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Case Report

7 Year Old Male with Acute on Chronic Back Pain

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- Epidermoid cyst
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Abstract

A 7-year-old previously healthy male presented with sudden worsening of a chronic lower back pain that had been present intermittently for 7 months requiring ibuprofen. His review of systems was essentially negative. Family history was negative for malignancies. Physical examination was only positive for a limping gait with tenderness in the right lateral lumbar region. CT of the lumbar spine, CBC, ESR, and CRP were normal. A lumbosacral MRI with and without contrast demonstrated a cystic structure, at the level of L3. After surgical excision, histology revealed a squamous epithelium lined cyst consistent with an epidermoid cyst. Low back pain is an uncommon but often serious presentation in children with an extensive list of differential diagnoses including musculoskeletal, neurologic, rheumatologic, oncologic, renal and infectious etiologies. Intraspinalepidermoid cysts are rare thin-walled inclusions lined by stratified squamous epithelium. Low back pain is the most common presentation of intraspinalepidermoid cysts. MRI is the imaging modality of choice and can distinguish intraspinalepidermoid cysts from other pediatric intraduralextramedullary spinal cord tumors. Spinal surgery is the mainstay of treatment and complete resection is usually curative.

ABBREVIATIONS

CT: Computed Tomography; **MRI**: Magnetic Resonance Imaging; **UTI**: Urinary Tract Infection; **ESR**: Erythrocyte Sedimentation Rate; **CBC**: Complete Blood Count; **CRP**: C Reactive Protein.

CASE PRESENTATION

During pediatric office visits, the complaint of low back pain presents a wide differential diagnosis. It requires a methodological assessment of the patient. We present the case of a 7-year-old previously healthy male child who presented with 4 days of sudden severe worsening of chronic lower back pain. Over the preceding 7 months, he had complained of back pain that was relieved intermittently by ibuprofen. This episode, however, occurred after his first game of basketball although there was no history of trauma. He presented with pain that was so severe he required admission to a local hospital intensive care unit for intravenous pain medication. His history is negative for rashes, hematuria, fever, weight loss, night sweats, numbness or constipation.

His only previous hospital admission was for a urinary tract infection (UTI) as a young infant. Laboratory work-up at that time included urine, blood, and CSF cultures. Voiding cystourethrogram demonstrated that he did not have vesicoureteral reflux (VUR), and he has not had recurrent UTIs. Family history was negative for malignancies.

On examination, vital signs were normal (pain = 4/10). His cardiac, respiratory, abdominal and genitourinary examinations were normal. His neurologic examination showed that he had normal sensation, reflexes, muscle strength, and no saddle anesthesia. Dermatologic exam did not reveal neurocutaneous findings. He had a limping gait with tenderness in the right lateral lumbar region. Straight leg raise would occasionally illicit tenderness but not with every exam. He had intermittent periods of screaming due to severe pain and distress.

He was evaluated with a CT of the lumbar spine which showed no abnormalities. His CBC, ESR, and CRP were normal. His pain persisted, requiring opiod analgesia and so he had a MRI was ordered. The lumbosacral MRI with and without contrast demonstrated a 2.2 cm x 1.3 cm x 1.4 cm nonenhancing T2 hyperintense intradural likely cystic structure at the level of L3. He underwent lumbar laminectomy during which a pearly white mass that was adherent to the nerve roots was visualized and excised. Histology revealed a squamous epithelium lined cyst consistent with an epidermoid cyst.

He had no major complications post operatively; he experienced a great reduction in the severity of the pain, improved gait and activity level, and no further episodes of

screaming or severe distress from pain. He was quickly advanced to regular diet required minimal post operative pain control and was discharged home on post operative day 4. In follow-up in neurosurgery clinic 4 months post operatively repeat MRI of the lumbar spine demonstrated no residual tumor (epidermoid cyst). Clinically he was doing well and had returned to all his normal activities including playing on a baseball team.

DISCUSSION

Low back pain is an uncommon presentation in children and warrants a thorough work-up for potentially serious causes. Multiple types of illness must be considered in the differential including musculoskeletal, neurologic, rheumatologic, oncologic, and infectious etiologies. Given the broad differential in such patients, a diagnosis can be hard to make without clear clues in the history or physical exam to guide clinical judgment. Here we report a 7-year old male who presented with low back pain and no other initial neurologic complaints. Initial assessment of his past medical history was benign with no obvious explanation of his current symptoms. The patient was eventually found to have an intraspinalepidermoid cyst after lumbosacral MRI was performed and the etiology of his condition became apparent after careful re-examination of early childhood illnesses.

Intraspinalepidermoid cysts are thin-walled inclusions lined by stratified squamous epithelium and constitute a rare pathologic finding that can either be congenital or acquired. The acquired forms account for approximately 40% of cases and are due to the implantation of epidermal cells into the subarachnoid space during procedures such as surgery, myelography, spinal anesthesia, and lumbar puncture [1]. These iatrogenic epidermal cysts were first described in 1956 as a complication of subarachnoid injections of streptomycin for treatment of tuberculosis meningitis [2]. The majority of iatrogenic cases were reported prior to the 1970s but their incidence has decreased due to the use of smaller lumbar puncture needles with stylettes [3,4].

Low back pain is one of the most common presentations of intraspinalepidermoid cysts [1,5]. This pain may be localized or radiate to the buttocks or lower extremities. Other common neurologic symptoms include leg paresthesia or weakness, gait disturbance, and urinary incontinence. Even in the absence of additional neurologic symptoms though, low back pain is a red flag in children and can provide evidence of an underlying intraspinal lesion.

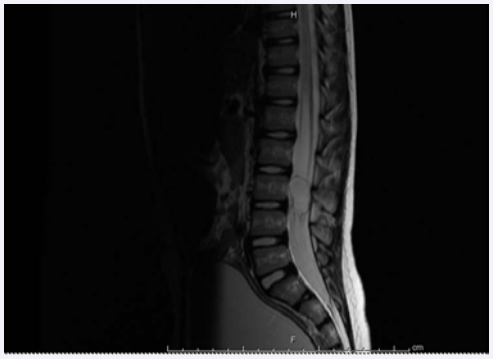

Figure 1 Lumbosacral MRI showing a nonenhancing T2 hyperintenseintradural, cystic structure at the level of L3.

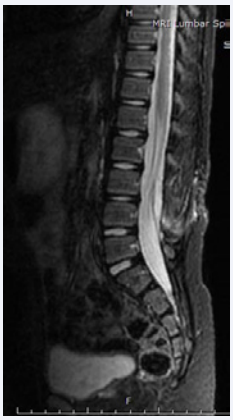

Figure 2 Post-surgical Lumbosacral MRI showing post-surgical changes and that the patient has had a laminectomy and evidence of resection of the mass.

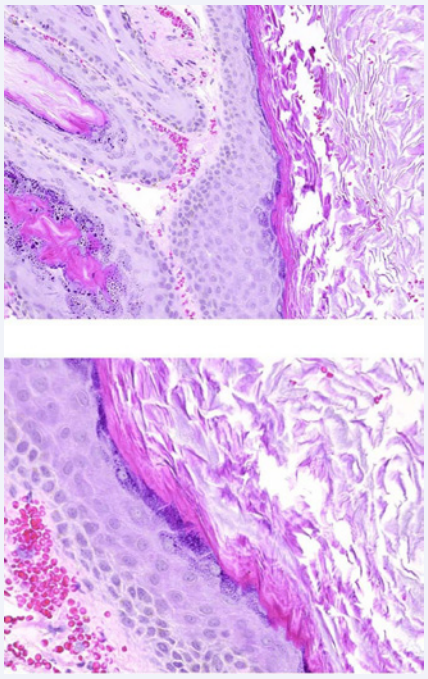

Figure 3 Hematoxylin and eosin stain at 20 x magnification and 40x magnification of surgically removed specimen showing squamous epithelium is seen surrounding a keratin-filled cyst that is consistent with an epidermoid cyst.

Lumbar puncture is the most common iatrogenic cause of intraspinalepidermoid cysts. The most common indication for lumbar puncture in recent case reports is part of an infectious work-up [1,4,6]. Children are also more commonly affected than adults [3]. This finding may be due to the preference of using needles without stylettes in neonatal units and children's hospitals [4]. Such needles make it easier for visualization of needle entry into the intradural space as there would be a rapid backflow of CSF. During the process, epidermoid cells can be carried in the

bore of the needle and implanted into the subarachnoid space [7-11]. The implanted cells slowly grow in the subarachnoid space until symptoms of nerve root compression become evident. The average latency period is approximately 7 years [12] but there are reports of periods as short as 18 months and as long as 40 years [12,13].

MRI is the imaging modality of choice for diagnosing intraspinalepidermoid cysts. These lesions appear isointense with T1-weighted images and hyperintense with T2-weighted images [14]. CT is usually non-contributory and there are characteristic features seen on MRI that help distinguish these lesions from other pediatric intraduralextramedullary tumors. They can be differentiated from neurofibromas, meningiomas, lipomas, dermoid tumors, and teratomas by lack of contrast enhancement [4,14]. Furthermore, they can be distinguished from dermoid tumors and lipomas by capsular enhancement and low signal on T1-weighted images [4,14].

Surgery is the mainstay of treatment for epidermoid cysts and is usually curative if all parts of the tumor can be removed. Care must be taken during the surgery to prevent cystic rupture and dissemination of cystic contents into the subarachnoid space which can lead to aseptic meningitis [15]. In a recent case series describing outcomes of 6 patients with intraspinalepidermoid cysts, all patients experienced pain relief and correction of other neurologic deficits over an average follow-up period of 58 months [16]. Additionally, none of these patients experienced a recurrence of their lesions. However, the authors of a larger case series of patients with either epidermoid or closely related dermoid cysts reported an approximate 17% recurrence rate [5]. Recurrence is more likely when the epidermoid cyst is not completely resected. This can occur when surgical removal is complicated by cyst adhesion to the caudaequina nerve roots or dura mater [17].

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