

Association of Occipital Dermal Sinus with Intracerebellar Epidermoid and Abscesses

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Abstract

Background: Congenital occipital dermal sinus with an underlying dermoid is a rare, benign lesion of embryological origin and may occur anywhere along the neuraxis. **Material and methods:** A case of occipital region dermal sinus associated with cerebellar abscess is described. **Result:** A-10- year-old girl was admitted with complaints of fever, headache and vomiting, Neurological examination revealed stiff neck with papilloedema involving both eyes. She had ataxic gaits with cerebellar signs. The computerised tomography revealed multiple cerebellar abscesses. In emergency, she underwent midline suboccipital craniectomy; with excision of abscess and developed aseptic meningitis in the postoperative period, which responded to antimicrobial therapy. **Conclusion:** A case suspected with meningitis should all be looked for presence of sinus along the spinal and cranial axis, which may give clue to presence of the underlying dermoid cyst and abscess. The treating physician should aware of such although rare clinical entity as timely surgical intervention can yield good neurological outcome.

Keywords

Epidermoid Cyst, Occipital Dermal Sinus, Cerebellar Abscess

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1. Introduction

Dermoid ‘tumor’ does not represents a true neoplasia but considered as inclusion cysts derived from ectodermal elements. [1-7] It is uncommon lesions, [9-12] constituting about 0.03-0.6% of total intracranial tumors [11–12]. It is postulated to originate as a result of incomplete disjunction’ during early period of gestation and, the residual tracts are associated with an inclusion tumor in 60-80% of the cases. [13]. Histopathologically dermoid cysts is well delineated lesion, containing lining of stratified squamous epithelium and show typical slow enlargement and progressive accumulation of thick, yellow- greenish brown material consisting of collection of desquamated epithelium linings, secretions of sebaceous gland, fat, and whorls of hair. [14, 15] However, presence of hair follicles, sweat glands and sebaceous gland in the lining cyst wall of dermoid cysts differentiates from common epidermoid cysts. Dermal

sinuses are abnormal communication between skin and deeper tissue.

Wright reviewed a total of 127 cases and observed sinuses in the three fourth cases were located along spine and only one third located along the cranium. [16] Dermoid cysts associated with dermal sinus tract can extend upto dura or superficial to it in 18% of the cases and in the rest of cases can further extend even to subdural or even deeper planes in the cranial cavity. [16]

Typically dermoid cysts represents as heterogeneous mass and presenting as hypodense lesion on plain computed tomography head scan and Hounsfield unit may be negative due to fatty content of cyst and frequently possess a thin rim of calcification. On MRI study cyst appear as hyperintense signal on T1W image and which becoming typically hypointense on T2W imaging. in addition curvilinear hypointense elements presence may be indicator of the presence of hair as the content of the dermoid cyst. [15]

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The occipital dermal sinus can extend up to variable depths and can extend either up to subcutaneous tissue, bone, extradural space or subdural space, cerebellar parenchyma or even rarely to the fourth ventricle. It can end either blindly or more commonly communicate with dermoid or epidermoid cyst. Authors are reporting a case of occipital dermal sinus associated with posterior fossa intracerebellar epidermoid in a school girl, which was associated multiple intracerebellar abscesses, the clinical feature, radiological and management is discussed.

2. Case Report

A-10- year-old girl was admitted in neurosurgical ward with progressive complaint of headache, fever, vomiting, and photophobia. Neurological examination revealed stiff neck with papilloedema involving both eyes. She had ataxic gaits. Bilateral cerebellar signs were present. An emergency cranial computerised tomography scan was carried out, which revealed three cerebellar abscesses, two in the vermis and one in the cerebellar hemisphere and another hypodense lesion in upper vermis. (Fig-1) with hyperdense capsule, showing contrast enhancement. Routine haematological test revealed leucocytosis. A dermal sinus was observed below the external occipital protuberance. An emergency surgery was carried out, with midline suboccipital craniectomy; attempt was made excise three abscesses. Five days after, she was taken up for re-surgery to excise the epidermoid. The cyst was adherent to the lower surface of tentorium, where the dermal sinus was ending into the epidermoid cyst. She had aseptic meningitis in the postoperative period, which responded to antimicrobial therapy.



Fig. 1. Non-contrast computed tomography scan of head revealing posterior fossa abscess in a 10-year girl with history of discharging dermal sinus in the occipital region.

3. Discussion

Dermal sinuses are ectodermal closure defect. It may extend into various depths, through the tissues of mesenchymal origin, skin, subcutaneous tissue, bone, or dura mater. These defects are usually produced before 3-5 weeks of intra-uterine life. The occipital dermal sinus with posterior fossa extension may be produced by failure of anterior neuropore closure defect towards the end of fourth week of gestation. The commonest site of dermal sinus is lumbosacral region; these are rare in the occipital region. [4] They are covered by stratified squamous epithelium with hair follicles. The sinus commonly ends in dermoid or epidermoid tumor. Although commonly located in the midline, the mal-development of the optic and otic vesicle shows the presence of dermoid in the midline but epidermoid off the midline in the cerebello pontine angle, parahypophyseal cistern, and trigone of lateral ventricle. [5]

An early defect with few differentiated cell produce dermoid cyst but if defect occurs relatively later in the embryonic life, with well-differentiated cell give raise to epidermoid cyst. [5-7, 9] Giuffre and Curatolo [2] reported after review of 46 cases in literature noticed intracerebellar and subdural extension was noted in 50% and 47% respectively, another 3 % cases ,the sinus tract were ending in extradural location. Matson also observed 18 cases with occipital dermal sinus noticed track ending in subdural (44%), intracerebellar (38%), in the 4 th ventricle (12%) and only 9% in extradural location. [10] Schijman et al [9] observed in study of their 7 cases, all ended in posterior fossa cyst epidermoid in 5 cases and dermoid in another two cases. [8, 9]

Mostly dermal sinus with associated dermoid or epidermoid presents with repeated episodes of meningitis, feature of mass effects or cerebella abscess. Subgaleal collection with spontaneous drainage of purulent material. However, most frequently a combination of symptoms is observed. The meningitis can be caused by *Staphylococcus aureus*,¹⁰ other organism incriminated are *Klebsiella*, *proteus* [9]

The diagnosis of these lesions in associated with dermal sinus is relatively easy. The skull x-ray may show bone defect usually situated at lithe level of the inion. [9] The CT scan may show the dermal sinus and associated dermoid and epidermoid cyst. These lesion needs differentiation from other commonly occurring neoplasm and abscesses of the posterior cranial fossa. These are hypodense with well-defined hyperdense margins, with slight enhancement following contrast administration. This enhancement is related to capsule and frequently inflamed surroundings. It is noted that the density of epidermoid cyst can vary between -22 to 18 HU, while dermoid cyst have a density between +4 to +18 HU. [5]

However, dermoid can demonstrate occasional atypical neuroimaging features. Satyarthee and Mahapatra reported compete hyperdense dermoid cyst revealed on CT scan imaging, located in the sellar and suprasellar region, which was operated and histopathology was consistent with dermoid cyst. [17] Dermoid located in the spine can show typical, however possibility of variable imaging findings should always be kept in mind. [18]

Regarding ideal management of associated epidermoid cyst with abscess, initially abscess removal should be carried out and after a gap of few days total en block removal of dermal sinus, its cutaneous orifice and epidermoid should be excised. However, complete excision of epidermoid and abscess can be done in one sitting. During surgery special care must be given to avoid spilling of cyst content because spilling of cyst content can lead to seedling of epithelial cells and secondary aseptic meningitis caused by cholesterol and fatty acid. However, total excision of lesion is frequently difficult owing to close adherence between cyst capsule and, more frequently the dermal sinus with torcula Herophili. So care is needed for careful dissection and preservation of torcula, which may be endanger if proper precaution is not taken. However, in our case also torcula was intact after complete excision of dermal sinus, its orifice and associated epidermoid cyst.

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