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Case Report

Possible Double Crush Syndrome Caused by Iatrogenic Acquired Lumbosacral Epidermoid Tumor and Concomitant Sacral Tarlov Cyst

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We describe a rare case of 42-year-old female who had possible double crush syndrome caused by iatrogenic spinal epidermoid tumor (ET) associated with lumbar puncture as well as concomitant sacral Tarlov cyst in close proximity. She presented with progressive left-sided perianal pain. She had a history of a Caesarean section with lumbar spinal anesthesia. Magnetic resonance imaging (MRI) demonstrated a relatively small intradural extramedullary solid lesion at L5/S1 level and cystic lesion at S2 level. We considered there were two different lesions, such as a tumor and Tarlov cyst; however, we could not rule out the possibility of a single lesion with two different components. Furthermore, there was a distinct compression at more than one locations along the course of the left S2 nerve root and we suspected possible double crush syndrome. We conducted tumor removal and the lesion turned out to be two different pathologies, such as an ET and Tarlov cyst. Both lesions were intraopertively pinching the left S2 nerve root at different sites as expected. The tumor was successfully removed and the cyst wall was imbricated and sutured. We need to take the possibility of ET into consideration if the patient underwent invasive spinal procedure previously. We also have to pay attention to the possibility of double crush syndrome if the nerve root possibly holding the responsibility for symptoms is compressed at two or more sites. This is the first report of possible double crush syndrome caused by acquired spinal tumor and congenital Tarlov cyst.

Keywords: double crush syndrome, epidermoid tumor, iatrogenic tumor, Tarlov cyst, lumbar puncture

Introduction

Epidermoid tumors (ETs) are benign tumors and rarely occur in the intraspinal region accounting for less than 1% of all primary spinal cord tumors.[1,2] The etiologies of spinal ETs are either congenital or acquired. Although congenital spinal ETs are often associated with spina bifida, spinal dysraphism, scoliosis, and cutaneous/dermal defects,[2–4] acquired spinal ETs are mostly caused by trauma or invasive procedures, such as lumbar punctures.[5] On the other hand, sacral Tarlov cysts are completely different and congenital pathologic lesions of the sacral nerve roots that are rarely symptomatic.[6] Spinal tumors and Tarlov cysts are sometimes difficult to differentiate if these pathologies are coexistent in close proximity to each other.[7]

We report a rare case of 42-year-old female who had possible double crush syndrome caused by iatrogenic spinal ET associated with lumbar puncture as well as concomitant sacral Tarlov cyst in close proximity. It was undetermined preoperatively whether there existed a single spinal tumor or two different pathologies, such as spinal tumor and Tarlov cyst; however, we found an ET and Tarlov cyst in the same vicinity intraoperatively. Both lesions were suspected of holding responsibility for her symptoms because the left S2 nerve root was pinched at two different sites by both of ET and Tarlov cyst on preoperative magnetic resonance imaging (MRI), namely possible double crush syndrome, which was made clear intraoperatively. Therefore, both lesions were operated at the same time. Her neurological symptoms completely disappeared immediately after surgery. We have to pay attention to the possibility of double crush syndrome and need to treat all relevant pathologies if the nerve root possibly holding the responsibility for symptoms is compressed at two or more sites by two or more lesions. This is the first report of possible double crush syndrome caused by acquired tumor and congenital Tarlov cyst.

Case Report

History

A 42-year-old female presented with progressive and severe left-sided perianal pain over a period of 10 months. She had not noticed urinary symptoms specifically until she was asked directly. She denied any history of trauma, infectious diseases, or surgery related to her spine. She had a history of a Caesarean section with lumbar spinal anesthesia. She remembered repetitive punctures of her lower lumbar spine at the time of spinal anesthesia. Spinal dysraphism or skin abnormalities were not observed by visual inspection in her lumbar/sacral region.

Examination

On her neurological examination, she exhibited full strength of muscle power and no evidence of a sensory deficit in her upper and lower extremities. She complained of characteristic numbness and pain in the perianal region only on the left side. Furthermore, she was found to have urinary symptoms, including urinary frequency and a feeling of

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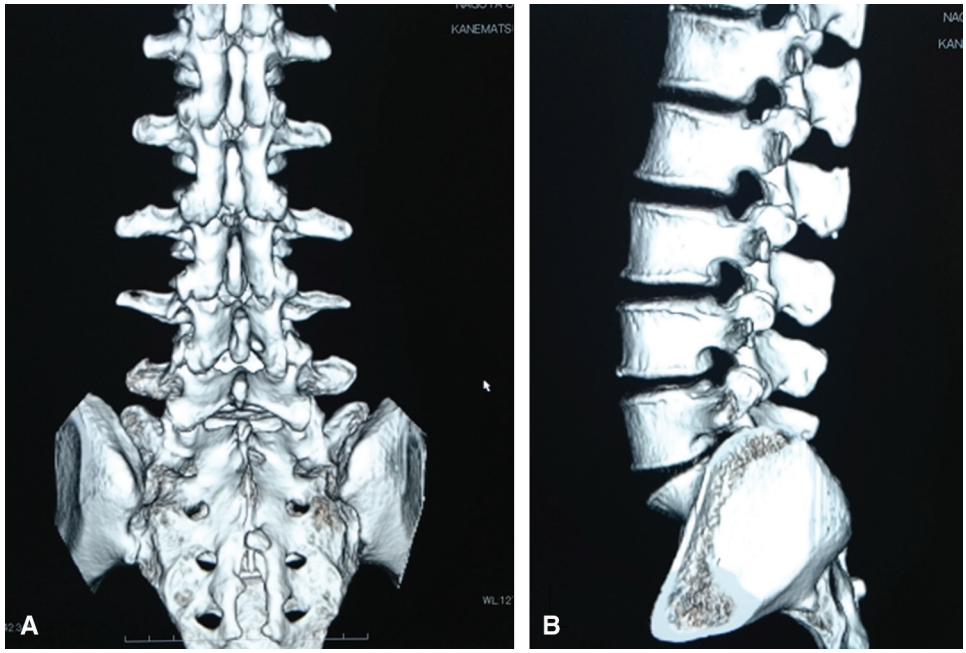

Fig. 1 CT of lumbosacral spine did not show any congenital bone anomaly. (**A**) AP view and (**B**) lateral view. CT: computed tomography.

residual urine. Reflexes were normal in the upper and lower extremities on both sides. Radiographs and computed tomography (CT) of lumbosacral spine did not show any congenital bone anomaly (Fig. 1). MRI demonstrated a relatively small intradural extramedullary solid lesion (28 mm × 17 mm × 17 mm) at L5/S1 level (Figs. 2A–2F, arrow) and cystic lesion (21 mm × 11 mm × 13 mm) at S2 level (Figs. 2A–2F, arrowhead). MRI with contrast material revealed rim enhancement of the solid lesion without any enhancement of the cystic lesion (Figs. 2C and 2D, arrow and arrowhead). We considered there were two different lesions, such as a tumor and Tarlov cyst; however, we could not rule out the possibility of a single lesion with two different components. Judging from her characteristic numbness and pain distribution in perianal region only on the left side with bladder dysfunction without leg symptoms, her symptoms were deemed being attributed to left S2 nerve roots impingement because the left S2 nerve root seemed to be squeezed at two different sites by both of solid lesion at L5/S1 level (Fig. 2F, arrow) and cystic lesion at S2 level (Fig. 2F, double arrowhead), namely possible double crush syndrome. Her symptoms were severe enough to interfere with her daily activities; therefore, surgical treatment was planned.

Operation

The tumor removal was performed using microscopy following a laminectomy from L5 to S2. The lesion turned out to be two different pathologies without continuity between them (Fig. 3A). These pathologies were presumptively diagnosed with ET (Fig. 3A, arrow) and Tarlov cyst (Fig. 3A, arrowhead) at this point based on the characteristic appearances intraoperatively. The left S2 nerve root was squeezed by the tumor capsule and there was mild adhesion between the nerve root and tumor capsule. The fragile capsule ruptured during the manipulation of ET (Fig. 3B, arrow). The spilled tumor contents were removed carefully and completely. The spinal canal was flushed with a large amount of water after complete resection of the tumor (Fig. 3C). Then, we proceeded to Tarlov cyst. The cyst seemed to be originating from the right S2 nerve root (Fig. 3C, arrowhead) and expanding into the left side to compress the left S2 nerve root (Fig. 3C, double arrowheads) at different locations from ET. Following incision into the thin cyst wall, cerebrospinal fluid (CSF) flowed out quickly and the right S2 nerve root was identified inside the cyst. The cyst wall was imbricated and sutured (Fig. 3D, double arrowheads).

Pathological findings

Histological examination of the tumor specimen demonstrated that the cyst wall was lined with stratified squamous keratinizing epithelium surrounded by an outer layer of collagenous tissue with the absence of skin adnexa (Figs 3E and 3F). Abundant keratin material was also observed. A diagnosis of ET was confirmed.

Postoperative course

She became completely free from preoperative perineal pain immediately after surgery. Her MRI showed complete resection of the tumor without recurrence at 2-year postoperative follow-up (Figs. 4A–4C).

Discussion

Considering solid and cystic lesion existed in close proximity to each other, we need to take both possibilities into consideration, such as a single cystic tumor like schwannoma[7] or a combined lesion of a spinal tumor and Tarlov cyst. Schwannomas tend to have cystic formation with a low

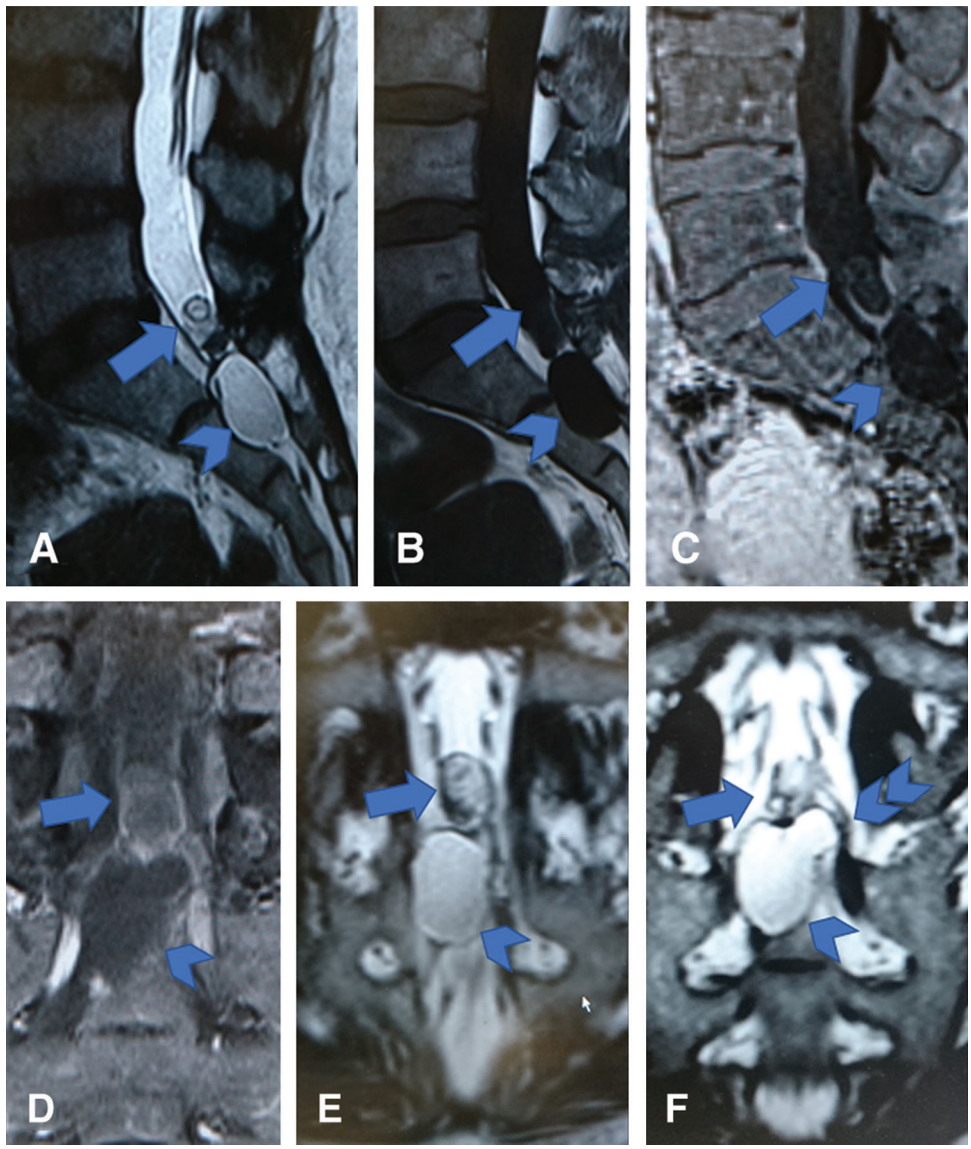

Fig. 2 MRI of lumbosacral spine: (**A**) sagittal view of T2WI, (**B**) sagittal view of T1WI, (**C**) sagittal view of gadolinium-enhanced T1WI, (**D**) coronal view of gadolinium-enhanced T1WI, (**E** and **F**) coronal view of T2WI. MRI demonstrates a small intradural extramedullary solid lesion (arrow, 28 mm × 17 mm × 17 mm) at L5/S1 level and cystic lesion (arrowhead, 21 mm × 11 mm × 13 mm) at S2 level. MRI with contrast material revealed rim enhancement of the solid lesion (arrow, **B, C, D**) without any enhancement of the cystic lesion (arrowhead, **B, C, D**). MRI: magnetic resonance imaging, T1WI: T1-weighted image, T2WI: T2-weighted image.

to intermediate signal intensity on T1-weighted images (T1WI) and a heterogeneous appearance with focal areas of hyper- and hypo-intensity corresponding to intratumoral cyst on T2-weighted images (T2WI).[8] Rim enhancement of cyst wall on gadolinium-enhanced MRI is characteristic of schwannomas. The present case showed rim enhancement of solid lesion mimicking a schwannoma; however, there was no enhancement of cystic lesion, indicating two different lesions. Spinal ETs are still difficult to diagnose particularly for small-sized tumors like the present case because they typically show negative contrast enhancement[8,9] and similar signal to CSF on MRI.[2,10–12] ETs can show atypical signal intensity changes, such as hyperintense signal on T1WI and hypointense signal on T2WI. This variability in signal characteristics might be related to the chemical state of cholesterol or the relative composition of cholesterol and keratin and makes preoperative diagnosis difficult.[8,9]

The etiologies of spinal ETs are thought to be both congenital and acquired. Acquired ETs were mostly reported in the region of the cauda equina (below the L1 level)[5,13] because lumbar punctures are usually performed around that level. In the present case, she did not exhibit any congenital spinal bone and skin anomaly. Epidermoid cells may have been introduced into the spinal canal at the L5/S1 level during lumbar punctures. The tumor location, which was exactly between spinous processes, corresponds with the insertion tract of the spinal needle and suggests the iatrogenic origin. Morita et al. found acquired ETs are more likely to be

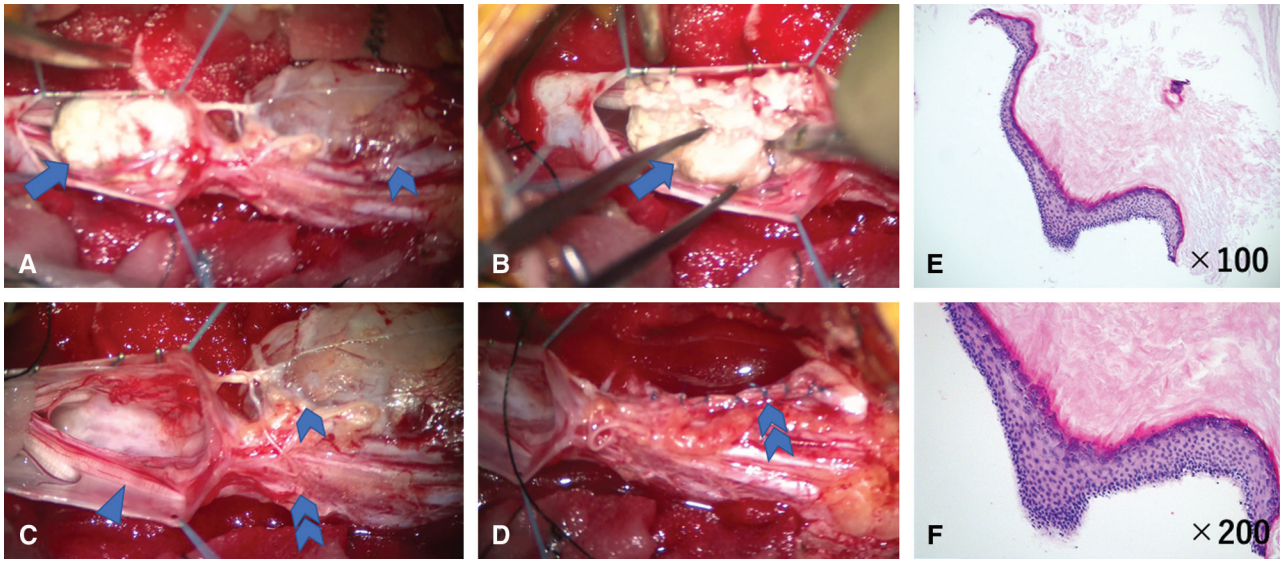

Fig. 3 Intraoperative photographs (**A–D**) and pathological findings (**E** and **F**). The lesion turned out to be two different pathologies and were presumptively diagnosed with ET (**A**; arrow) and Tarlov cyst (**A**; arrowhead). The left S2 nerve root was squeezed by the tumor capsule. There was a mild adhesion between the thin tumor capsule and the left S2 nerve root and the fragile capsule ruptured during the manipulation of the tumor (**B**; arrow). The spilled tumor contents were removed carefully and completely (**C**) S1 nerve root was squeezed by ET (**C**; triangle). Tarlov cyst was tense (**C**; arrowhead), compressing the left S2 nerve root at different location from ET (**C**; double arrowheads). Following incision into the thin cyst wall, CSF flowed out and the nerve root was identified inside the cyst. The cyst wall was imbricated and sutured (**D**; double arrowheads). Histological examination demonstrated that the cyst walls lined with stratified squamous keratinizing epithelium (**E**; HE, magnification ×100), surrounded by the outer layer of collagenous tissue with the absence of skin adnexa (**F**; HE, magnification ×200). A diagnosed of epidermoid cysts was confirmed. CSH: cerebrospinal fluid, ET: epidermoid tumor, HE: hematoxylin and eosin.

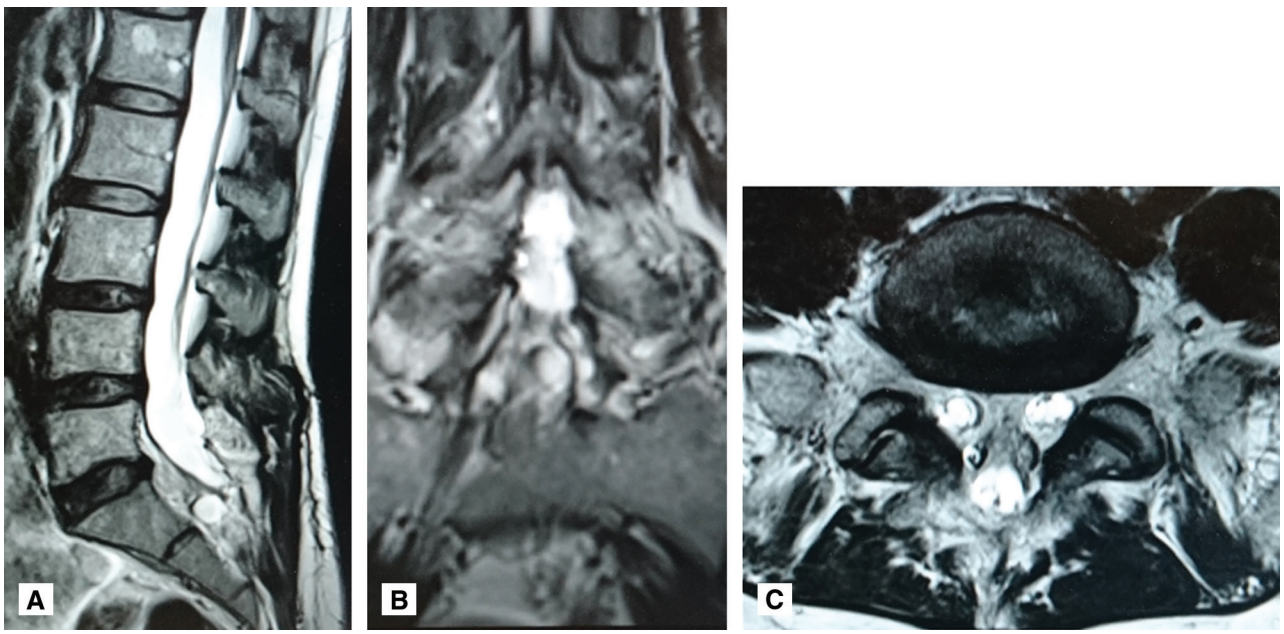

Fig. 4 MRI of lumbosacral spine 2 years after surgery: (**A**) sagittal view of T2WI, (**B**) coronal view of T2WI, (**C**) axial view of T2WI. MRI shows complete resection of ET and no recurrence of Tarlov cyst. ET: epidermoid tumor, MRI: magnetic resonance imaging, T2WI: T2-weighted image.

symptomatic compared to congenital ones. Although the clinical symptoms of spinal ETs tend to be variable and nonspecific with slow progression,[2,14] the present case followed an acute course and the ET is relatively small for the patient's significant perianal pain. This is because there was a distinct compression at two different locations along the course of left S2 nerve root caused by both of acquired ET and congenital Tarlov cyst, called possible double crush syndrome.[15] The

patient complained of characteristic unilateral (left-sided) perianal pain and urinary symptoms without leg pain, numbness or weakness indicating absence of S1 nerve root symptoms even as S1 nerve root was squeezed by ET alone at L5/S1 level as confirmed intraoperatively (triangle to show S1 nerve root in Fig. 3C). S2 nerve root was suspected of being compressed by both of ET and Tarlov cyst on preoperative MRI, which was also confirmed intraoperatively. We thought subclinical left S2 nerve compression by congenital Tarlov cyst became symptomatic with multiple nerve impingement by both of ET and Tarlov cyst following progression of acquired ET though S1 nerve root was not symptomatic with single compression. Therefore, we have to be careful not to underestimate the presence of congenital lesion or anomaly in close proximity to acquired lesion and recommend both pathologies be treated at the same time.

Surgical treatment is required when the patients develop refractory neurological deficits. The surgical strategy for the present case consists of removal of ETs and imbrication of Talrov cyst because of the multiple compression of left S2 nerve root. Complete excision is an essential part of surgical treatment for ETs.[16] However, the ETs' capsules are very fragile, and often adhere to the nerve sheath.[17–19] Local recurrence is reported especially after subtotal excision.[2,11,14] It is suggested that incomplete removal of basal germinal cells of the tumor induce tumor recurrence and produce an inflammatory reaction leading to chemical meningitis.[2,10] Eventually, we were able to achieve complete resection of the tumor's capsule with emptying the contents of tumor capsule as well as successful imbrication of Tarlov cyst. Fortunately, the patient did not develop anti-inflammatory reaction or neurological deterioration postoperatively. There was no recurrence at 2-year follow-up with a good neurological outcome. However, long-term follow-up is required for a potential risk of tumor recurrence.

Conflicts of Interest Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

The analysis of the case was conducted in accordance with the "Ethical Guidelines for Medical and Health Research Involving Human Subjects."

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