Rare disease

Intradiploic epidermoid cyst overlying the torcula: a surgical challenge

Amad Naseer Khan,¹ Salema Khalid,¹ Syed Ather Enam²

¹Medical School, Aga Khan University, Karachi, Pakistan;

²Department of Neurosurgery, Aga Khan University, Karachi, Pakistan

Correspondence to Amad Naseer Khan, amadnaseer@gmail.com

Summary

Intradiploic tumours are rare, slow growing tumours that can present in many different ways, including a painless lump, tenderness, headache and rarely with focal neurological signs. The authors present the case of a gentleman in whom the tumour presented in an unusual location and presented a surgical challenge. The major take home message from this case report is that an epidermoid cyst overlying the torcula is to be approached with great caution and care and is without doubt a surgical challenge.

BACKGROUND

Cranial epidermoid cysts are slow growing, uncommon benign tumours which constitute less than 1% of all cranial tumours.^{1 2} Of these cranial epidermoid tumours, 75% are located intradurally and 25% are located within the diploic spaces.³ They develop from displaced dorsal midline ectodermal rest cells between the 3rd and 5th gestational weeks during the neural tube closure.⁴ They vary in size, location and rate of growth. Their most common presentation is a long-standing, asymptomatic lump on the head. Headache and focal tenderness may be present. Rarely, large lesions may be associated with focal neurological signs.⁵

Intradiploic epidermoid cysts are rare in themselves let alone those overlying the torcula herophili or confluence of sinuses – the connecting point of the superior sagittal sinus, straight sinus and occipital sinus.⁴ We could only find two cases^{5 6} in our literature review which reported an intradiploic epidermoid cyst overlying the torcula. Also, various studies have looked at the different radiological³ and neurosurgical^{5 7–12} aspects of these tumours but none have presented it as a testing surgical procedure. We present the case report of an intradiploic epidermoid cyst of the occipital bone overlying the torcula which served as a surgical challenge.

CASE PRESENTATION

Case history

A 50-year-old man who was a known case of benign prostatic hyperplasia presented to our clinic in May 2009 with a 4-year history of an on and off headache. The pain was diffuse, had no radiation, was sharp in character, gradual in onset and had been worse for the past 3 months. The pain had no aggravating or alleviating factors and currently had no associated symptoms. Over the previous 4 years, he had been treated for allergic rhinosinusitis by a GP and then an ENT specialist and even visited a General Surgeon when he felt that his headache was associated with a lump at the back of his head. On examination however, the surgeon could not appreciate any mass. Finally, he consulted

a neurologist who ordered a CT scan of his head which showed a mixed attenuating lesion with widening of the diploic space in the occipital bone with a possible breach in the outer table suggestive of a sinus formation (figure 1A). These findings were suggestive of an intradiploic dermoid/epidermoid cyst. A screening MRI to further evaluate the lesion was obtained after neurosurgical consultation (figure 1B,C).

Examination

His general physical and systemic examinations including his neurological examination were all unremarkable. With regards specifically to signs of raised intracranial pressure, funduscopy was performed and no papilloedema was noted. No mass was palpable at the posterior aspect of the skull and no tenderness was present.

INVESTIGATIONS

Imaging

CT showed a mixed attenuating lesion with widening of the diploic space in the occipital bone with a possible breach in the outer table suggestive of a sinus formation. These findings were suggestive of an intradiploic dermoid cyst (figure 1A). A screening MRI brain was done which showed a high signal intradiploic lesion seen in the left occipital region which was highly suggestive of an intradiploic tumour. There was no evidence of involvement of the brain parenchyma except for the mass effect created by the lesion figure 1B,C. One year postop MRI of the brain showed no evidence of recurrence. Both MRIs appeared to be normal, barring the normal postoperative changes that were to be expected figure 2A,B.

The rest of his routine blood tests were within normal limits.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of intradiploic epidermoid cysts includes dermoid cyst, hemangioma, eosinophilic granuloma and, in the orbitofrontal region, cholesterol

Figure 1 (A) CT findings highly suggestive of an intradiploic dermoid cyst. (B,C) A screening MRI brain showing a high signal intradiploic lesion in the left occipital region highly suggestive of an intradiploic tumour.

Figure 2 (A,B) One year postop MRI of the brain showed no evidence of recurrence. Both MRIs appeared to be normal, barring the normal postoperative changes that were to be expected.

granuloma.⁸ Differentiation between dermoid and epidermoid cysts is based on histological examination, as was done in our case, which confirmed the diagnosis of an epidermoid cyst. Dermoid cysts are more frequently diagnosed in childhood and epidermoid cyst in adult life. Dermoid cysts are commonly located in the orbital region and the midline.¹³ Hemangiomas usually have a typical, but non-specific appearance of a honeycomb or radiating sunburst pattern. The main differential diagnosis in children is eosinophilic granuloma. Atypical epidermoid cysts are not characteristic, being difficult to differentiate them from other lytic skull lesions.¹⁴

TREATMENT

Operation

A left suboccipital craniotomy was performed and the tumour excised. At operation a semicircular scalp flap was lifted and a thinned shell-like outer table of the skull was revealed. Craniectomy around the tumour was done in a manner that the tumour was totally excised along with a 1.5 cm margin of healthy bone. The bone flap along with the tumour mass was carefully detached and lifted off the underlying dura, sinuses and torcula. On removing the thinned-out outer table of bone mass, semisolid cheesy material and small amount of haemorrhage were encountered and sent for histological examination. Its inner table appeared very thin and the underlying dura mater was intact throughout. The inner cyst wall was found to be overlying the torcula and extending to the left sagittal sinus. Special care was taken not to damage the underlying dura, as damage to the torcula, an area of immense sensitivity and importance had to be avoided at all costs. Gelfoam was placed over the dura mater and venous sinuses in order to form a protective layer over the torcula. For the cranioplasty, the bone flap was evacuated of the tumour and then autoclaved. Polymethyl methacrylate (bone cement) was filled in the defect and drilled to match the shape and contour of the bony defect. Finally, it was sutured in place using silk sutures.

Pathological findings

Histologically, the diagnosis of an epidermoid tumour was confirmed. The removed tumour also contained some acellular eosinophilic material and the cyst wall had a foreign body giant cell reaction with cholesterol clefts and sheets of histiocytes.

OUTCOME AND FOLLOW-UP

Postoperative course

The patient had an uneventful postoperative course. One year postop MRI of the brain (figure 2A,B) showed no evidence of recurrence. Both MRIs appeared to be normal, barring the normal postoperative changes that were to be expected.

DISCUSSION

Although clinically insignificant, distinguishing between epidermoid and dermoid cysts can only be done histologically, as was done in our case. Most intradiploic epidermoid cysts manifest clinically as small, asymptomatic lumps in the scalp. The most common symptoms are tenderness or

headache. Intracranial hypertension, seizures, traumatic rupture or focal neurological signs have been described in patients with large cysts.² In our patient none of these symptoms or signs were present though he did complain of headache. Our patient had a cranial epidermoid cyst but can they also grow intracranially as well. Intracranial epidermoid tumours are most common in the cerebellopontine angle and suprasellar and parasellar regions.¹⁵

Interestingly, we could only find two case reports that had an intradiploic epidermoid tumour that involved the torcula.^{5 6} Our patient had the tumour overlying but not yet invading or compressing the torcula, unlike the aforementioned cases. Needless to say, the torcula is an area of great importance and damage to it intraoperatively had to be avoided by all means. Thus, this case was a surgical challenge and had to be approached with great caution and care. The various measures we took are already mentioned in the 'operation' section and include the meticulous and cautious removal of the cyst in order to avoid damage to the underlying dura. Secondly, we autoclaved the bone cement in order to sterilise it as there was no room for infection in such a delicate area and finally we placed gelfoam over the bony defect at all times to avoid iatrogenic damage intraoperatively.

Various authors have emphasised complete removal of the cyst,^{10 11} which is something we adhered to in our case. On the patients' 1 year follow-up visit an MRI was done which showed no signs of recurrence postoperatively. Also, subsequent follow-up visits till the writing of this case report have been unremarkable. This lack of recurrence of the tumour, thus supports complete cyst removal.

Learning points

- ▶ Intradiploic tumours are rare, slow growing tumours that can present in many different ways, including a painless lump, tenderness, headache and rarely with focal neurological signs.⁸
- ▶ The major take home message from this case report is that an epidermoid cyst overlying the torcula is to be approached with great caution and care.
- ▶ It is without doubt a surgical challenge.
- ▶ Complete cyst surgical removal is the favoured treatment option.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Ciappetta P**, Artico M, Salvati M, *et al*. Intradiploic epidermoid cysts of the skull: report of 10 cases and review of the literature. *Acta Neurochir (Wien)* 1990;**102**:33–7.
2. **Guridi J**, Ollier J, Aguilera F. Giant intradiploic epidermoid tumor of the occipital bone: case report. *Neurosurgery* 1990;**27**:978–81.
3. **Narlawar RS**, Nagar A, Hira P, *et al*. Intradiploic epidermoid cyst. *J Postgrad Med* 2002;**48**:213–14.
4. **Tubbs RS**, Oakes WJ. The torcular herophili is not synonymous with the confluence of sinuses. *Neuroanatomy* 2002;**1**:14.
5. **Abramson RC**, Morawetz RB, Schlitt M. Multiple complications from an intracranial epidermoid cyst: case report and literature review. *Neurosurgery* 1989;**24**:574–8.

6. **Lam CH**, Solomon RK, Clark HB, *et al.* Reversal of increased intracranial pressure with removal of a torcular epidermoid: case report. *Neurosurgery* 2001;**48**:929–32.
7. **Rengachary S**, Kishore PR, Watanabe I. Intradiploic epidermoid cyst of the occipital bone with torcular obstruction. Case report. *J Neurosurg* 1978;**48**:475–8.
8. **Han G-Y**, Won Y-S, Yang J-Y, *et al.* Intradiploic epidermoid cyst of the skull. *J Korean Neurosurg Soc* 2005;**38**:69–70.
9. **Atabay H**, Bardakci SI, Ersahin Y, *et al.* Giant primary intradiploic occipital bone epidermoid tumour: a case report. *Turkish Neurosurgery* 1994;**4**:165–8.
10. **Curarino G**, Rutledge JC. Temporoparietal dermoid cysts with intracranial extension. *AJNR Am J Neuroradiol* 1988;**9**:385–7.
11. **Cushing H**. A large epidermal cholesteatoma of the parietotemporal region deforming the left hemisphere without cerebral symptoms. *Surg Gynecol Obstet* 1922;**34**:557–66.
12. **Salazar J**, Vaquero J, Saucedo G, *et al.* Posterior fossa epidermoid cysts. *Acta Neurochir (Wien)* 1987;**85**:34–9.
13. **Curarino G**, Rutledge JC. Temporoparietal dermoid cysts with intracranial extension. *AJNR Am J Neuroradiol* 1988;**9**:385–7.
14. **Arana E**, Latorre FF, Revert A, *et al.* Intradiploic epidermoid cysts. *Neuroradiology* 1996;**38**:306–11.
15. **Vion-Dury J**, Vincentelli F, Jiddane M, *et al.* MR imaging of epidermoid cysts. *Neuroradiology* 1987;**29**:333–8.

This pdf has been created automatically from the final edited text and images.

Copyright 2011 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Khan AN, Khalid S, Enam SA. Intradiploic epidermoid cyst overlying the torcula: a surgical challenge. *BMJ Case Reports* 2011;10.1136/bcr.08.2010.3245, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow