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Case Report

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Presacral Schwannoma Mascquerading as a Uterine Leiomyoma

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ABSTRACT

Presacral tumors are a rare group of tumors located posterior to the rectum in the retrorectal or presacral space. Due to the lack of specific symptoms and difficult anatomic localization, they present a diagnostic and management challenge for the surgeons. We present a case of retrorectal schwannoma in a young female who was initially misdiagnosed as a uterine fibroid on ultrasound and taken up for myomectomy. The tumor was later successfully excised by a combined abdominosacral approach. The clinical features and management of presacral tumors are discussed in this article.

Introduction

Presacral tumors are uncommon lesions that can be difficult to diagnose because of their nonspecific signs and symptoms. Neurogenic tumors, including schwannoma (Neurilemmoma), account for 10% of all presacral lesions. Approximately 85% of neurogenic tumors are benign. Complete surgical excision is the choice for management, and the long-term prognosis is good.

Case Report

A 21-year-old female was referred to our institution with mild lower abdominal pain and impairment complaints for the last three years. She was unmarried and nulliparous. She had attained menarche at 14 years and her menstrual periods were normal. Two months back, as per her treatment records, she was diagnosed with a large pelvic leiomyoma on ultrasonography in a peripheral hospital and was taken up for a myomectomy. There was a large pelvic mass be-

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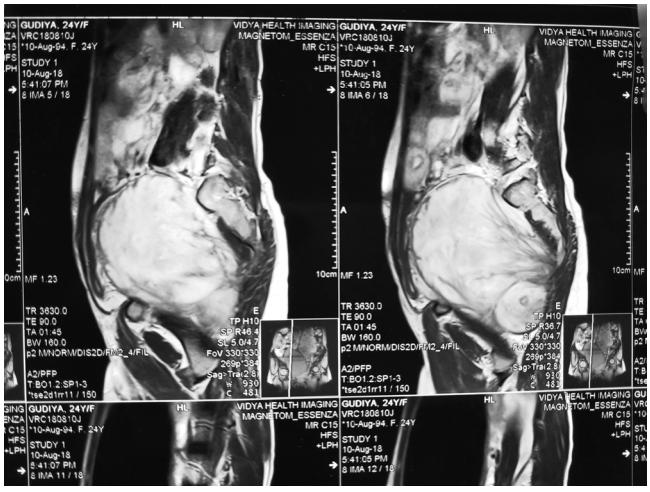

Figure 1. Sagittal section of T2-weighted MRI Pelvis showing a large soft tissue mass in the presacral region. The mass is seen compressing the urinary bladder and uterus.

hind the rectum displacing the rectum and uterus anteriorly at laparotomy. The urinary bladder, uterus, fallopian tubes, and ovaries appeared normal. The surgery was abandoned, and the patient was referred to us. No biopsy had been taken from the mass.

On examination, a scar on the Pfannenstiel incision appeared healthy. A palpable lower abdominal mass reached about 10 cm above the pubic symphysis. Per vaginal examination was not done as the patient was unmarried. On per rectal examination, there was a firm non tender mass palpable mass posteriorly beginning about 5 cm above the anal verge and the upper limit of the mass could not be reached. The CT scans and MRI of the abdomen showed a 16x14x12 cm sized, mildly and heterogeneously enhancing soft tissue mass in the presacral region, causing anterior displacement and compression of the uterus, cervix, vagina, ovaries, and urinary bladder without any obvious infiltration (Figure 1). Significant compression over the distal rectum and anal canal led to a severe proximal dilatation of the fecal matter-filled rectum and sigmoid colon (Figure 2). The mass was causing bony erosion of the sacrum with a small intrascrotal extension of the lesion through the lower presacral foramina on the right side, suggestive of neoplastic etiology (Figure 3).

The patient was taken up for surgery using a combined abdominosacral approach. The abdomen was opened by a lower midline laparotomy with the patient in the "sloppy lateral position," permitting both abdominal and sacral access. The rectum and descending colon were grossly distended, and the fecal matter was evacuated by manual pressure after passing a rectal catheter. The tumor was approached following the mobilization of the sigmoid colon and was separated posteriorly from the presacral fascia.

Due to the larger tumor size, care was taken to preserve the S3 nerve root and the anterior division of the internal iliac artery. The next portion of the surgery was initiated through a sacrococcygeal incision concerning the external sphincter, and the tumor was dissected free from the rectum. The can-

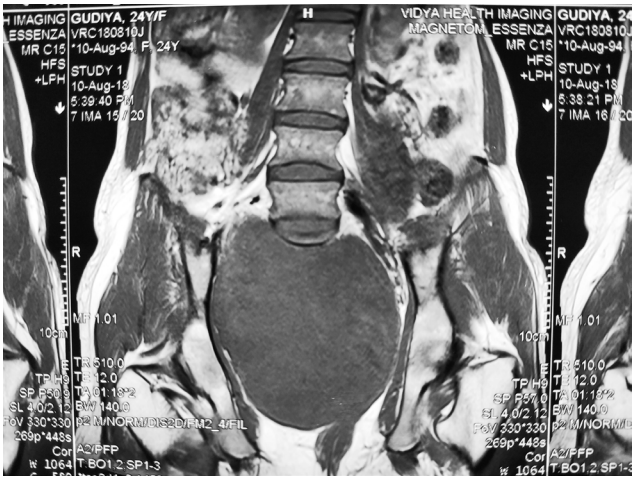

Figure 2. Coronal Section of T2-weighted MRI Pelvis showing a large pelvic mass.

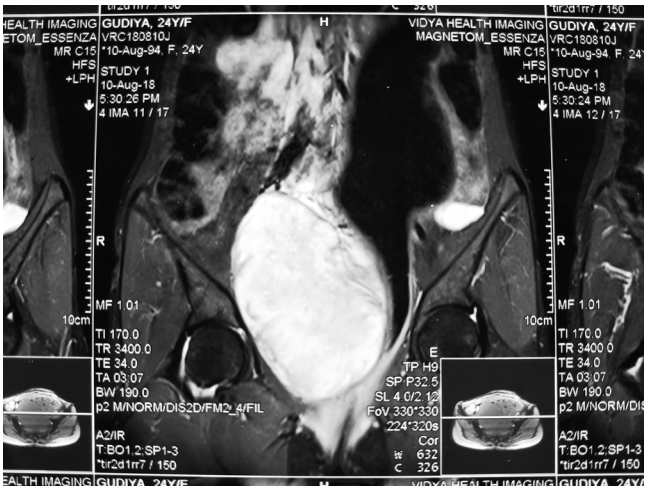

Figure 3. Coronal section of T2-weighted MRI Pelvis showing the pelvic mass causing gross dilatation of rectum.

cer was delivered through the abdominal incision. Suction drains were placed in the presacral space, and the abdominal and perineal wounds were sutured in layers. The postoperative period was uneventful, and the patient was discharged on the sixth postoperative day. The patient is asymptomatic after a follow-up of two years.

Discussion

The presacral or retrorectal space represents the continuation of the retroperitoneum into the pelvis. This potential space is located between two anatomical structures, the presacral fascia of the sacrum (Waldeyer's fascia) and the parietal peritoneum of the posterior abdominal wall. In adults, this site contains retrorectal fat, loose connective tissue, lymph nodes, the median sacral vessels, the superior rectal vessels, and sympathetic and parasympathetic branches [1].

If these neural and vascular structures are harmed or injured, the rectoanal physiology is seriously affected and may cause substantial musculoskeletal and/or neurologic morbidity. Notably, if the S3 nerve is damaged bilaterally, the external sphincter malfunctions, leading to various degrees of incontinence. Congenital tumors are the most common presacral lesions, 55% to 70% of all lesions in the presacral area. These include developmental cysts, chordomas, and anterior meningoceles. Neurogenic tumors originate from the peripheral nerves and are the second most common presacral tumors after congenital tumors and account for 10% of all presacral lesions [2]. Approximately 85% of neurogenic tumors are benign. Benign lesions include neurofibromas, neurilemomas (schwannomas), and ganglioneuromas, whereas malignant lesions include neuroblastomas, ganglioneuroblastomas, ependymomas, and malignant peripheral nerve sheath tumors (malignant schwannomas, neurofibrosarcomas, and neurogenic sarcomas). These tumors tend to be slow-growing and cause minimal or nonspecific symptoms; therefore, they may be of considerable size at the time of diagnosis. Presacral tumors, especially benign ones, are asymptomatic for a long time or produce minimal or nonspecific symptoms, such as vague abdominal pain, constipation, paradoxical diarrhea, rectal tenesmus, and sexual dysfunction [3, 4]. Presacral schwannomas may mimic uterine myomas due to their proximity to the uterus and image density similar to muscle.

Similarly, like a leiomyoma, they may undergo degeneration, focal necrosis, or calcification. Casey et al. reported a case of a 47-year-old woman taken up for laparoscopic hysterectomy wherein the pedunculated myoma on ultrasound turned out to be a retroperitoneal schwannoma on fine-needle aspiration cytology [5].

Özkardeş reported a similar misdiagnosis, and the hysterectomy was abandoned, and a biopsy showed a schwannoma [6]. Chen et al. reported a case of multiple fibroid uterus with a left anterior 7 cm pedunculated myoma [7]. During the operation, the mass was excised and was firm, solid, and well-capsulated. The mass adhered to the bladder serosa and was close but not connected to the uterus. A total hysterectomy was performed because of multiple small uterine fibroids. The final pathologic diagnosis of the tumor was a cellular schwannoma located in the retroperitoneum in their case [7].

Presacral neural tumors are rare and hardly ever encountered by gynecologists who may not be equipped to handle them operatively due to a lack of experts in the field. Per rectal examination must be done in unmarried females with pelvic tumors as per vaginal examination is omitted due to ethical issues. Such assessment will help clinically differentiate an anteriorly located pelvic tumor from a retro rectal one. Preoperative MRI/CT is not routinely done before sur-

gery for leiomyomas but may aid in diagnosing these rare tumors so that appropriate surgical management may be planned. Schwannomas are heterogeneous at MR imaging, with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, and tend to have small cystic areas and a thin pseudo capsule [8].

CT is used to reveal any destruction of the bone cortex, the solid or cystic nature of the lesion, and whether the tumor is infiltrating any neighboring viscera. MRI, being more specific than CT, may provide a detailed view of the anatomical correlations and histology of the tumor and neural tissue involvement.

Surgical resection is the treatment of choice for all presacral tumors, even if they are asymptomatic. Surgical resection is necessary due to the patient's risk of underlying malignancy or malignant transformation, spontaneous infections of cystic lesions, obstruction to vaginal delivery in females, constipation, and chronic discomfort. The surgical excision may be accomplished through three approaches: anterior (abdominal), posterior (sacral), and combined abdominosacral. The selection of a specific approach depends on tumor characteristics (nature, size, and location) [2].

The potential infiltration of the sacrum, pelvic sidewall, and adjacent structures should also be considered. Lesions extending above the S4 level are usually resected through the anterior approach, while lower lesions through the posterior. In case the upper extent of the lesion is palpable on rectal examination, the lesion can be resected transracially [9].

A combined abdominosacral approach is preferred when the distal margin of the tumor is lower than S3 and the cephalic margin higher. Furthermore, this approach is applied to cases where the neoplasms invade the rectum or adjacent structures. Distinct benefits linked to the combined system may be the better visualization of structures through the anterior incision and the improved exposure of the nerve roots via the posterior approach. An intraoperative digital rectal examination is vital for all three approaches to avoid rectum injury while dissecting the tumor. For trans-sacral and collaborative approaches, at least unilateral S3 and all of the S1-S2 nerve roots should be preserved and protected.

Gynecologists rarely encounter neural tumors. Per rectal examination must be done in unmarried females with pelvic tumors to differentiate an anterior tumor from a retro rectal one. Preoperative MRI must be used whenever there is a doubt regarding the etiology of a tumor close to the uterus or one that appears to be arising from the uterus but is pedunculated so that appropriate surgical management can be planned with the help of medical experts in the field.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles are considered in this article. The participants were informed of the purpose of the research and its implementation stages. They were also assured about the confidentiality of their information and were free to leave the study whenever they wished, and if desired, the research results would be available to them. A written consent has been obtained from the subjects. Principles of the Helsinki Convention was also observed.

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Conflict of interest

The authors declared no conflict of interest.

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