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Amad Naseer Khan

Salema Khalid

Ather Enam

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BMJ Case Reports

Intradiploic Epidermoid Cyst Overlying the Torcula - A Surgical Challenge.

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TITLE OF CASE

Intradiploic Epidermoid Cyst Overlying the Torcula - A Surgical Challenge.

AUTHORS OF CASE *Please indicate corresponding author by* *(*after the author's name*)

Amad Naseer Khan*, Salema Khalid, Syed Ather Enam MD

SUMMARY Up to 150 words summarising the case presentation and outcome

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. We 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 Why you think this case is important – why you decided to write it up

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 spaces³. 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 signs³.

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 sinus⁴. We could only find two cases^{5,16} in our literature review which reported an intradiploic epidermoid cyst overlying the torcula. Also, various studies have looked at the different radiological³ and neurosurgical⁵⁻¹² aspects of these tumours but none have presented it as a testing surgical procedure. We present the case report of an intradiploic epidermoid cyst overlying the torcula which served as a surgical challenge.

CASE PRESENTATION **Presenting features**, medical/social/family history

Case History:

History: A 50 year old man who was a known case of Benign Prostate Hyperplasia presented to our clinic in May of 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 breech in the outer table suggestive of a sinus formation (Fig 1a). These findings were

suggestive of an intradiploic dermoid/epidermoid cyst. A screening MRI to further evaluate the lesion was obtained after neurosurgical consultation (Figs 1b & 1c). *Examination:* His general physical and systemic examinations including his neurological examination were all unremarkable. With regards specifically to signs of raised intra-cranial pressure (ICP), fundoscopy was performed and no papilloedema was noted. No mass was palpable at the posterior aspect of the skull, and no tenderness was present.

INVESTIGATIONS *If relevant*

<u>Imaging:</u>

<u>Fig 1a</u>.CT showed a mixed attenuating lesion with widening of the diploic space in the occipital bone with a possible breech in the outer table suggestive of a sinus formation. These findings were suggestive of an intradiploic dermoid cyst.

<u>Figs 1b & 1c:</u> 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.

Figs 2a & b: One year post-op MRI of the brain showed no evidence of recurrence. Both MRIs appeared to be normal, barring the normal post-operative changes that were to be expected.

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

DIFFERENTIAL DIAGNOSIS If relevant

The differential diagnosis of intradiploic epidermoid cysts includes dermoid cyst, hemangioma, eosinophilic granuloma and, in the orbitofrontal region, cholesterol granuloma⁷. 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 midline¹³. Hemangiomas usually have a typical, but nonspecific 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 lesions¹⁴.

TREATMENT If relevant

<u>Operation</u>: A left sub-occipital 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, semi-solid cheesy material and small amount of hemorrhage 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 do 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.

<u>*Pathological Findings:*</u> Histologically, the diagnosis of an epidermoid tumour was confirmed. The removed tumour also contained some acellular eosinophillic material and the cyst wall had a foreign body giant cell reaction with cholesterol clefts and sheets of histiocytes.

OUTCOME AND FOLLOW-UP

<u>Postoperative course</u>: The patient had an uneventful postoperative course. One year post-op MRI of the brain (Figs 2a & 2b) showed no evidence of recurrence. Both MRIs appeared to be normal, barring the normal post-operative changes that were to be expected.

DISCUSSION *including very brief review of similar published cases (how many similar cases have been published?)*

Although clinically insignificant, distinguishing between epidermoid and dermoid cysts can only be done histologically, as was done in our case. Most intradiploic epidermoid cysts are 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 cysts². 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 regions¹⁵.

Interestingly, we could only find two case reports that had an intradiploic epidermoid tumour that involved the torcula^{5,16}. 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 intra-operatively 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 sterilize 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 intra-operatively.

Various authors have emphasized complete removal of the cyst^{9,10}, which is something we adhered to in our case. On the patients' one 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/TAKE HOME MESSAGES 3 to 5 bullet 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 signs⁷.
- 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.

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Figure cantions
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