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Case Report

Giant schwannoma of the pelvis: A case report

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Abstract

Schwannomas are benign nerve sheath tumors most commonly seen in cranial and peripheral nerves. Pelvic schwannomas are particularly unusual. They are slow-growing tumors and detected incidentally. We report a giant presacral schwannoma in a 74-years-old woman who presented with urinary incontinence. Abdominal ultrasonography and computed tomography pointed out a large well-circumscribed solitary mass arising from right adnexial region to the epigastric area without any sign of ascites and bilateral hydronephrosis. Although sacrouterin leiomyoma was considered preoperative diagnosis pelvic retroperitoneal mass was detected in an operation and frozen section was made It reported mesenchymal tumor with a high probability of schwannoma. Since the tumor invade strictly the major vessels, the mass was removed by enucleation in order to not to lead further morbidity and mortality. Final pathology revealed Schwannoma.

Key Words:

Diagnosis, pelvic cavity, pelvic mass, schwannoma

Introduction

Schwannomas, benign neurogenic tumors derived from peripheral nerve sheaths, are rarely found in pelvic cavity [1]. The clinical and radiological features are not specific to the tumor, so preoperative diagnosis is challenging. Giant retroperitoneal schwannomas can lead to diagnostic dilemma since they can mimic genital tract malignancies. Pelvic schwannomas are often considered to be urologic diseases or gynecologic masses [2]. We report a case of female pelvic schwannoma located in the presacral region emphasizing its surgical difficulty due to its proximity to large vessels of pelvis. A large presacral encapsulated mass measuring 15 cm mimicking sacrouterine leiomyoma was noted by laparotomy.

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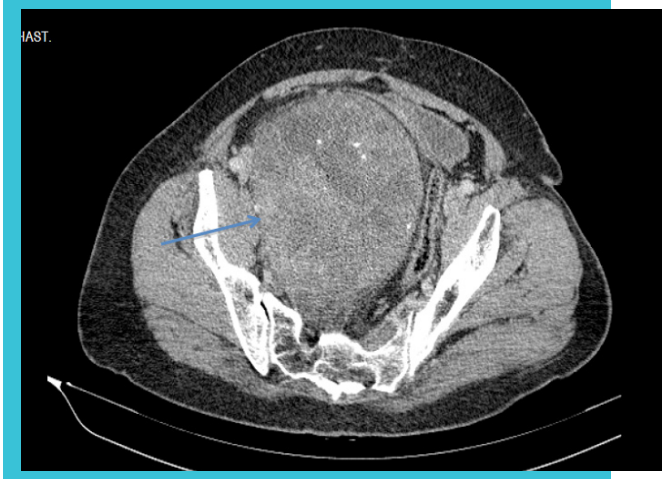
Case Presentation

A 74-year-old postmenopausal woman, presented with urinary incontinence and underwent examination in a clinic. The patient's only complaint was urinary incontinence that had been going on for one year and it was without any signs of neurovascular deficit. Her obstetric history included four childbirths, three of them natural childbirths whereas one of them was a cesarian section. Her physical examination and bimanual pelvic examination were performed there were noticed that a 15-20 cm fixed pelvic mass reaching to umbilicus. Preoperative routine laboratory analysis including complete blood count tests, urine analysis and several tumor markers such as carcinoembryonic antigen, α-feto-

protein, and CA- 19.9,CA-125 and CA-15.3 were all within normal limits. Abdominal ultrasonography revealed grade 2 hydronephrosis in left kidney, grade 3 hydronephrosis in right kidney with the suspicion of mass effect, absence of ascite, marginalized bladder due to compression and a pelvic heterogeneous well-circumscribed solitary mass extending to the umbilicus (craniocaudal measurement 153 mm, transverse measurement 117 mm) composed of cystic formations. Computerized tomography (CT) pointed out a well-circumscribed large mass arising from right adnexial region to the epigastric area without any evidence of lymphadenopathy (Figure 1). There were multiple, small calcifications within the tumor. No pathological fluid accumulation in abdominal cavity was detected. These imaging features indicated the high possibility of a benign tumor. Explorative laparotomy was carried out. The abdomen was opened through a midline incision. Genital organs, uterus, bilateral tubes and ovaries were atrophic. A large presacral encapsulated mass measuring 15-20 cm was noted and at first look it thought to be a leiomyoma originated from sacrouterine ligaments. When the exploration continued the mass was recognized that there was no relation between the tumor and the sacrouterine ligaments, but the mass was originating from sacroccocygeal area. Right common iliac and external iliac arteries with corresponding veins were stretched by the tumor. While dissecting the tumor from right common iliac vessel vascular injury occurred and repaired. A frozen section was performed during the surgery which was reported as mesenchymal tumor with a high probability of schwannoma. Since the tumor was firmly adhered to the sacrum and giant vessels of pelvis, the mass was partly removed by enucleation. The surgery duration was 3 hours and the estimated blood loss was 2.4 L. and this was replaced by 7 units of cross matched red blood cells intraoperatively. The patient's postoperative hemoglobin level was 9.4 gr/dl. Uterus and ovaries with their supplying vessels were preserved. Final pathology revealed a 15x 10x 8 cm yellow to gray-white soft tissue consistent with schwannoma. Histologically, sections showed neoplastic proliferation including spindle-shaped cells and histiocytes in large hyalinization areas with focal calcifications (Figure 2). Cystic degeneration and nuclear atypia was also observed which were associated with ancient schwannoma. Immunhistochemically, tumor cells had diffuse expression of S-100 protein but were negative for Vimentin, Actin, Caldesman, F-8 and Desmin (Figure 3). Postoperatively, the patient recovered fully on the eight day and discharged. The patient was taken to follow-up procedure.

Figure 1.



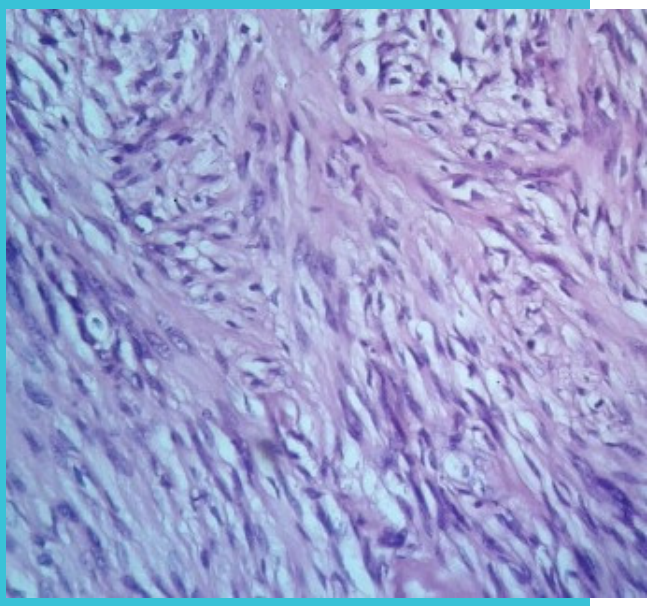
Computed tomography (CT) pointed out a well-circumscribed large mass arising from right adnexial region to the epigastric area (arrow)

Discussion

Schwannomas originating from peripheral nerve sheaths are rarely found in pelvis accounting for 0.5% of reported cases. Although they can occur sporadically, they can be combined with some genetic disease such as Von Recklinghausen disease. It affects the females usually between the ages of 20 to 50 years [3]. Schwannomas are generally benign in nature with low possibility of malignant transformation or recurrence after complete excision [4]. Malignant schwannomas act as high grade sarcomas and recurrence rates and distant metastasis are very high. Since schwannomas are asymptomatic until they reach huge size, they are diagnosed during medical investigations for unrelated symptoms [4]. They can cause chronic pelvic pain, constipation, dysuria, urinary incontinence and non specific neuritic pain [5]. In our case the pelvic schwannoma was detected incidentally while the patient was being evaluated for urinary incontinence. Due to absence of specific symptoms, preoperative diagnosis of pelvic schwannomas are challenging. Moreover, there is no distinguishing features on pelvic ultrasound, CT scans and magnetic resonance imaging (MRI) and giant size can make it difficult to detect exact origin of the tumor. Schwannomas are typically well-circumscribed with enhancement of capsule after contrast administration. MRI with gadolinium enhancement is believed to be superior to CT in

demonstrating degenerative changes and tumor origin. But radiographic studies are still inadequate in differentiating benign and malignant disease. Since in our case there was no preoperative diagnostic tool for malignancy, we did not plan any core biopsy under guidance before operation. Schwannomas can be misdiagnosed as ovarian carcinoma, ovarian cyst, psoas abscess and uterine leiomyomas [6,7]. Exact diagnosis is performed histopathologically.

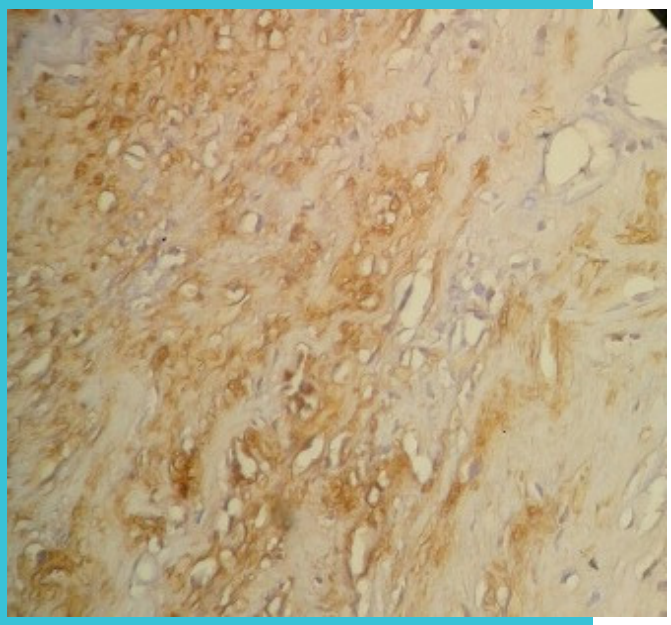
Figure 2.



Histologic features of the resected schwannoma. Spindle cells in interlacing fascicles (x200)

Macroscopically schwannomas are encapsulated, solitary and well-circumscribed tumors. When the Schwann cell nuclei are large, hyperchromic and multilobed without mitotic figures, then this is called nuclear atypia and it's a characteristic feature of ancient schwanoma. In case of large schwannoma, it may be composed of cyst formation, hemorrhage, calfication and hyalinatization indicating impaired vascular supply. Immunohistochemistry is positive for S-100 and negatif for CD-34 [8]. 100 positive immunostaining confirms the neuroectodermal origin of the tumor. Treatment is complete removal of tumor, since recurrence has reported in few patients due to incomplete surgical excision [9]. The most commonly used surgical technique is median laparotomy, since these tumors are often large in size. Care must be taken during dissection of the tumor from adjacent structures because the tumor can be in close

Figure 3.



Immunohistochemical findings. S-100 expressing in Schwann cells (x200)

proximity to the major pelvic vessels. Abandon hemorrhage may be life-threatening. In our case right common iliac vein was damaged during dissection and massive transfusion was needed. So the complete removal could not be performed. It is important that the sufficient amount of blood products must be readily available preoperatively. Laparoscopic dissection may be appropriate for these tumors as tumors are generally solitary, encapsulated and do not invade nearby structures providing easy dissection and moreover, magnification of anatomic view may facilitate the excision by laparoscopy [10]. However, many pelvic schwannomas are incidentally diagnosed, laparotomy is considered initially for pelvic mass. In summary, this case report describes a large presacral schwannoma in the pelvic cavity. Preoperative evaluation was insufficient to diagnose schwannoma due to lack of unique features. Definitive diagnosis was made histopathologically. Although incidence is rare, in the evaluation of pelvic masses schwannomas should be considered in differential diagnosis.

Acknowledgement
None

Declaration of Interest
None

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