Pelvic pain from a giant presacral Tarlov cyst successfully obliterated using aneurysm clips in a patient with Marfan syndrome

Case report

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Patients with Marfan syndrome used to succumb early in life from cardiovascular complications. With the current rapid advance in medical and surgical care, such patients may now have near-normal longevities. Consequently, rare late-life complications are emerging in these patients and represent challenges to clinicians for their diagnoses and treatments. The authors report a rare case of pelvic pain and genital prolapse from a giant presacral Tarlov cyst in a 67-year-old patient with Marfan syndrome.

This 67-year-old Caucasian female presented with progressively severe pelvic pain, intermittent explosive diarrhea, and dysuria. Physical and bimanual examination demonstrated genital prolapse and a nontender, cyst-like mass fixed in the midline. She underwent ultrasound, CT, and eventually MRI evaluations that led to the diagnosis of a giant (6.7 × 6.4 × 6.6 cm) Tarlov cyst originating from the right S-2 nerve root sleeve/sacral foramen with intrapelvic extension. She underwent S1–S2 and S2–S3 laminectomy with obliteration of the Tarlov cyst using aneurysm clips. Postoperatively, her pelvic pain and bowel symptoms resolved and the bladder symptoms improved. The 3-month follow-up CT of abdomen/pelvis demonstrated resolution of the cyst.

The present case illustrates that clinicians caring for elderly patients with Marfan syndrome need to increasingly recognize such unusual late-life complications. Also, these large Tarlov cysts can be simply and effectively obliterated with aneurysm clips.

Key Words • Marfan syndrome • Tarlov cyst • aneurysm clip • sacral

In his original paper of 1896, Antoine Bernard-Jean Marfan described congenital deformities of the extremities in a 5-year-old girl with a hereditary medical history.11 Although the syndrome now bears his name, Marfan made no reference to the other cardinal involvement of ocular and cardiovascular systems typically seen in these patients. More contemporary accounts of Marfan syndrome detail the many facets of the syndrome;2 however, the associated neurological disturbances tend to occur at older ages and are therefore not as well recognized.24 With the current rapid advance in medical and surgical care, patients with Marfan syndrome may now enjoy near-normal life expectancies.8 Consequently, rare late-life complications of this condition are emerging and represent clinical challenges for diagnosis and treatment. Increasingly, neurosurgeons encounter specific problems among patients with Marfan syndrome due to the common occurrence of defects in the dura mater in such patients.6 Prompt recognition of the associated neurological complications is crucial, and neurosurgical treatment protocols are needed. We report a rare case of pelvic pain, genital prolapse, and bowel and bladder symptoms arising from a giant presacral Tarlov cyst in a 67-year-old patient with Marfan syndrome.

Case Report

History and Presentation. This 67-year-old Caucasian woman with Marfan syndrome presented with an 8-year history of progressively severe pelvic pain, intermittent explosive diarrhea, and dysuria. Physical and bimanual examination demonstrated genital prolapse and a nontender, cyst-like mass fixed in the midline. Ultrasonographic examination of the patient’s pelvis demonstrated a 6.7 × 6.4 × 6.6–cm solid, fluid-filled cystic mass with internal septation near the right ovary. A CT scan of the abdomen/pelvis revealed a large Tarlov cyst originating from the right S-2 nerve root sleeve/sacral foramen with
intrapelvic extension (Fig. 1 left). Magnetic resonance imaging showed cystic dilation of multiple sacral nerve root sleeves consistent with Tarlov cysts (Fig. 2 upper and lower). The largest was noted on the right at S-2, extending anteriorly into the pelvis and exerting extrinsic mass effect on adjacent loops of bowel and other pelvic contents.

**Operation and Postoperative Course.** The patient underwent S1-S3 laminectomy to expose the neck of the cyst (Fig. 3 upper). The Tarlov cyst neck was successfully obliterated with aneurysm clips (Fig. 3 lower). The intrapelvic cyst cavity was subsequently entered, and aspiration of the cyst fluid was performed with suction (Fig. 3 lower). The aneurysm clip obliterated the neck of the cyst along with the accompanying nerve root although the patient exhibited no postoperative neurological deficit. Postoperatively, her pelvic pain and bowel symptoms resolved and the bladder symptoms improved. The 3-month-follow-up CT scan of the abdomen/pelvis demonstrated resolution of the cyst (Fig. 1 right).

**Discussion**

Marfan syndrome, an autosomal-dominant inherited syndrome caused by **fibrillin-1 (FBN1)** gene mutations, represents one of the most commonly inherited connective tissue disorders. As a major component of microfilaments, which are abundantly expressed in most tissues and organs, fibrillin-1 has a ubiquitous distribution in connective tissue. Therefore, patients with Marfan syndrome have signs and symptoms that affect many organ systems, including cardiovascular, ocular, skeletal, dermal, pulmonary, and dural systems.

The clinical diagnosis of Marfan syndrome currently relies on a combination of major and minor clinical signs and symptoms. While rare in the general population, dural ectasia is the most prevalent feature of this syndrome and may be evident in more than 90% of patients. Dural ectasia has been classified as a major criterion for the diagnosis of Marfan syndrome. The severity of dural ectasia seems to correlate with age. Fibrillin-1 constitutes the scaffold for deposition and maturation of tropoelastin in elastin fibers of the dura mater, and this may explain the progressively severe dural ectasia with age. The prolonged gravity and pulsation effects from CSF in patients with Marfan syndrome likely explain the progression of dural ectasia with age and the gradual erosion of the vertebral bodies (scalloping). If the altered dural sac dilates and protrudes through the neural foramina, arachnoid cysts, meningocele, and meningeal cysts may develop.

With advances in medical and surgical care, the mean life expectancy of patients with Marfan syndrome has significantly increased. The neurological complications related to the altered spinal meninges in these patients typically occur later in life. Because of the increased life expectancy, such neurological complications are emerging and represent clinical challenges for their diagnoses and treatments. Many of the Marfan syndrome pathologies related to the spinal meninges, such as dural ectasia, arachnoid cysts, and meningeal cysts, often do not
Pelvic pain from a giant presacral Tarlov cyst

First reported by Tarlov in 1938, Tarlov cysts are currently classified as Type II meningeal cysts. Although the majority of Tarlov cysts are asymptomatic, a small subset of these sacral perineural cysts may become symptomatic and warrant treatment. Clinical symptoms include radicular pain, sensory disturbances, motor deficits, abdominal pain, impotence, bladder/bowel dysfunction, and intracranial hypotension if the cyst ruptures. Currently, there are no established guidelines or consensus with regard to the best management option for symptomatic Tarlov cysts. Nonsurgical treatment includes medical therapy (analgesic, nonsteroidal antiinflammatory medications, and steroids) and physical therapy. Surgical treatment strategies conceptually involve either diversion of CSF flow (aspiration or shunt procedures) or a direct microsurgical approach. Several authors have observed that surgical treatment may accomplish better clinical outcome in patients with radicular symptoms, bladder/bowel dysfunction, and cyst diameters exceeding 1–1.5 cm. Such surgical methods consist of decompressive laminectomy, cyst neck or wall resection, cyst fenestration and imbrication, or cyst shrinkage via bipolar cautery. Because of the dural thinning and fragility from ectasia, symptomatic Tarlov cysts in patients with Marfan syndrome represent a particular challenge for neurosurgical intervention. A direct surgical approach is associated with a high risk of dural tears and osseous complications. The advantage of our approach over other surgical methods is that it is less likely to tear the dura while accomplishing direct obliteration of the connection between the thecal sac and cyst. Other approaches may accomplish the same goal without using an aneurysmal clip, but are more likely to cause dural tearing, especially in patients with Marfan syndrome.

Fig. 3. Intraoperative photographs. Upper: After S1–2 and S2–3 laminectomy, the neck of the Tarlov cyst (asterisk) was exposed. Lower: The neck of the cyst was successfully obliterated with 2 aneurysm clips. The intrapelvic cyst (cystic cavity indicated by asterisk) was subsequently entered, and aspiration of the cyst fluid was performed. Orientation for both images: top of image, caudad; bottom of image, cephalad; right side of image, patient’s left side; left side of image, patient’s right side.

produce any symptoms; however, radicular, myelopathic, and urinary symptoms from such pathologies have been reported in patients with this syndrome. In addition, spontaneous spinal CSF leaks are much more commonly observed in patients with Marfan syndrome.

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Conclusions

Advancements in medical and surgical treatment have resulted in significantly increased life expectancy for patients with Marfan syndrome. Increasingly, neurosurgeons encounter neurological complications that typically occur later in life and relate to the spinal meninges, such as dural ectasia, arachnoid cysts, and meningeal cysts. Prompt recognition of the associated neurological complications, and development of neurosurgical treatment protocols are becoming essential. We have reported the successful treatment of a giant symptomatic presacral Tarlov cyst in a patient with Marfan syndrome using aneurysm clips.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: H Wang. Acquisition of data: H Wang. Analysis and interpretation of data: H Wang. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: H Wang. Approved the final version of the manuscript on behalf of all authors: H Wang.

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