Case Report

A rare case of intracranial dermoid cyst in a young woman – A case report

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Abstract

Intracranial dermoid cysts generally occurring along the midline are rare. They are benign, congenital, slow-growing cystic lesions located inside the skull. They account for<1% of all primary intracranial tumors and are more common in females in the first three decades of life. Many intracranial dermoid cysts are asymptomatic and are found by chance when brain imaging is carried out for other reasons. Clinical presentation usually relates to compression of adjacent structures or spontaneous rupture of the cyst. The signs and symptoms may range from headaches, seizures to cerebral ischemia. On CT imaging these lesions usually appear as well-defined lobulated midline masses with low attenuation and hyperintense on T1-weighted MRI imaging. We hereby report a case of an 18-year-old female presented with history of seizures involving right upper limb which spread to other limbs associated with frothing and tongue bite- 3 episodes since 2 months. She also had cleft lip. On examination CT images showed hypodense lesion in intrahemispheric region in frontal lobe and MRI image with contrast showed hyperintense T2W, hypointense T1W/FLAIR lesion. A diagnosis of an intracranial dermoid cyst in the intrahemispheric region of frontal lobe was made and the patient was advised a surgical excision of the cyst.

Key words

Intracranial Dermoid Cyst, Hyperintense lesion, Seizures.

Introduction

Intracranial dermoid cysts are slow growing tumor-like malformations arising from the trapped ectodermal tissue during closure of the neural tube. Cysts are lined with stratified squamous epithelium and contain keratin, cellular debris and elements of dermis, including hair and sebaceous glands [1].

A common misconception is that dermoid cysts contain adipose tissue. This is not the case, as lipocytes are mesodermal in origin, and dermoid cysts by definition are purely ectodermal. A dermoid cyst with adipose tissue would be a teratoma [2].

Embryologically, they are derived from two germ cell layers: the ectoderm and the mesoderm. Dermoid cysts typically appear along the midline plane of the human body, corresponding to sites of embryonic fusion [3]. They make up 0.025% to 0.04% of all intracranial tumors, and are seen in 1 in 2500 live births [4].

Clinical features

Intracranial dermoid cysts can have varying clinical features depending on their location, size, and pressure on surrounding structures. Common symptoms include: 1) headaches, 2) seizures, 3) visual disturbances, 4) cranial nerve dysfunction, 5) hydrocephalus, 6) cognitive decline.

Large cysts in the frontal lobes can cause cognitive decline, personality changes, and behavioral abnormalities. It's important to note that some cysts may be asymptomatic and discovered incidentally on brain imaging.

Average time between the onset of symptoms and diagnosis of dermoid cysts has been reported at 8.5 years the previous century, allowing copious time for a variety of clinical symptoms to appear [5].

Diagnosis

Both CT and MRI scans are considered to be the diagnostic methods of choice for dermoid cysts. Radiological features associated with dermoid cysts include the characteristic signal intensities, absence of perilesional edema, and the presence of well-defined margins to the cyst [5]. Dermoids typically appear as hypodense areas on CT. On MRI dermoids are usually hyperintense on both T1- and T2- weighted images [1]. Dermoid cysts when unruptured show typical radiological features and knowing these features are necessary for their diagnosis (**Table - 1**).

Table - 1: Radiological features of an unruptured dermoid cyst [6].

Modality		**Radiological features**
Magnetic Resonance Imaging	**T1**	Typically high signal intensity with chemical shift artefacts
	T2/PD	Variable signal intensity ranging from low to high
	FLAIR	Heterogeneously Hyperintense
	T2*GRE/SWI	Hypointense with “blooming artefacts”
	CE-T1	Commonly do not enhance
Computed Tomography		Hypodense lesion due to fat component commonly do not enhance

T1-T1-weighted images, T2 – T2-weighted images, PD- proton density-weighted images, T2*GRE – T2*-weighted gradient echo images, SWI – Susceptibility-weighted images, CE-T1 – contrast-enhanced T1-weighted images

Differential Diagnosis

There are other intracranial lesions that can have similar clinical features or radiological findings as intracranial dermoid cysts. These include epidermoid cysts, arachnoid cysts, craniopharyngioma, pituitary adenoma, meningioma, and metastatic tumors. Epidermoid cysts lack the characteristic oily or cheesy

contents seen in dermoid cysts. Arachnoid cysts do not contain hair or sebaceous material. Craniopharyngiomas have a different appearance on imaging studies and do not contain sebaceous or hair material. Pituitary adenomas can mimic the appearance of intracranial dermoid cysts on imaging studies, but do not contain sebaceous or hair material. Meningioma's have a different appearance on imaging studies and do not contain sebaceous or hair material. Metastatic tumors are usually multiple and have a different clinical presentation compared to intracranial dermoid cysts. A biopsy or surgical resection of the lesion may be necessary to confirm the diagnosis of an intracranial dermoid cyst.

Treatment and prognosis

Surgical excision is the treatment of choice if symptomatic. However, complete resection is difficult as not all tissue can be removed, especially from around cranial nerves and vessels. Recurrence is therefore not uncommon, although growth is typically slow and many years can elapse without new symptoms.

Case report

An 18-year-old female patient presented with history of seizures involving right upper limb which spread to other limbs associated with frothing and tongue bite- 3 episodes 2 months ago.

History of presenting illness: Patient was apparently asymptomatic 2 months ago when she had 3 episodes of seizures involving right upper limb associated with frothing and tongue bite. No history of fever, vomiting, headache, loss of consciousness, blurring of vision and diplopia.

Past history: No history of similar complaints in the past, No history of HTN/ DM/ CAD/ CVA/ TIA/ CRHD in the past.

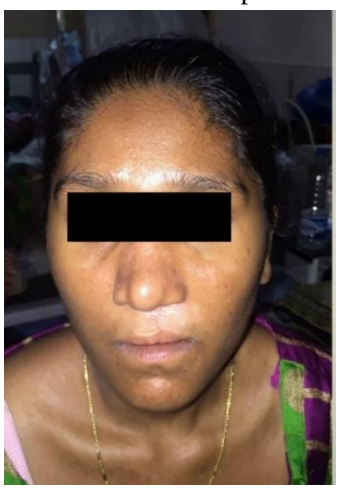
Personal history: Mixed diet, Normal appetite, Adequate sleep, Bowel and bladder movements regular, No addictions, No known drug allergies.

Family History: Not significant.

General Examination: Patient was conscious, coherent and cooperative. There was no pallor/ icterus/ cyanosis/ jaundice/ clubbing/ pedal edema or lymphadenopathy.

On examination a cleft lip was present (**Figure - 1**).

Figure - 1: Patient with cleft lip.



Vitals: Temp: 97.3F PR-92/min, regular, normal volume, equal on both sides and no radio-femoral delay. RR:18 cpm, BP – Right Upper Limb 100/70 mm Hg.

Systemic examination:

CNS - Intellectual functions: - Patient was conscious/ coherent/ co-operative, oriented to time, place and person. Memory, recent and remote Intact. Speech – Normal, Cranial nerves - all cranial were nerves normal. Motor system, sensory system, cerebellar system were normal. Skull and scalp – normal, No neck stiffness, pupils - NSRL, plantar- Normal. Gait - Normal, Peripheral nerves – not thickened.

CVS: S1, S2 +, No murmurs heard.

Lungs: B/L air entry +, No additional sounds.

PA: Soft, no hepatosplenomegaly

Investigations:

CBP: Normal,ESR–20 mm, RBS: 95 mg, Blood urea: 34 mg, serum creatinine: 0.8 mg, Serum electrolytes - Na+ -140 meq/ l, K+ - 3.4 meq/l, Cl- 106 meq/l. ECG - Normal. CXR- Normal.

CT brain: Hypodense lesion noted in interhemispheric region in frontal lobe.

MRI brain with contrast (**Figure - 2A and 2B**): Hyperintense T2W, Hypointense T1W/FLAIR lesion, with restriction of DWI and no post contrast enhancement along midline interhemispheric fissure causing indentation of anterior body of corpus callosum.

Figure - 2A and 2B: MRI Brain: Segittal (A) and Axial (B) T2 weighted images demonstrating hyperintense midline lesion in the interhemispheric fronto-parietal region.

Figure – 2A:

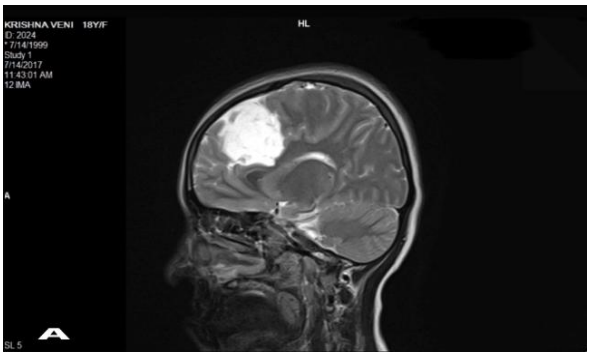
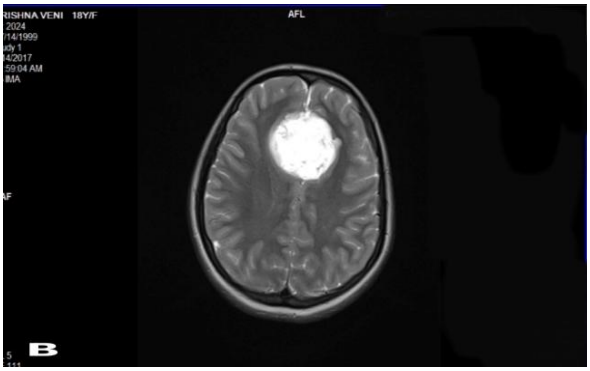


Figure – 2B:

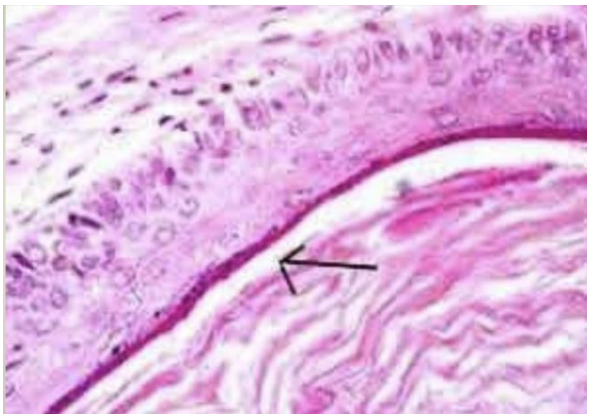


S/O Dermoid Cyst in Brain.
Histopathological Examination (**Figure – 3**): Fibrous wall lined by keratinizing squamous epithelium with skin adnexa. Cyst contains squamous epithelium, hair, sebum.

Diagnosis: Intracranial dermoid cystin the interhemispheric region of frontal lobe.

Treatment: Surgical excision of the cyst done.

Figure - 3: H and E stained tissue from the cyst wall showing fibrous wall lined by keratinizing squamous epithelium with skin adnexa.



Discussion

Background

Intracranial dermoid and epidermoid cysts have not been acknowledged until E. Bostroem first recognized in 1897 the epithelial origin of the "tumeur perlees" that was described by Cruveilhier in 1829 [5]. Ever since these discoveries, several reports of dermoid and epidermoid cysts have been published [7].

Characteristic presentations

Dermoid cysts are thick walled cysts lined by keratinized squamous epithelium and constitute 0.1% to 0.2% of all intracranial tumors. They arise from the inclusion of ectopic embryonic rests of the ectoderm in neural tissue. The cyst material contains skin appendages like hair follicles, hairs, sweat glands, sebaceous glands, teeth or nails [8]. Dermoids are predominantly midline lesions, arising commonly in diploe of the fontanel extradurally and in parasellar region intradurally [1]. Extradural intracranial dermoid cysts, when compared to those located intradurally, are a much rarer entity [5].

Although dermoid tumors develop from the embryonic period, symptoms may not occur until adulthood due to their slow growth [9]. Many intracranial dermoid cysts are asymptomatic and are only found incidentally. Often there is a history of vague symptoms, with headache and seizure being most common [10]. The clinical presentation depends on the location of the cyst.

Posterior fossa dermoid cysts present early where as supratentorial dermoid cysts present in the 3rd to 4th decade of life [9].

Our patient presented at 2nd decade of her life with recurrent seizures. Despite their benign nature, dermoid cysts have a high rate of morbidity and mortality if ruptures [11].

The rupture cause dissemination of the cholesterol debris from the cyst into the subarachnoid and intraventricular spaces, may cause chemical meningitis [12]. However, with the early radiological detection of dermoid cysts, devastating events such as rupture of the cyst are rarely reported [5]. Other complications of dermoid cysts include the development of dermal sinuses, which are channels connecting the dermoid cyst with the skin. Lesions as such most commonly present in the lumbar and occipital regions [13].

Both CT and MRI scans are considered to be the diagnostic methods of choice for dermoid cysts [5]. MRI scan usually shows hyperintense on T1-weighted images lesion with fat content but it sometimes can appear more heterogeneous with minimal enhancement because of the presence of calcifications, hair, epithelial debris, and sebaceous secretion [14]. CT scans can easily depict dermoid cysts as discrete lesions of low density, a characteristic which is consistent with their fatty content. Other imaging modalities, such as plain skull film, may show a well-defined, lucent lesion with ring-like or egg shell calcification, as well as bony defects, if present [5].

Management

Complete surgical resection of dermoid cysts is the only effective treatment for prevention of future recurrence and complications [15]. In some instances, complete resection might be difficult due to the cyst's extensive fibrous adhesions into the adjacent neurovascular structures [16]. Despite this, total resection of the cyst along with the adhesions and sinuses is crucial to avoid recurrences [5].

Conclusion

Intracranial dermoid cysts are rare, slow-growing tumors made up of embryonic tissue. They are generally benign, but can cause issues due to their size and location. The symptoms can range from non-life threatening such as headaches to morbid conditions such as cerebral ischemia and death. Although death is a rare case it causes recurrent seizures and infective meningitis if the cyst ruptures. Treatment usually involves surgical removal and may include radiation therapy in certain cases. The prognosis is usually good with low rates of recurrence after successful removal. However, complications such as infection, bleeding or damage to surrounding brain tissue can occur. A multi-disciplinary team should manage the cyst with careful consideration of the patient's symptoms, medical history and imaging findings.

Here we report a rare case of intracranial dermoid cyst in a young woman who had recurrent episodes of seizures of 2 months duration. Radiological examination by MRI of brain shows a Hyperintense interhemispheric midline lesion of the fronto-parietal region. Radical Surgical excision of the cyst is done. She recovered completely and there are no further seizures.

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