Case Report

Late onset Proteus Mirabilis Meningitis and Subdural Abscess in a Boy With Lumbosacral Epidermoid Tumor: Case Report

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Abstract
Dermal sinus tracts have been reported all along the midline neuroaxis, with the majority being located in lumbar and sacral areas. They are usually associated with split cord malformations, tethered cord and inclusion tumors (dermoid, epidermoid, teratoma). Dermal sinus tracts with concomitant dermoid or epidermoid tumors are frequently associated with recurrent meningitis, but spinal subdural abscess is rare. The authors report a 2 years old boy with lumbar epidermoid tumor, late onset Proteus mirabilis meningitis and spinal intradural abscess, who had a history of a dermal sinus tract removal one year before his admission. The importance of early diagnosis, proper surgical and medical treatment and close follow up in such patients is stressed.

Keywords: Dermal sinus tract, epidermoid tumor, meningitis, pediatric, Proteus mirabilis, subdural abscess

INTRODUCTION
Congenital spinal dermal sinus is a stratified squamous epithelium-lined tract extending from the skin overlying the spine inward to varying depths. The sinus may terminate on or near neural structures and often present in childhood with skin findings, progressive weakness or sensory loss in the lower extremities, gait abnormalities, bowel and bladder dysfunction and/or infection¹. Dermal sinus tracts (DSTs) have been reported all along the midline neuroaxis, with the
majority being located in lumbar and sacral areas\(^{(1,11)}\). They are usually associated with other pathologies, such as split cord malformations, tethered cord and inclusion tumors (dermoid, epidermoid, teratoma)\(^{(1)}\).

Intraspinal epidermoid tumors are rare intradural extramedullary tumors, representing less than 1% of all intraspinal tumors\(^{(7)}\). They are usually congenital in nature and when compared to dermoid tumors they are rarely associated with a DST\(^{(2)}\). The connection between intraspinal epidermoid cysts and the skin via DST provides direct access for bacteria on the skin to reach the interior cysts which may result in recurrent episodes of bacterial meningitis. However, spread of infection within the subdural space is rare. Spinal subdural abscess with a coexistent epidermoid tumor is even rarer and only few case reports have been published\(^{(2,5)}\). We report a 2 years old boy with lumbar epidermoid tumor complicated with late onset Proteus mirabilis meningitis and spinal intradural abscess, who had a history of dermal sinus tract removal operation one year before his admission. We intend to point out the importance of early diagnosis, proper surgical and medical treatment and close follow up in patients harbouring epidermoid tumors complicated with subdural abscess.

**CASE PRESENTATION**

A 2 years old boy presented to our pediatric emergency clinic with progressive difficulty in walking, deterioration of consciousness, and fever. His parents admitted that these symptoms started one week prior to his admission and progressively worsened. His past medical history was unremarkable except for surgical removal of a sacral dermal sinus tract one year ago in another center (Figure 1). Physical examination revealed a temperature of 39.5°C, heart rate of 110/minute, respiratory rate of 25 /minute, and blood pressure of 100/60 mm Hg. Neurological examination revealed a Glasgow Coma Scale (GCS) score of 14 (M:6, V:5, E:3), stiff neck, positive Brudzinski and Kernig signs and paresis in lower extremities. The hemogram showed a WBC count of 20,000/mm\(^3\) with 75 % neutrophils (Hb: 12 g/dL, Hematocrit:36%). Cranial computerized tomography (CT) revealed no pathological signs. Spinal magnetic resonance imaging (MRI) showed an intradural, septated, peripherally enhancing lesion with heterogeneous density, extending from L3 to S1 (Figure 2a-c). Subdural abscess was considered in the diagnosis and subsequently the patient underwent emergency surgery. After L3 and L4 laminotomy and durotomy, microsurgical removal of the lesion along with the covering whitish, stinky purulent abscess was done. The cavity was irrigated with saline and the dura was closed in a watertight fashion. The histopathological examination was consistent with an epidermoid tumor complicated with an abscess (Figure 3). The cultures of the abscess material demonstrated Proteus Mirabilis. The antibiogram tests showed sensitivity to Meropenem and Teikoplanin, which was started at 300 mg IV daily in 3 doses and 200 mg/d in one dose consequently. At postoperative 7th day, the patient had a generalized tonic clonic seizure, nausea and vomiting. The emergent cranial CT showed hydrocephalus and an external ventricular drainage was applied. Phenobarbital at 4mg/kg/day was started. The patient's general physical condition and neurological symptoms improved rapidly. On the 14th day of ventricular drainage the CSF biochemistry and cultures were in normal limits and the patient's physical and neurological examination revealed no pathological findings. The ventricular drainage is stopped to check if the patient would tolerate the removal of the catheter, however one day later the patient showed symptoms of raised intracranial pressure with hydrocephalus on cranial CT. Subsequently, the patient underwent ventriculoperitoneal shunting. On 7th week
of his admission antibiotic treatment was stopped and the patient was discharged without symptoms and signs. The neurological and radiological examinations were normal in the follow-up period and his last control MRI at postoperative 5th year disclosed no pathological findings (Figure 4).

Figure 1: The thoracolumbar MRI sections prior to surgical removal of the sacral dermal sinus tract were obtained from the patients’ parents after his admission. On T2 weighted sagittal MR images the course of the dermal sinus tract and the intradural mass is clearly seen.

Figure 2: MRI sections showing intradural, septated, peripherally enhancing lesion with heterogeneous density, extending from L3 to S1 (a. T1-weighted sagittal; b. T2-weighted sagittal; Gadolinium enhanced T1-weighted c. axial and d. sagittal sections).
DISCUSSION

Despite their harmless external appearance, DSTs may cause serious clinical problems if they are underestimated and not properly managed. Patients typically present with infection and apart from meningitis, rare but serious conditions such as focal intradural accumulation of pus and abscess can also be encountered. Thus, early diagnosis and treatment of the DST along with the associated coexistent pathologies is crucial for preventing permanent neurological deficit. The present case points out the potential harmful complications of underestimating a DST and its’ associated lesions. For instance the patient was operated one year ago for DST removal in another clinic and when his preoperative MRI scans were reevaluated, the tumoral lesion is clearly detected.
However, the parents admit that they were not informed about the tumoral mass neither before or after the operation.

The current recommendations state that infants less than 4-6 months of age harbouring cutaneous signs over the spine should be evaluated with spinal ultrasonography which can easily detect DSTs. MRI is the gold standard diagnostic technique for detecting the course of the tract and associated pathologies such as intradural tumors, tethered cord and split cord malformations. However, even MRI has limitations in showing the true extent of the associated pathologies with DSTs and this fact have been reported before. Ackerman and Menezes retrospectively analysed 28 patient with DSTs and reported that 14 of 24 patients that did not have sinus tracts appreciated on preoperative MRI scans had the pathologies observed intraoperatively. Radmanesh et al. reported that 40% of 35 patients operated for DST had tight filum terminale noticed intraoperatively which was evident in only 23% of preoperative MRIs. Based on these findings, the current accepted treatment of DSTs is surgical removal of all dermal sinuses and intradural exploration except the ones located at the sacrococcygeal area. The dura should be opened, intradural extension of the tract should be evaluated and any associated pathologies should be treated accordingly. The removal of DST alone does not prevent the potential harmful complications as reported in the present case.

Spinal abscesses are associated with high morbidity and mortality and they most frequently involve the epidural rather than subdural space. Spinal subdural abscesses are rare with approximately 70 cases reported in the literature. The most frequent age is between 60 and 70 years and the majority of the patients reported in the literature have one or more predisposing conditions, such as diabetes mellitus, alcoholism, tumors, iv drug abuse, degenerative joint disease, or anatomical abnormalities of the spinal cord or vertebral column. Dermal sinuses with concomitant dermoid or epidermoid tumors are frequently associated with recurrent meningitis and rarely with spinal subdural abscess. Subdural abscess formation due to direct contamination occurs when microorganisms invade the area via the fistula ending in the subdural space. However spinal subdural abscess is exceptionally rare in patients where the DST removed one year ago such as the present case.

The main symptoms of spinal subdural abscess include headache, irritability, prolonged fever and headache which may accompany neurological signs in late stages such as motor and sensory loss in the lower extremities and sphincter disturbances and deterioration of consciousness. It is impossible to predict the progressive course of the disease thus, urgent radiological and biochemical investigations along with a detailed neurological examination are crucial to reach definite diagnosis. The present case presented with prolonged fever progressive neurological deficit including paraparesia in lower extremities and finally deterioration of consciousness occurring on the day of his admission. Urgent radiological and biochemical investigations and surgical resection along with appropriate antibiotic treatment resulted in a successful outcome.

Contrast enhanced MRI of the spine remains the radiological investigation of choice in spinal subdural abscess with concomitant epidermoid tumor. Epidermoid tumors are hyper-intense on T2-weighted sections and hypo-intense on T1-weighted sections with peripheral contrast enhancement. However abscesses have similar MRI appearances with slightly higher peripheral contrast enhancement in T1-weighted sections. The interpretation of MRI appearance of...
an epidermoid tumor and concomitant abscess formation is difficult and arachnoidal adhesions and clumped nerve roots may additionally complicate this process\(^{(12)}\). In most cases, the definite diagnosis can only be made during surgical resection and after histopathological examination of the lesion. Preoperative biochemical test findings such as high WBC count and CRP levels along with the history and physical examination of the patient may aid in differential diagnosis. If MRI is not available, spinal contrast enhanced CT may be an alternative diagnostic method.

Spinal epidermoid tumor complicated with subdural abscess is an emergency condition and surgical removal of the tumor and drainage of the pus should be done as soon as the diagnosis is established. Infection further complicates surgical removal of the tumor by causing dense arachnoidal adhesions thus it is important to hold back from attempts to attain total removal of the scarred capsule wall. After local irrigation with saline, the dura should be closed in watertight fashion. Besides histopathological examination, the removed material should always undergo microbiological analysis. Wide spectrum antibiotics should be started and later adjusted according to the antibiogram test results. Previous reports and literature reviews on spinal subdural abscess revealed Staphylococcus Aureus as the most frequent causative agent\(^{(9,13,14)}\) while Proteus mirabilis was also reported in cases with abscesses associated with dermal sinus\(^{(10)}\). Our case exhibited Proteus mirabilis as the causative agent which was successfully treated with Teikoplanin+Meronem combination therapy. Early diagnosis and prompt surgical intervention with appropriate antibiotic treatment offer the best chance of functional recovery as presented in this case.

Hydrocephalus is a well-documented complication of post infectious meningitis\(^{(11)}\) as well as spinal dermoid cysts\(^{(8)}\). The proposed mechanisms include impaired CSF absorption at the cerebral convexities caused by increased levels of proteins, fibrinogen or degradation products of blood in the former and arachnoiditis or ventriculitis due to the dissemination of cyst contents in the latter\(^{(8)}\). Our patient exhibited both pathologies and the definite cause cannot be determined. However in our opinion more important than the etiology is diagnosing and treating the condition when it happens. Even after surgery the neurological status of the patient should be closely monitored and acute hydrocephalus should be included in differential diagnosis when raised intracranial pressure symptoms commence.

In conclusion, we present the diagnosis and successful treatment of a rare case of epidermoid tumor complicated with Proteus mirabilis meningitis and subdural abscess and later with hydrocephalus. The patient had a history of surgical DST removal one year prior to his admission. Thus simple excision of a DST will not obviate associated intradural pathologies which may cause potentially devastating results. The dura should always be opened and intradural extension of the lesion must be carefully evaluated. Spinal epidermoid tumor complicated with subdural abscess is an emergency condition and surgical removal of the tumor and drainage of the pus along with wide spectrum antibiotic treatment should be done as soon as the diagnosis is established. Postoperatively the neurological status of the patient should be closely monitored due to the risk of hydrocephalus.

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REFERENCES