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Occipital Dermal Sinus Tract Causing Craniospinal Infection: Case Report and Review of Literature

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Abstract **Background:** Congenital dermal sinus tract (DST) is a rare entity of cranial or spinal dysraphism that may occur anywhere from nasion and along the midline neuraxis from occipital to sacral regions. Craniospinal infection caused by occipital DST is even rarer. Because of their scarcity, these lesions are not well documented in the literature, often mixed with dermal sinuses in other location or other dysmorphic features. This paper reports a unique case of an infant presented with craniospinal abscesses resulting from occipital dermal sinus tract. **Methods and materials**: In this paper, we report a case of a 16-month-old girl presented with high grade fever, vomiting and lethargy. She had a discharging occipital skin lesion. Her diagnosis was "Occipital DST with Cerebellospinal Abscess", which was treated successfully by excision of the DST and cerebellar abscess. Histopathological examination revealed a dermoid cyst. She received 8 weeks of parenteral antibiotic treatment with a good outcome. **Results**: Occipital DST is a rare condition. Its clinical presentation varies and clinical suspicion is required. Early neurosurgical intervention is important to prevent the risk of potential complications such as abscess and bacterial meningitis. **Conclusion**: This case highlights the importance of early recognition and evaluation of midline craniospinal cutaneous stigmata in infant. Further neurosurgical assessment with radiological investigations are recommended for early detection and management. Once diagnosed is made, surgical intervention and appropriate antibiotic therapy are the mainstay of treatment.

Keywords: *occipital dermal sinus tract, abscess, dermoid, meningitis*

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1. Introduction

Congenital dermal sinus tract is an epithelium-lined tract that extends from an opening in the skin through deeper tissues [1]. It is a form of craniospinal dysraphism that may take place during neurulation when the neural groove closes to form the neural tube on Day 26 of gestation [2]. Dermal sinus tract was first described by Ogle in 1865 [3]. Spinal DST with staphylococcal meningitis was reported by Moise in 1926 in a young boy [4].

Dermal sinus tracts may occur anywhere along the craniospinal axis and the majority of these lesions occur in the lumbar (41%) or lumbosacral region (35%) followed by the occipital and thoracic regions (10%) and cervical (1%) respectively [2,3]. They can be found in association with other pathologies, such as inclusion tumours (e.g., dermoid, epidermoid, teratoma), split cord malformations and tethered cords [4].

We report an uncommon, but successfully treated case of infected occipital DST with cerebellospinal abscess and meningitis. This case highlights the importance of early recognition and evaluation of midline craniospinal axis in children with meningitis and DST.

2. Case Report

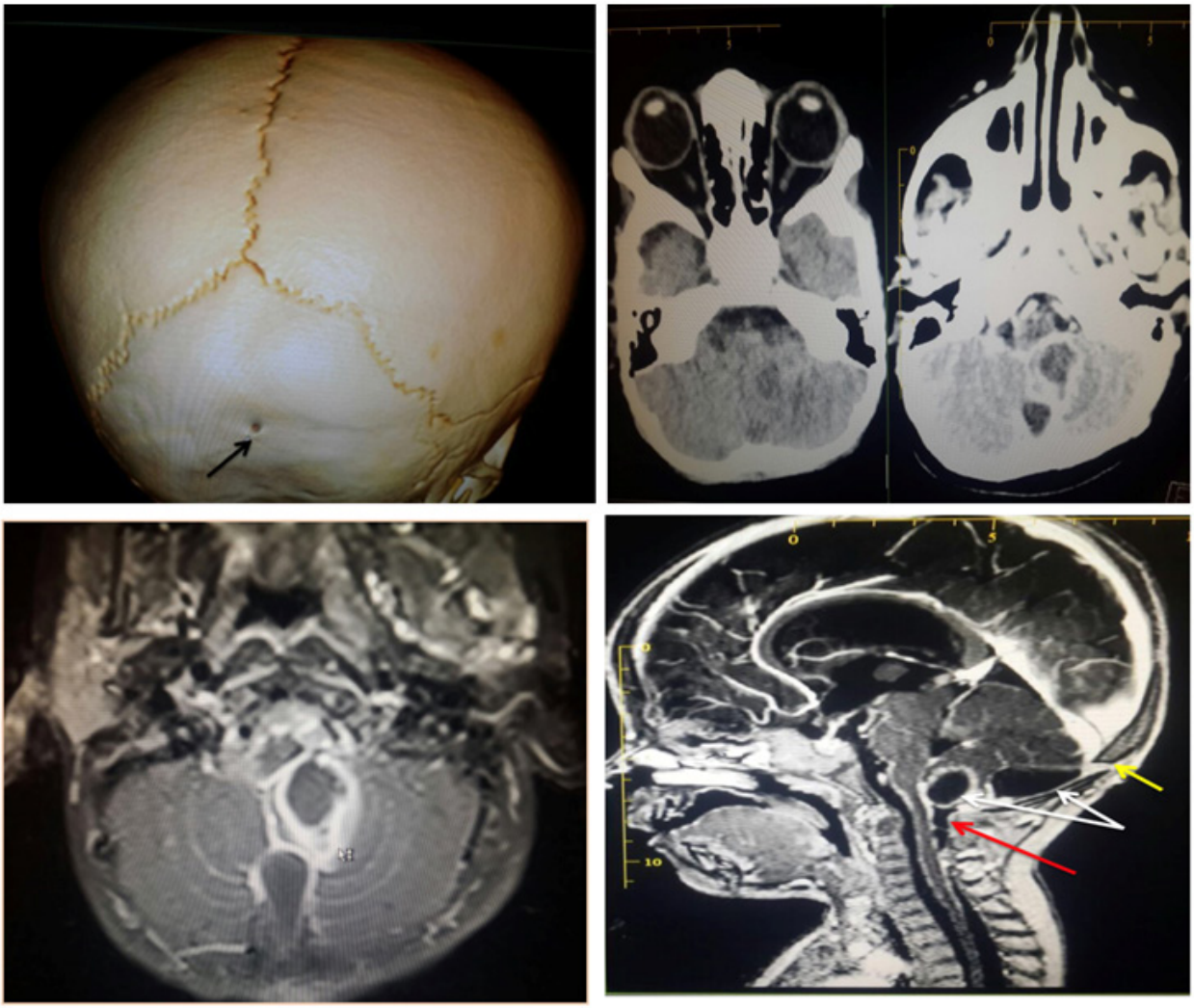
A 16-month-old female was born full term with normal Apgar score and neurological assessment. The case was referred to our hospital with 3 days history of fever, vomiting, lethargy, irritability and poor feeding. There was no history of convulsions. According to the mother, child has a skin lesion in the occipital region that was present since birth and is discharging intermittently.

Earlier, she had been admitted in another hospital because of meningitis. During that admission, her laboratory studies were unremarkable except for high white cell count in blood with neutrophilia. Parents refused lumbar puncture then, head CT was done and reported as normal. Ceftriaxone, vancomycin and acyclovir were empirically prescribed for 14 days. There was clinical improvement so patient was discharged home. Two weeks later patient condition deteriorated. She was re-admitted in another

second hospital and was found to be febrile and tachycardiac. Her head circumference was 47 (at 50th centile). Lumbar puncture was done and revealed WBC 270 cells/mm^3 (0-20/mm^3), protein 2.59 mg/dL (15–60 mg/dL), glucose 3.5 mmol/L (2.5 - 4.4 mmol/L), Concomitantly, blood glucose was 6.2 mmol/L. Cerebrospinal fluid (CSF) microscopy and culture were unremarkable. Head CT done and showed multiple hypodense areas in the left cerebellum with rim enhancement. There was an extradural cystic lesion that communicates through a sinus to the occipital bone. There was soft tissue seen in the occipital area adjacent to the sinus (Fig. 1A-B-C). The findings were representing an infected dermoid cyst with secondary cerebrospinal abscess collection. Patient was then transferred to our service for MRI and further evaluation and management.

On admission to our hospital, general examination revealed a small midline occipital skin lesion. There was no discharge and no signs of inflammations. Anterior fontanelle was lax and nuchal rigidity was not present. C-reactive protein was elevated 96 mg/L (<5 mg/L) as was the number of WBC 13,000 cells/mm^2.

Both contrast and non-contrast MRI were done (Figure 1D-E). Non-contrast MRI revealed a well-defined rounded lesion measuring 1.3cm seen in the left cerebellum. The lesion was associated with moderate degree of perilesional edema with compression of the brainstem and forth ventricle resulting in obstructive hydrocephalus. Contrast MRI showed diffuse enhancement of the leptomeninges and enhancing lesion in the left cerebellum and upper cervical spine (Figure 1E). The findings were suggestive of cerebellospinal abscess and diffuse meningitis.

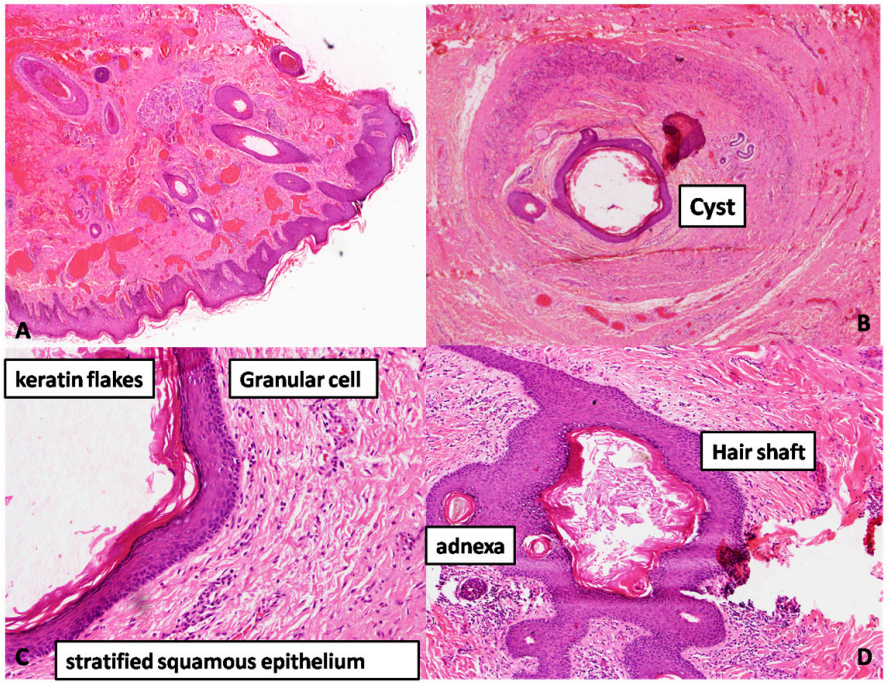


a. 3D reconstruction CT head. Black arrow showing the bony defect of the sinus tract in the mid occipital bone.
b. Plain and contrast CT head showing a ring enhancing left cerebellar abscess
c. MRI brain; Axial T1 with contrast showing multiple enhancing lower cerebellar abscesses.
d. MRI brain: Sagittal T1 with contrast showing multi-loculated cerebellar abscesses (white arrows) extending to the upper posterior cervical spinal canal (red arrow). The enhancing bony sinus tract is also shown (yellow arrow). There is also evidence of lepto-meningeal enhancement.

Figure 1.

Patient underwent sub-occipital craniotomy and excision of dermoid sinus and cerebellar abscess. Intra-operatively pus came out along with hair and cheesy materials. There was a dermoid which was excised but surgery could not be continued as patient had hemodynamic instability.

Histopathological examination of the specimen revealed fragments of hair bearing skin and underlying subcutaneous tissue. Dermoid cyst is lined by stratified squamous epithelium and containing keratinous material was seen. The wall contain pilosebaceous units (Figure 2).



a. Section shows skin and subcutaneous tissue
b. A cyst is noted in the dermis of skin
c. The cyst is lined by stratified squamous epithelium, Granular cell layer is present, Cyst lumen contains keratin flakes
d. Cyst wall also has adnexa and lumen also has hair shaft

Figure 2.

Sagital T1 post contrast done one year after the surgery showing the complete resolution of the located abscess with mild glottis changes in the lower cerebellum.

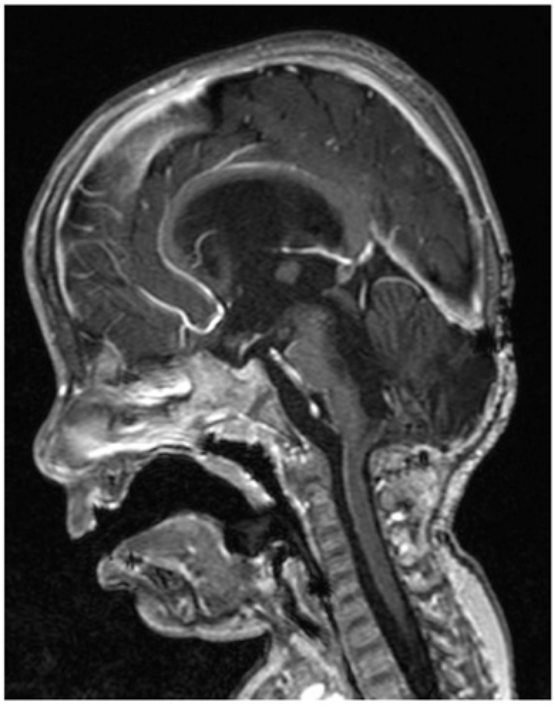


Figure 3.

Postoperatively, the patient was placed on vancomycin, ceftriaxone and metronidazole. When final culture sensitivity showed presence of staphylococcus aureus, cloxacillin was added instead of vancomycin. The antibiotics were given for 8 weeks.

Patient had repeated vomiting and became drowsy. Hence, head CT scan done and showed significant dilatation of lateral ventricles. Urgent programmable VP shunt was inserted. Follow-up examination at 1, 4, 9, 12 months revealed normal neurological functions. Figure 3 showing Sagittal T1 post contrast done one year after the surgery showing the complete resolution of the located abscess with mild glottis changes in the lower cerebellum.

4. Discussion

A congenital DST is a rare entity of craniospinal dysraphism which represents a cutaneous depression or tract that communicates between the surface of the skin and deeper structures [2]. Different theories exist regarding the embryology of dermal sinuses. It was thought to be the result of failure of the neuroectoderm to separate from the overlying cutaneous ectoderm at the end of neurulation [5].

Approximately 60% of congenital DSTs end in a dermoid or epidermoid tumours, since ectodermal, or mixed ectodermal and mesodermal elements can be trapped along the tract [6].

In the literature, nine cases of occipital dermal sinuses were described (Table 1) [7].

Posterior fossa dermoid cysts were classified into four groups depending on whether they were extradural or intradural and on the degree of development of the dermal sinus, whether absent, partial or complete: (1) extradural dermoid cyst with a complete sinus, (2) intradural dermoid cyst without a dermal sinus, (3) an intradural dermoid cyst with an incomplete dermal sinus, and (4) intradural dermoid with a complete dermal sinus [7].

Table 1. Summary of the nine Cases of Occipital Dermal Sinus with Extra Dural Dermoid Cyst Published in the Literature

Serial number	Authors/ Year	Age/Sex	Clinical presentation	Imaging	Cranial and spinal involved	Organism	Localization	HPE
1	Martin/1943 [17]	2y/M	Raised ICP	Plain radiographs	Cranial only	-	Interdural	Dermoid and epidermoid tumors
2	Logue/1952 [18]	2y/M	Occipital scalp swelling	Plain radiographs, ventriculography	Cranial only	Staph. Aureus, Bact. coli.	Epidural	Posterior fossa dermoid cysts
3	Maston/1969 [19]	2.5y/F	Meningitis, Occipital scalp swelling	Plain radiographs	Cranial only	Staph. Aureus	Epidural	-
4	Schijman/1986 [20]	3y/F	Meningitis, Occipital scalp swelling	CT scan	Cranial only	Staph. Aureus	Interdural	Epidermoid cyst
5	Rubin/1989 [21]	27y/M	Raised ICP	CT scan	Cranial only	Staph. Aureus	Epidural	Torcular compression
6	Martinez/1992 [22]	2y/F	Occipital scalp swelling, local infection	CT scan	Cranial only	-	Epidural	Empyema
7	Goffin/1993 [23]	7y/M	Occipital scalp swelling	CT scan, fistulography	Cranial only	-	Interdural	Cerebellar abscesses + dermoid cyst
8	Martinez/1997 [24]	6m/F	Occipital scalp swelling	MRI	Cranial only	-	Epidural	Sinus hemorrhage
9	Martinez/1997 [24]	2y/F	Occipital scalp swelling	MRI	Cranial only	-	Interdural	Sinus hemorrhage
8	Akhaddar/2001 [7]	14m/M	Acute raised ICP, seizure, meningitis	CT scan	Cranial only	Staph. Aureus	Epidural	Cerebellar abscesses
9	Gurdeep/2009 [6]	20m/F	Meningitis	CT scan /MRI	Cranial only	Propionibacterium avidum, peptrostreptococcus and enterococcus faecalis	Extradural	Multiple cerebellar abscesses + dermoid cyst arising from the 4th ventricle
10	Our case	16m/F	Discharging Occipital scalp swelling Meningitis, craniospinal abscess	MRI	Craniospinal	Staph. Aureus	Intradural	Cerebellospinal abscesses + dermoid cyst

ICP: intracranial pressure _ M/F: Male/Female _ y: years _ m: months _ HPE: Histopathology examination.

Cutaneous markers such as skin dimple, nevi, hemangioma, pigment changes, subcutaneous lipoma and others can be found in 86% of occult spinal dysraphism [8]. Tethered cord, bifid lamina, split cord malformation, inclusion tumors such as epidermoid and dermoid cysts, tight filum terminalis and myelomeningocele can be seen together with dermal sinus tracts [3].

A history of clear fluid discharge with intermittent swelling or recurrent episodes of infection are confirmatory features of DST [9]. As found in our patient, the presence of a skin dimple with intermittent discharge in the occipital area should always be subject to investigations. Dermal sinus may serve as a tract that allows seeding of bacteria to the spine and cerebellum [10]. These lesions predispose to intraspinal dermoid formation and infective complications such as spinal abscess or meningitis [9]. Repeated meningitis of unknown origin due to infected dermal sinus tract was also reported [4]. Rarely, dermoid cysts may present with raised intracranial pressure or seizures [7].

Dermoid cyst may contain stratified squamous epithelium, hair follicles and keratin debris [6]. Absence of sebaceous and sweat glands indicate that the dermoid cyst may have contain less lipid or liquefied cholesterol [1,11].

Staphylococcus aureus is the most common responsible organism for infected DSTs [2], as found in our case. E coli, propionibacterium avidum, peptrostreptococcus and enterococcus faecalis were also reported [2,6].

Beside the clinical examination, occipital or spinal DSTs can be diagnosed by different modalities of imaging, such as plain radiographs, sonography, computed tomography (CT) and magnetic resonance imaging (MRI). Plain x-ray and CT may not show abnormal findings [4]. However, plain radiograph is a good modality in cases where DST is associated with dermoid tumour as it can differentiate it from lipoma by a well-defined hypodensity with occasional calcifications [5]. Spinal sonography can detect DSTs, position of conus, and other associated spinal anomalies in newborns and small infants [12]. It is an excellent initial tool for screening spinal dysraphisms and early use of sonography allows early surgical interventions which in turn, prevent complications associated with DSTs [4,13]. CT scan is the best modality in detecting the associated bony tract, and it also can reveal the exact location of the dermoid cyst, its relationship with the adjacent sinus and any related cranial defect [5]. Unless infected, the cyst does not enhance with contrast [5]. Computed tomogram also defines the complications of abscess formation and hydrocephalus [5]. Contrast enhancement of the associated posterior fossa abscess of an infected dermoid cyst is seen in both CT and MRI [5]. Dermoid typically show high T1 and T2 signals and when infected, have restricted diffusion on DWI and ADC imaging [5].

MRI is the imaging modality in diagnosis and assessment of both DST and associated dermoid cyst [5]. MRI could help in determining the surgical approach by delineation of the sinus tract, its extension into deeper tissues and its association with cysts, abscesses or venous anomalies [14]. The MR characteristics of dermoid cyst are hypointense on T1-weighted imaging, hyperintense on T2-weighted imaging and peripheral enhancement with gadolinium administration [15].

Early diagnosis and prompt surgical intervention with appropriate antibiotic therapy offer the best chance for functional neurological recovery and decrease the risk of complications.

Our patient received systemic antibiotics for 8 weeks according to bacterial sensitivity, other authors recommend the use of antibiotics for four weeks in case of cerebellar abscess caused by infected DST [5,16].

Craniospinal abscess may recur and require other operations. Therefore, it is recommended that regular follow-up by clinical and radiological examinations should be done.

In conclusion, craniospinal abscess due to occipital DST is a rare condition and high index of suspicion is required for diagnosis. Cutaneous markers have critical role in detecting occult craniospinal dysraphism and require further investigations. The development of craniospinal abscess by contamination through the occipital DST indicates the importance of early excision of tract. Once diagnosis of craniospinal abscess is made, prompt surgical excision of DST, drainage of abscess and appropriate antibiotics are the mainstays of the treatment.

Abbreviations

DST: dermal sinus tract, MRI: Magnetic resonance imaging, CSF: Cerebrospinal fluid.

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