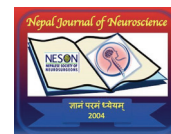


Intra-diploic epidermoid cyst: An iceberg lesion

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Abstract

Intraosseous epithelial inclusion cysts of the skull, presenting as lytic defects, constitute a very small percentage of the primary intracranial tumours. We report a 45-year-old female patient presented with a small scalp swelling in the occipital region. During initial exploration, the surgeon suspected an intracranial extension due to a large bony defect at base and inability to reach the inferior margin of the swelling, consequently the procedure was abandoned. Imaging revealed a well-defined, mixed density lesion with sharp marginated bone defect involving both the outer and inner tables of the occipital bone. Subsequently the lesion was approached through a right occipital craniotomy. Pearly white, flaky contents of the lesion along with the capsule was excised completely. The case is presented by virtue of not only the rarity of the variant but also to highlight the importance of timely intervention by a neurosurgeon after adequate investigation and in a tertiary care setting.

Key words: Epidermoid, Intra-diploic, Scalp lesions.

Introduction

Intraosseous epithelial inclusion cysts of the skull, presenting as lytic defects, constitute a very small percentage of the primary intracranial tumors.¹ These cysts are derived from ectodermal cells of cranium and are lined by stratified squamous epithelium. Although rare, their common locations include frontal, parietal and occipital bones.² We intend to report a case of intra-diploic epidermoid cyst of occipital bone presenting as a small scalp swelling in a middle aged female patient. The case is presented by virtue of not only the rarity of the variant and site of occurrence but also to highlight the importance of timely intervention by a neurosurgeon after adequate investigation and in a tertiary care setting.

Case Report

A 45-year-old female patient presented to a local Primary Healthcare Centre (PHC) with a small scalp swelling in the occipital region. She was not advised any imaging and after routine blood examination, excision of swelling was planned under local anaesthesia. Intraoperative identification of intracranial extension was made by the surgeon as the inferior margin of the swelling could not be reached and also by palpation of the huge bone defect following which the procedure was abandoned midway and the patient was referred to our centre for further management.

On examination, a residual swelling which was partially excised in its superficial portion was noticed along with an open cavity extending intracranially via the bone defect. There was active bleeding from the residual lesion which was sutured and the patient was shifted for imaging.

Imaging

Contrast Enhanced Computerised Tomography (CECT) revealed a well-defined, mixed density lesion with hypodense and an isodense component in the right occipital region. Lesion measured 4.2 (Cranio caudal) x 3.3 (Antero posterior) x 3.6 (transverse) cm. A sharp marginated bone defect was noted involving both the outer and inner tables of the occipital bone.

Lesion displayed a thin, mildly enhancing membrane on the inner aspect causing mild mass effect on right posterior parietal and occipital lobes. Enhancing dural vessels were seen along this membrane (Figure 1).

Magnetic Resonance Imaging (MRI) of the brain confirmed the CECT findings in terms of size and site of

Access this article online

Website: <https://www.nepjol.info/index.php/NJN>DOI: <https://doi.org/10.3126/njn.v18i3.34433>

HOW TO CITE

Punia P, Chugh A, Gotecha S. Intra-diploic epidermoid cyst: An iceberg lesion. *NJNS*. 2021;18(3):69-72.

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ISSN: 1813-1948 (Print), 1813-1956 (Online)



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location and revealed a T1 hyperintense and T2 mixed intensity lesion with peripherally enhancing membrane on the inner aspect. Restricted diffusion was evident on Diffusion Weighted Imaging (DWI). Thinning and destruction of inner and outer tables was noted and there was no perilesional oedema. Keeping in mind the clinical picture and imaging findings, a diagnosis of an intra diploic epidermoid cyst in the right occipital region was made and the patient was taken up for definitive surgery.

Intraoperative findings

Lesion was approached through a right occipital

craniotomy wherein the margins of bone defect were nibbled away to gain a wide access to the lesion. Pearly white, flaky contents of the lesion along with the capsule were identified and excised completely. Dura mater was intact and the capsule was easily separable from the dura. Patient underwent cranioplasty using artificial cranial bone graft (Figure 2).

Histopathological examination (HPE) revealed fibrocollagenous tissue and stratified squamous epithelium with a granular layer composed of keratinous flaky material. Mild haemorrhage and lymphocytic infiltration was also noted (Figure 3).

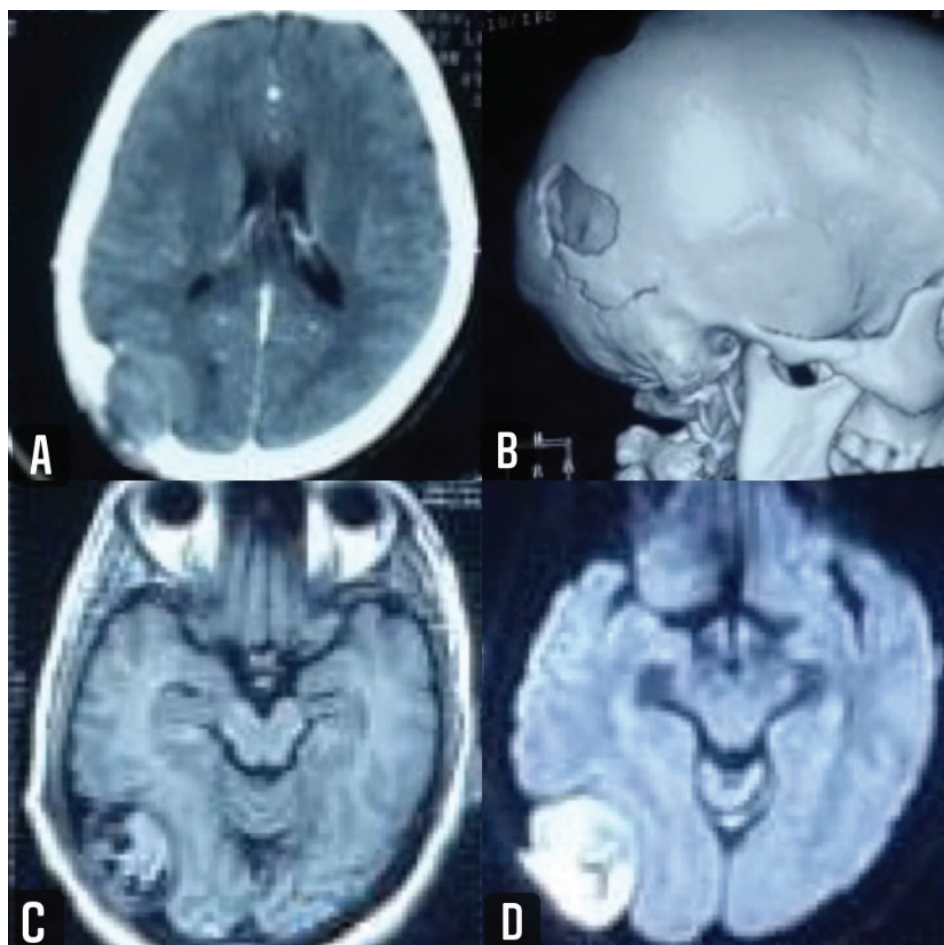


Figure 1a: Contrast Enhanced Computed tomography (CECT) brain, axial view showing non contrast enhancing lesion in the right parieto occipital region with minimal peripheral contrast enhancement.

1b: 3D reconstructed bony window of CT brain showing bone defect at the site of lesion.

1c: T1 weighted Magnetic Resonance Imaging (MRI), axial view showing a hyperintense lesion in the right parieto occipital region.

1d: Diffusion Weighted Imaging (DWI) showing restricted diffusion in the lesion.

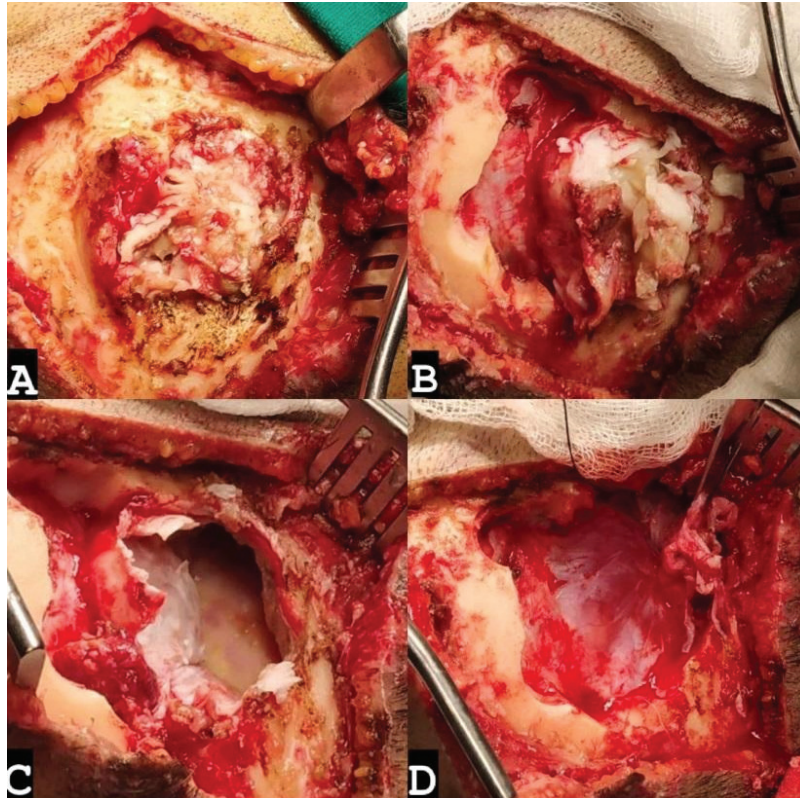


Figure 2a: Intraoperative photograph showing exposure of the scalp lesion, its size and whitish contents.
2b: Intraoperative photograph after widening of bone defect via craniotomy, thus revealing the intracranial part of the lesion.
2c: Intraoperative photograph showing pearly, white capsule of the lesion after decompression and evacuation of its contents.
2d: Intraoperative photograph showing capsule of lesion being dissected off dura and thinned out dura.

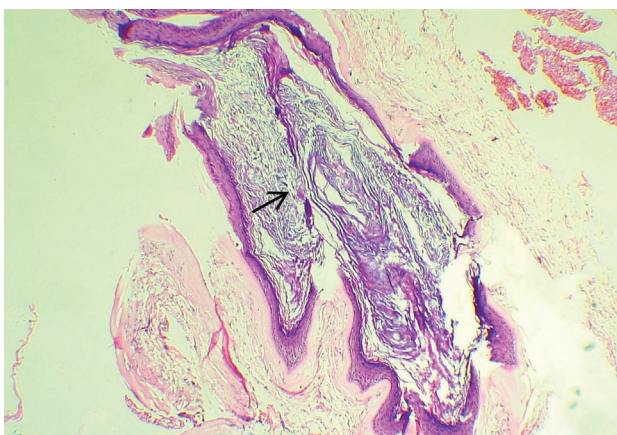


Figure 3: Photomicrograph showing cyst wall lining of squamous epithelium with presence of granular cell layer.

Discussion

Cushing first described a diploic epidermoid cyst in 1922.³ These cysts are benign soft tissue tumours that develop from abnormal sequestration and inclusion of the surface ectoderm along the lines of skin fusion.⁴

Intra-diploic epidermoid cysts are very rare, accounting for <3% of all intracranial epidermoid cysts.⁵ These cysts grow very slowly and usually present as painless bony swelling under the scalp.⁶ Prior et al in their study of 234 patients with dermoid and epidermoid swellings of the scalp demonstrated a very small ratio (9.28%) of epidermoid cysts that caused full thickness erosion of bone.⁴ Swisher, in his article titled “epidermoid cyst of the skull causing displacement of brain” stated that there may be little or no clinical evidence apart from a painless swelling below the scalp, occasionally associated with a palpable bone defect.⁷

Because of the ability of these cysts to enlarge, patients are thought to be at higher risk of constant cranial erosion and expansion into the epidural space.⁸ Although the above said hypothesis is largely accepted and supported by the natural history of the disease, Prior et al in their largest series of dermoid and epidermoid cysts of the scalp, failed to find any association between age at surgery and bone erosion and thus suggested a more complex physiopathology of cysts producing full thickness cranial erosion, such as a deeper localization of the original site of

entrapment of the surface ectoderm.⁷

CECT and MRI are the best investigations to make an accurate diagnosis and these lesions typically appear as hypodense, non-enhancing lesions and on MRI demonstrate high signal intensity in T1 weighted images and a variable T2 weighted signal and sometimes, the cyst contents can be hyperdense, mimicking a haemorrhage.⁹ No or minimal peripheral contrast enhancement may be seen. Diffusion restriction is typically seen on DWI and it is considered the best imaging sequence in diagnosing epidermoid cysts.⁶

Gross total excision remains a cornerstone for the treatment of epidermoid cysts. As proposed by Cushing, the aim of surgery is complete removal of tumour, together with its capsule, which must be carefully dissected from the bone and dura mater.³ Complete resection is to be aimed, as the only living and growing part of the intradiploic epidermoid cyst is its capsule which must be excised in its entirety to avoid recurrence.⁶ Skardowa in his article on benign paediatric cranial vault tumours observed constant enlargement of tumours in 1/3rd of their patients and advocated complete tumour excision as it not only provides a complete recovery but also the material for an ultimate diagnosis.¹⁰

This case also brings forth the importance of proper clinical and radiological evaluation not only to assess the tumour in its entirety but also aid in proper planning. Had the patient been subjected to imaging at the first presentation, the full extent of lesion would have been demonstrated and an unnecessary surgery would have been avoided. The surgeon was probably misled by the smaller extracranial sized scalp lesion whereas the much larger and aggressive intracranial part was hidden in plain sight like an iceberg and the identification of the same intraoperatively led to abandonment of the procedure.

The authors stress on the use of radio imaging even for visibly small lesions on scalp to gain all around knowledge about the tumour and extrapolate the same to the surgical modality to aid in complete resection.

Conclusion

Cranial epidermoid is a fairly common entity and intra-diploic variant of the same is not commonly seen in neurosurgical practise. These lesions may present as a small scalp lesion which should not be judged based on its apparent size as these lesions are not infrequently known to have a bigger intracranial extension like an iceberg and they should incite a doubt in the surgeon's mind to hold the knife and further investigate the patient as being forewarned is being forearmed.

Conflict of Interest: None

Source(s) of support: None

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