

Dura-Based Intracranial Epidermoid Cyst in a Young Adult Female: Report of A Rare Case

Dr. Mallika Pal¹, Dr. Indranil Das², Dr. Susmita Mukhopadhyay³, Dr. Rathin Hazra⁴, Dr. Suman Ghosh⁵

¹Associate Professor, Department of Pathology, NRS Medical College and Hospital, Kolkata, India

^{2,3}Demonstrator, Department of Pathology, NRS Medical College and Hospital, Kolkata, India

^{4,5}Assistant Professor, Department of Pathology, NRS Medical College and Hospital, Kolkata, India

*Corresponding author

Dr Indranil Das

Email: buntyrules83@gmail.com

Abstract: Intracranial epidermoid cyst is a rare occurrence. Here, we present a case of eighteen year old young female who presented with chief complaints of swelling in the left occipital region. No other complaint was present. Both computed tomography and magnetic resonance imaging showed space occupying lesion eroding inner and outer tables of skull bone at left occipital region and abutting meninges. Craniotomy and removal of the mass was done and the tissue was subjected to histopathological examination which revealed a squamous epithelium lined cystic structure containing lamellated keratinous material. No dermal appendage was present in the cyst wall. So the diagnosis of epidermoid cyst was given. No further treatment was given to the patient and no abnormality was detected on her first follow up visit.

Keywords: Epidermoid cyst, dermoid cyst, keratin, dermal appendages, cyst.

INTRODUCTION

Epidermoid cyst, or pearly tumour, is congenital in origin and accounts for about 1% of intracranial tumours [1]. It is a benign extracerebral intradural lesion and in about 40% of cases is located in the cerebellopontine angle, representing the third most frequent lesion, after acoustic neuroma and meningioma [2]. Here we present a rare case of extracerebral dura based epidermoid cyst in a young adult female arising in the left occipital region which is not a common site of presentation.

CASE HISTORY

An 18 years old female presented with chief complaints of swelling over the left side of the upper part of occipital region for last four years. The swelling was gradually increasing in size. There was no history of headache or pain over the swelling. Initially it was approximately 2 cm x 2 cm in size but attained the size of 4 cm x 3 cm over the period of four years. Any history of trauma was absent. CT scan of brain suggested focal osteolytic lesion measuring 16mm x 13mm being filled up with soft tissue in the left occipital region along with focal bulge of the overlying scalp (Figure 1). Thin connection of the lytic lesion with the underlying extra axial space had also been noted as well. Following that MRI brain was done and it suggested 23mm x 22mm x 13mm mildly enhancing space occupying lesion at left occipital bony calvarium eroding its inner and outer tables and abutting meninges (Figure 2). The patient underwent craniotomy and excision of left occipital SOL followed by refreshening of the bony margin. Grossly it was a well encapsulated

greyish white cystic mass measuring 30mm x 15mm (Figure 3) and containing greyish white oily material on cut section. Microscopic examination of the tissue showed the presence of lamellated keratin material in a cystic cavity lined by keratinizing squamous epithelium. No dermal appendage was present in the wall of the cyst (Figure 4 and 5). So the diagnosis of intracranial epidermal cyst was given and further treatment was given. On her first follow up visit at one month, neurological status of the patient was evaluated and no abnormality was detected.

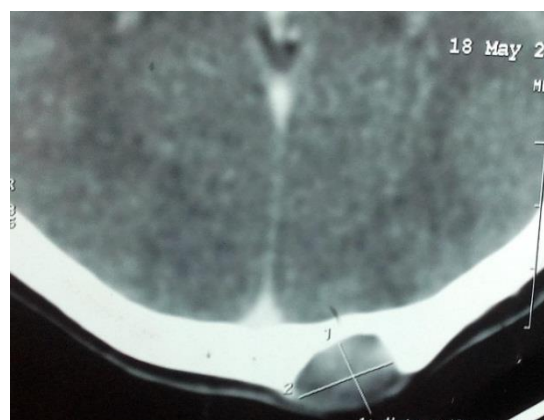


Fig-1: CT scan of brain



Fig-2: MRI of brain

Figure 1 shows CT scan of brain revealing focal osteolytic lesion in the left occipital region along with focal bulge of the overlying scalp and figure 2 shows MRI brain revealing mildly enhancing space occupying lesion at left occipital bony calvarium eroding its inner and outer tables and abutting meninges

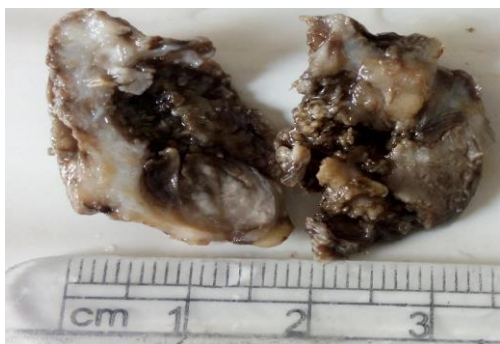


Fig-3: Gross appearance of the resected specimen

Figure 3 reveals gross appearance of the resected specimen with greyish white cystic mass containing greyish white oily material.

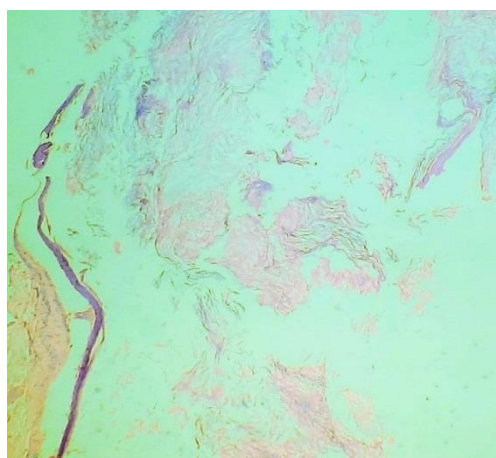


Fig-4: Histopathological examination of the tissue shows a cystic structure lined by stratified squamous epithelium without any dermal appendage in subepithelial tissue (HE x 100)

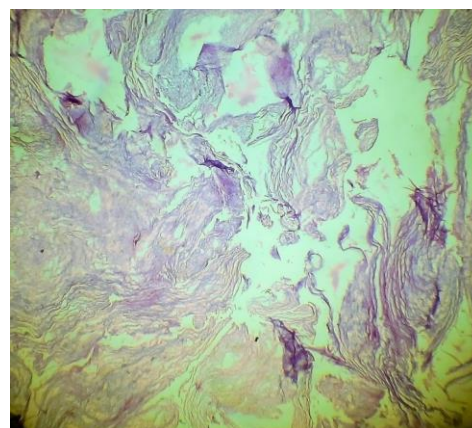


Fig-5: Cystic cavity contains lamellated keratin (HE x 400)

DISCUSSION

Two major variants of ectodermally derived neuraxis cyst are recognised, both lined by keratinising squamous epithelium. The epidermoid cyst, by definition, is devoid of cutaneous adnexal structures and filled by friable, often lamellated keratinous debris that radiates a pearly sheen as viewed through the thin, fibrous lesional capsule. Dermoid cysts, by contrast, are endowed with skin appendages. But both of them are maldevelopmental in origin. They probably arise from entrapment of surface ectodermal element along with developing central nervous system during closure of the neural groove or formation of secondary cerebral vesicle [3]. Acquired variants of epidermoid cyst resulting from iatrogenic or traumatic implantation of cutaneous tissues in the cranial or spinal subdural space are also well documented [4, 5]. Most common age groups of presentation of intracranial epidermoid cysts are young adulthood or middle age and our case also presented in her 18 years of age. Whereas intracranial dermoid cysts are common in childhood or adolescence.

Cysts of epidermoid type though are widely distributed along the neuraxis, most of them are located intracranially, cerebello-pontine angle being the most common location. Rarely may they occur in posterior fossa, parasellar region, suprasellar region, ventricular system, pineal region or in the cerebral hemisphere. In view of that, location of the cyst in our case is also not so common in occurrence. But one important thing to mention is that intraspinal epidermoid cysts are usually extramedullary and intradural in location. On the other hand, dermoid cysts usually occur in the midline. Clinical manifestations of both dermoid and epidermoid cysts are mainly due to mass effects but sign and symptoms of chemical meningitis may occur due to cyst rupture and spillage of irritating keratinous and lipid rich debris into ventricular system or subarachnoid space [3].

In most patient with cerebello-pontine angle cysts, the symptoms are subjective (tinnitus, headache and facial paresthesia) and functional. In epidermoids of

parasellar region, patients usually presents with diplopia and seizures. Similar presentation may occur in cysts of suprasellar region. Posterior fossa epidermoids usually presents with seizures whereas headache and gait disturbances are common in fourth ventricle cysts [6]. Fortunately nothing of these happened in our case. CT scan brain shows non-enhancing hypodense lesion. MRI is little more informative and its interpretation depends on presence of lipid, cholesterol and keratin. Usually, this lesions display hypointensity on T1-weighted images, with no gadolinium enhancement. T2-weighted images show a nonhomogeneous high signal intensity lesion. This kind of images can best define the full extent of the lesion, demonstrating any associated oedema. Epidermoid cysts are constituted by a thin capsule with fine internal strands, which may surround rather than displace neuro-vascular structures [2, 7]. In our case, neither CT scan nor MRI was much informative.

Neoplastic transformation of epidermoid cyst is well documented but a rare event. Most common malignancy that arises is squamous cell carcinoma [8]. Cannon *et al.* reported one rare case of primary intracerebral osteosarcoma arising within epidermoid cyst [9].

Differential diagnosis of Rathke cleft cyst with extensive squamous metaplasia and cystic craniopharyngioma may come in epidermoid cyst arising in the suprasellar region. The former usually harbour scattered mucicarmophilic cells of cuboidal or low columnar epithelium atop their squamous island and there is no evidence of the advanced keratinisation typical of epidermoid cyst. The formation of cytoplasmic keratohyaline granules, typical of the epidermoid cyst's maturing squames is absent in craniopharyngioma. However we could not indulge in such types of differential diagnosis due to location of the cyst in our case.

Treatment modality in case of intracranial epidermoid cyst is only surgical excision. There is no role of radiotherapy or chemotherapy in these cases. Extent and approach of surgical removal depends on the size as well as location of the lesion. To conclude, intracranial epidermoid cyst is not so common occurrence and may be asymptomatic or may present with variety of symptoms related to its location. Radiologically it may give an impression of intracranial mass lesion. But histopathological examination will suffice to prove its benign nature and relieve patient's anxiety.

KEY MESSAGES

Intracranial epidermoidcyst is a rare benign cystic lesion of central nervous system. But it may give rise to a variety of symptoms owing to its location and may result in patient anxiety. Proper clinicoradiological

evaluation and judicious histopathological examination can give exact diagnosis and help in proper management of the patient.

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