternating movements, finger-nose-finger test and heel-shin test were impaired on the left side. Romberg test was negative. She also had difficulty in performing tandem walking – swaying towards left side.*

CT Brain showed well defined hypodense lesion in the posterior fossa with hydrocephalus. MRI brain showed, T1 heterointense, T2 hyperintense 3.5 X 4 cm lesion in the posterior fossa Compressing left cerebellar hemisphere . The lesion showed diffusion restriction and was not enhanced with contrast suggestive of an - epidermoid.

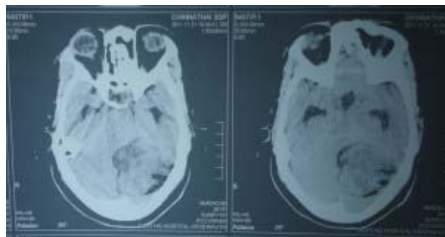


Figure 1 preoperative plain CT brain showing hypodense posterior fossa lesion with dilated temporal horns of lateral ventricle



Figure 2.a MRI brain T1 sagittal image showing heterointense posterior fossa lesion

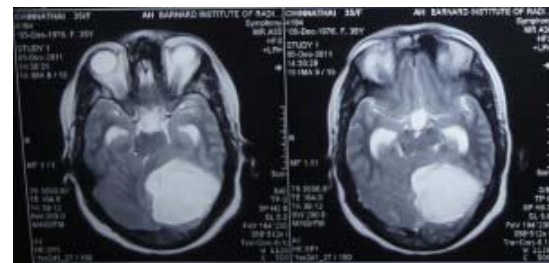
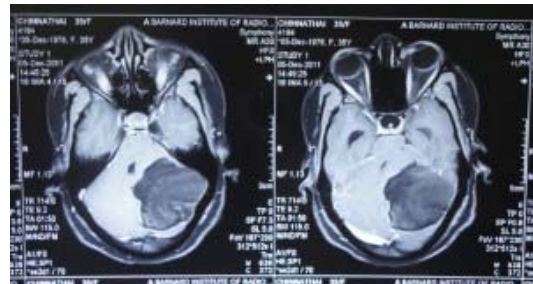


Figure 2.c MRI brain showing T2 hyperintense posterior fossa lesion

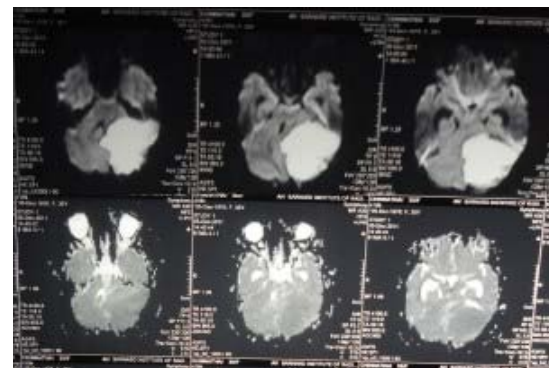


Figure 2.d MRI brain DWI and ADC showing diffusion restriction



Left paramedian suboccipital craniectomy was done. Intraoperatively the capsule was in dull white colour and on opening the capsule there were hairs admixed with tooth paste like pultaceous material.

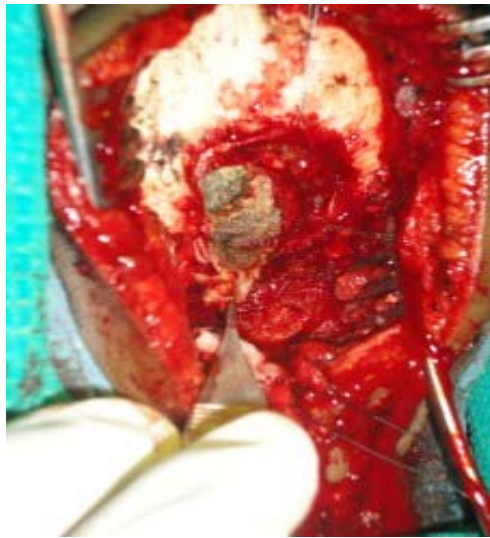


Figure 3.a Intraoperative image showing opening of capsule revealing pultaceous material



Figure 3.b Intraoperative image showing hairs admixed with pultaceous material
Figure 3.c Magnified view showing hairs admixed with pultaceous material

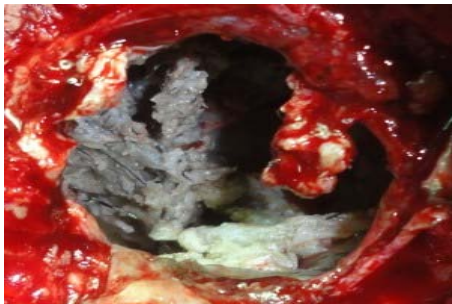


Figure 3.d Intraoperative image showing total excision of the cyst

Histopathological examination showed Dermoid cyst lined with stratified squamous epithelium with hair follicle, sebaceous glands. Postoperative period was uneventful.

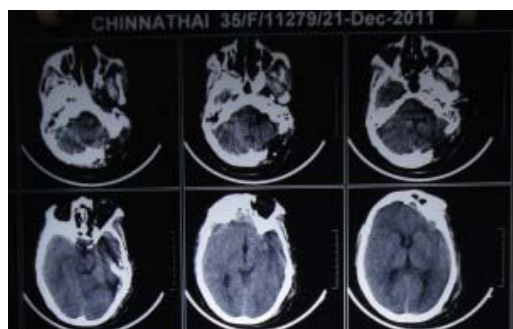
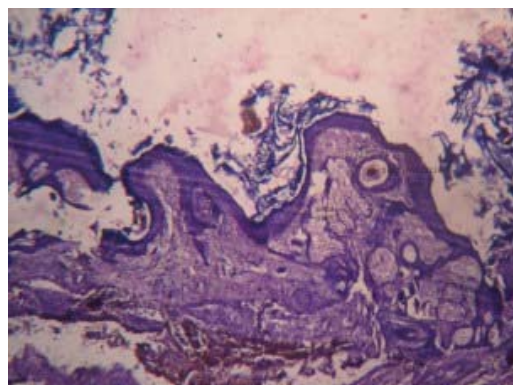


Figure 4. HPE showing Dermoid cyst lined with stratified squamous epithelium with hair follicle, sebaceous glands.

Figure 5. Post operative CT brain showing Left paramedian suboccipital craniectomy status with total excision of the lesion

Discussion:

Dermoid cysts share many characteristics with epidermoid cysts. They are also clinically and biologically benign, with the main problem at initial evaluation being related to mass effect on neural structures in a tight space (intracranial or spinal). They represent a developmental malformation, with the defect in gastrulation² affecting the surface ectoderm and causing a secondary disruption of neural tube closure. Epidermoid cysts contain epithelial cell debris and keratin, whereas dermoids contain elements of the dermis, such as hair and hair follicles and apocrine, sebaceous, or sweat glands.

Intracranial dermoids are rare congenital lesions that account for 0.04% to 0.6%^{1,4,5} of all intracranial tumours. The Dermoid cysts can be present anywhere along the embryonic fusions apart from midline. It can also occur in the Sylvian fissure. Intracranial dermoid cysts have been reported to occur in the cerebellopontine angle, cerebellar vermis, fourth ventricle, parasellar region, and frontal and frontotemporal cisternal spaces^{11,12}.

Patients with dermoid cysts are generally seen at a younger age than those with epidermoids. Gormley and co-workers noted an average age at diagnosis of 15 years for dermoid cysts versus 35 years for epidermoids³. As with epidermoid cysts, there is a female preponderance, and patients may initially be seen with local neural deficits, headache, or meningitis. Hydrocephalus and sometimes cerebellar abscesses are the principal manifestations^{1,7,8}. Posterior fossa dermoid cysts should be considered in all children with recurrent meningitis and in children with occipital skin lesions,

especially dermal sinus^{1,8}. Dermoid tumors have been reported in association with Klippel-Feil syndrome^{4,5}. Apart from epidermoid the differential diagnosis to be kept in mind is Teratoma⁹.

On CT, the lesions are usually hypodense and avascular and do not exhibit contrast enhancement. The MRI characteristics of dermoids are also similar to those of epidermoids^{1,6,9}, with perhaps even more signal heterogeneity. Relaxation time on these images may be variable, depending on the fat content of the lesion. The tumours usually show high signal levels on both T1- and T2-weighted images⁶. Because dermoid tumours are generally more solid than epidermoid tumours, they are less likely to grow between neurovascular structures and tend to demonstrate more of a local mass effect. Surrounding parenchymal oedema is lacking. Rare cases of dermoid cysts showing contrast enhancement and even an enhancing mural nodule have been reported⁶. Although it was previously thought to be uniformly fatal, dermoid cyst rupture can result in different patterns of distribution of the spilled contents, as visualized on MRI. Most commonly, fat droplets may be seen throughout the subarachnoid or intraventricular space. Another pattern is that of localized dissemination in the sulci causing widening, perhaps contained by pia or inflammatory tissue. Spinal dermoid cysts have been manifested as hydrocephalus secondary to CSF obstruction by fat droplets¹⁰. An extradural cranial lesion shows the typical bony erosive changes seen with epidermoids.

Surgical extirpation is the treatment of choice of dermoids and may be somewhat less problematic than removal of epidermoid tumors because of the firmer consistency of dermoids.

Complete excision decreases the risk for both postoperative chemical meningitis and tumor recurrence. Tumor recurrence has been reported with both cranial and spinal dermoids¹⁰. Although surgical resection of recurrent tumors may be more difficult, this should not influence the decision to reoperate. The surgeon must be cognizant of adherence of the tumor capsule to vascular and neural structures, damage to which can be devastating. This consideration may dictate a more conservative surgical approach. As might be predicted, surgical treatment of recurrent cysts remains controversial. There is no current role for radiotherapy or chemotherapy in the treatment of these tumors.

References:

1. Youmans 6th edition, chapter 136, page 1525
2. Dias MS, Walker ML: The embryogenesis of complex dysraphic malformations: a disorder of gastrulation?. *Pediatr Neurosurg* 1992; 18:229-253.
3. Gormley WB, Tomecek FJ, Qureshi N, et al. Craniocerebral epidermoid and dermoid tumours: a review of 32 cases. *Acta Neurochir (Wien)*. 1994;128:115-121
4. Turgut M: Klippel-Feil syndrome in association with posterior fossa dermoid tumour. *Acta Neurochir (Wien)* 2009; 151:269-276.
5. V.V. Pai et.al (2007) Posterior Fossa Dermoid Cysts in Association with Klippel-Feil Syndrome: Report of Three Cases. *AJNR* 28 Nov-Dec 2007
6. Brown JY, Morokoff AP, Mitchell PJ, et al: Unusual imaging appearance of an intracranial dermoid cyst. *AJNR Am J Neuroradiol* 2001; 22:1970-1972.
7. Bayram Çrak et.al *Journal of Pediatric Neurology* 2004; 2(3): 161-164 Cerebellar dermoid cysts with hydrocephalus
8. Valentine Logue and Kenneth Till et al. *J. Neurol. Neurosurg. Psychiat.*, 1952, 15, 1.
9. Anne G. Osborn, MD, FACR. *Diagnostic Imaging Brain*.
10. Cavazzani P, Ruelle A, Michelozzi G, et al: Spinal dermoid cysts originating intracranial fat drops causing obstructive hydrocephalus: case reports. *Surg Neurol* 1995; 43:466-469.
11. S. Sethi et al : Ruptured sylvian fissure dermoid presenting with MCA infarction : a rare case report. *The Internet Journal of Radiology*. 2010 Volume 12 Number 1. DOI: 10.5580/1429
12. Hidenori sugano et al. Temporal lobe epilepsy caused by dermoid cyst. *Neurol Med Chir (Tokyo)* 46,206-209, 2006.