Case Report

Fatal *Streptococcus melleri* Meningitis Complicating Missed Infected Intramedullary Dermoid Cyst Secondary to Dermal Sinus in a Saudi Child

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Summary

A spinal intramedullary abscess secondary to an infected dermoid cyst is rare, and it has a poor prognosis, unless diagnosed and treated promptly. We report a 12-month-old patient with a spinal intramedullary abscess with secondary to a dermoid cyst resulting from a dermal sinus, despite a clearly defined opening at the lower back with discharge of a purulent material from a dermal sinus tract seen on the lower back before the patient had become symptomatic and showed meningeal signs. The patient was managed as a case of meningitis until he had complications that endangered his life, and then further radiological evaluation was done to delineate the underlying pathology. This case illustrates the importance of the recognition and evaluation of skin markers because of the potential for intradural extension and a frequent association with other dysraphic abnormalities. It also emphasizes the importance of early diagnosis and treatment of spinal intramedullary abscess.

Key words: intramedullary abscess, *Streptococcus melleri*, dermal sinus.

Introduction

Congenital midline paraspinal lesions, mostly located in the lumbosacral area, are widely recognized as markers of occult spinal dysraphism. Congenital dermal sinuses represent cutaneous depressions or tracts that communicate between the surface of the skin and deeper structures. They are often accompanied by other cutaneous stigmata, various dysraphic abnormalities or intraspinal tumours. They become symptomatic because of infection or associated mass lesions. Spinal intramedullary abscesses are rare, and they have a poor prognosis, unless they are diagnosed and treated promptly [1]. We report a fatal case of intramedullary abscess associated with dermoid cyst secondary to dermal sinus.

Case Report

A 1-year-old boy from Saudi Arabia was admitted to the hospital, as he had been suffering from vomiting and diarrhoea for 1 week. He was brought to the emergency room and managed as a gastroenteritis case; he was rehydrated with intravenous fluids and discharged home. However, the child’s health did not improve, and after 3 days, he developed fever and lethargy. He was taken again to the emergency department. His examination at that time revealed a sick-looking febrile child with a temperature of 39°C. He had stiffness on the neck, and the reflexes over the upper and lower limbs were exaggerated. A small sacral dimple was noticed with greenish discharge. An urgent computed tomography scan of the brain was requested, and it showed dilated ventricles without hydrocephalus. A full septic work-up was done, including complete blood count, blood culture and lumbar culture. The white cell count was 15900 cells/mm³, with 75% polymorphonuclear cells. The lumbar puncture drained the pus-like CSF, which was sent to the laboratory. He was treated empirically on ceftriaxone 100 mg/kg/day and vancomycin 30 mg/kg/day. The patient’s case was registered as a case of meningitis, and he was admitted to the paediatric intensive care unit. In the microbiology laboratory, the CSF specimen was pus-like in nature on naked-eye examination and not suitable for cell count or latex testing. The Gram stain
revealed many pus cells and gram-positive cocci. The specimen was cultured on blood agar, chocolate agar, Todd Hewitt Broth and cooked meat broth. After 24 h, the plates showed no growth, whereas both broths were turbid and offensive. Gram stain of the broth showed gram-positive cocci and pleomorphic gram-negative bacilli. Aerobic and anaerobic subculture of the cooked meat and Todd Hewitt Broth after 48 h showed growth of *Streptococcus merrillii* and *Bacteroides fragilis* on the anaerobic plates but no growth on aerobic plates. Accordingly, metronidazole, 2.5 mg/kg/dose given every 8 h, was added to the antibiotic regimen. Blood culture was negative after 48 h of incubation. On the third day of hospitalization, the patient had several episodes of hypotension and bradycardia. Meropenem was added to cover the possibility of resistant organism, and ceftriaxone was discontinued. Despite antibiotic coverage, the boy continued to have fever. On the fifth day of hospitalization, he had weak pulses with cold extremities and poor perfusion. As septic shock was anticipated, dopamine 10 mcg/kg/min and epinephrine 0.05 mcg/kg/min were added. Regardless, his condition deteriorated and he was ventilated electively. Magnetic resonance imaging (MRI) of the brain and the spine was done on day 6 of hospitalization to delineate the anatomy of the central nervous system tract for the possibility of sinus tract connection from the skin to the spine and to rule out possible intra spinal abscess collection. The MRI of the brain and the spine revealed the following findings: the post-contrast images showed significant leptomeningeal enhancement along the surface of the brain and posterior fossa as well as on the visualized part of the spinal cord, with areas of nodular enhancement seen along the left sylvian fissure, in keeping with meningitis. No intracranial abscess could be visualized (Fig. 1). Images of the lower spine showed subcutaneous soft-tissue swelling, with sinus tract extending from the subcutaneous tissue to the spinal canal at the level of the sacral vertebrae S3 associated with spinal bifida. Abnormal signal intensity involving lower spinal cord was seen at T12–L2 level and expanding below that level till S3, which appears heterogeneously faint, high T2 and low on T1WI with significant post-contrast enhancement of the periphery of the lesion as well as leptomeningeal enhancement. Findings described are suggestive of an intraspinal mass lesion (dermoid) with a dermal sinus complicated by abscess formation in the lower spinal canal (Fig. 2). MRV demonstrates no evidence of dural sinus thrombosis. On the seventh day of hospitalization, an urgent exploration of the spinal cord was done by the neurosurgeon. The operative findings showed an infected intramedullary dermoid cyst (abscess) extending from the level of lumbar vertebrae 3 (L3) till the level of sacral vertebrae 3 (S3). A midline incision encircling the intramedullary abscess with aspiration of pus was performed. Dissection of the dermal sinus down to the dural matter, followed by laminectomy of L3, L4, L5, S1, S2 and S3, was done. Spina bifida was discovered from L5 till S4. Dural graft using lumbar fascia was closed tight with dura seal. The condition of the patient deteriorated after the operation. He had increased intracranial pressure with bradycardia. An external ventricular drain was inserted. Urgent computed tomography scan of the brain was done, which showed evidence of severe brain oedema, global brain ischaemia and brain injury. The patient had cardiac arrest on the ninth day of admission; cardio respiratory resuscitation was done, but the patient died despite aggressive resuscitation trials.

**Discussion**

Cutaneous alterations as indirect signs of occult spinal dysraphism can be found in ~86% of instances. Cutaneous stigmata that may indicate an underlying dysraphism of the lumbosacral spine include sacral cutaneous dimples, pigment changes, nevi, haemangiomata, dermal sinuses, subcutaneous lipomas and other mesenchymal tumours, hypertrichosis or skin tags and even tail-like cutaneous structures, the so-called human tail [1].

Congenital dermal sinuses represent cutaneous depressions or tracts that are lined by stratified squamous epithelium with surrounding dermal tissue. They may occur anywhere along the craniospinal axis, and the majority of these lesions occur in the lumbar or lumbosacral region, followed by the occipital and thoracic regions, respectively [2]. They are often accompanied by other cutaneous stigmata, various dysmorphic abnormalities or intraspinal tumours [3]. Approximately half of all dermal sinuses have associated dermoids or epidermoids, usually at the termination of the tract [4].

As a result, this tract could be a potential route of bacterial dissemination to the central nervous system. It can cause meningitis, infect the spaces around the spinal cord or even infect the spinal cord itself, basically when the latter comes in direct contact with the sinus [5]. Congenital dermal sinus is implicated as the leading cause of intramedullary spinal cord abscess in children. Intramedullary abscess is an extremely rare neurosurgical condition. Fewer than 100 cases have been reported, with <40 cases reported in paediatric patients, since this condition was first described by Hart in 1830 [6].

Intramedullary spinal cord abscess may present at any age, but children aged <5 years are more likely affected. This infection may result in a considerable damage to the spinal cord, and the neurological outcome may remain disappointing. MRI with gadolinium contrast is the gold standard of investigation before surgical planning. It revealed the lesion in all
cases, with cord enlargement and enhancement of the abscess wall [7].

Medical presentation of an acute intramedullary spinal cord abscess is consistent with fever, back pain, a partial or total transverse myelitic picture and increased white blood cells and erythrocyte sedimentation rate. Patients with chronic intramedullary spinal abscesses are less likely to present with fever or leucocytosis, and their symptomatology can mimic an intramedullary spinal tumour. A review of the literature reveals that patients with symptoms for <4 days have a 90% mortality rate, whereas those with symptoms for >1 week have a 67% mortality rate. Our patient had a dramatic course, with symptoms present for <1 week [8].

Cerebrospinal fluid analysis in these patients generally shows an elevated protein and variable white cell count, and although this is not specific, it should raise the possibility of an infective cause. Although 30% of cases are microbiologically sterile, a diverse range of organisms have been isolated, including Staphylococcus, Streptococcus pneumoniae, Hemophilus, Proteus, Listeria, Actinomycyes, Pseudomonas cepacia, Mycobacterium tuberculosis, S.milleria and Streptococcus mansonii. Anaerobes are rare, but multiple organisms are found frequently [8, 9]. Our case exhibited multiple organisms

Fig. 1. The lateral ventricles appear dilated with normal appearance of the fourth ventricle. There is an area of low signal intensity on T2 and FLAIR seen in the posterior horn of the left lateral ventricle, which shows blooming artifact on gradient echo images, in keeping with small intraventricular haemorrhage.
S. milleria and anaerobes B. fragilis). We were unable to find any primary infection area for the meningitis and the intramedullary abscesses in this patient, except the infected dermal sinus. It is advisable that once the diagnosis has been made, prompt decompressive laminectomy, myelotomy and surgical drainage, along with appropriate antibiotics, are the mainstays of treatment [6, 10]. In our patient, the outcome was poor because the diagnosis and surgical intervention were delayed.

Conclusion
Cutaneous markers have a crucial role in the detection and diagnosis of occult spinal dysraphism and require further investigation on initial observation. The development of spinal intramedullary abscesses by contamination through the dermal sinuses indicates the importance of early excision of such congenital lesions.

Although intramedullary spinal cord abscess is a rare disease, we should have knowledge of its existence because misjudgement and deferring adequate treatment may lead to an unfavourable outcome, and even to death.

References