

Presacral Schwannoma

Schwannomas are rare neoplasms arising off Schwann cells. Most of them are benign. Acoustic schwannoma is the most famous tumor of the family. Retroperitoneal schwannomas form 1-5% of all retroperitoneal primary tumors. Pelvic schwannomas consist 1% of all schwannomas. Thus, they are coming too far after the head, the neck, and the upper extremities ones. Nevertheless, there are only 21 cases of presacral schwannoma reported in the english medical literature.

Case report

A 40-year-old male patient presented with a history of both Crohn's disease and left ureteral calculi. Crohn's disease dates to 2 years ago. Whereas, the left ureteral calculi were first put in evidence 2 months ago. It was during the ultra sound screening when a pelvic mass was seen occupying the presacral space.

The ultra sound study was undetermined about the nature of the mass. In contrast, CT scan with contrast infusion showed a round well- demarcated mass, measuring 8 cm, occupying the presacral space, deviated a little bit to the right of the midline, pushing forward the other pelvic organs (rectum, bladder). The mass was inhomogeneous in taking the contrast. [Figures \(1\) & \(2\)](#).

The decision was made to totally remove the tumor via anterior midline approach. Supported by the clear contour of the mass, the integrity of the sacrum, the absence of any neural signs and symptoms, I did not expect any difficulty in operating on.

Smoothly run out the time of the operation. Two hours and the mass has been removed by enucleation, and been outside on a lateral table. Blood loss was less than 100 ml. The patient has discharged the second postoperative day; [Figure \(3\)](#).

Contrary to all preoperative differential diagnosis, the pathologic analysis of the resected specimen was "benign schwannoma"; [Figure \(4\)](#).

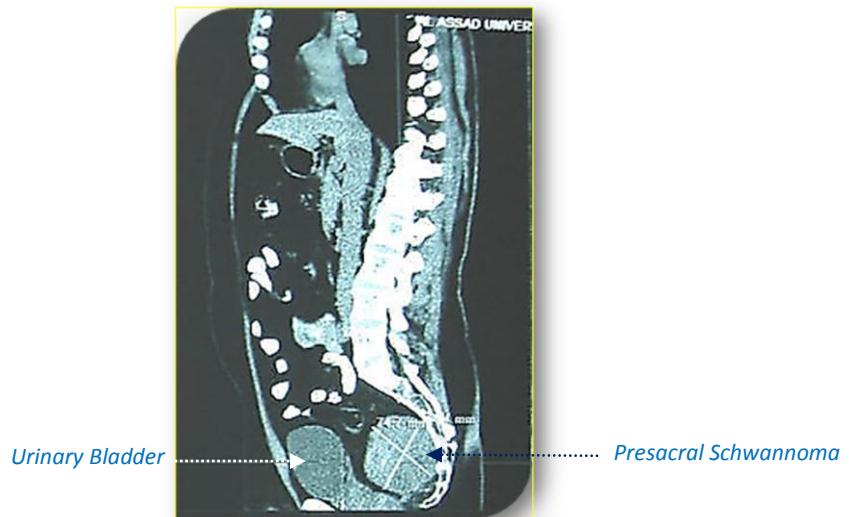


Figure (1)
CT- Scan with Contrast Infusion
Sagittal Section

Round well demarcated mass in the presacral space, measuring 8 cm, and pushing forward the bladder.

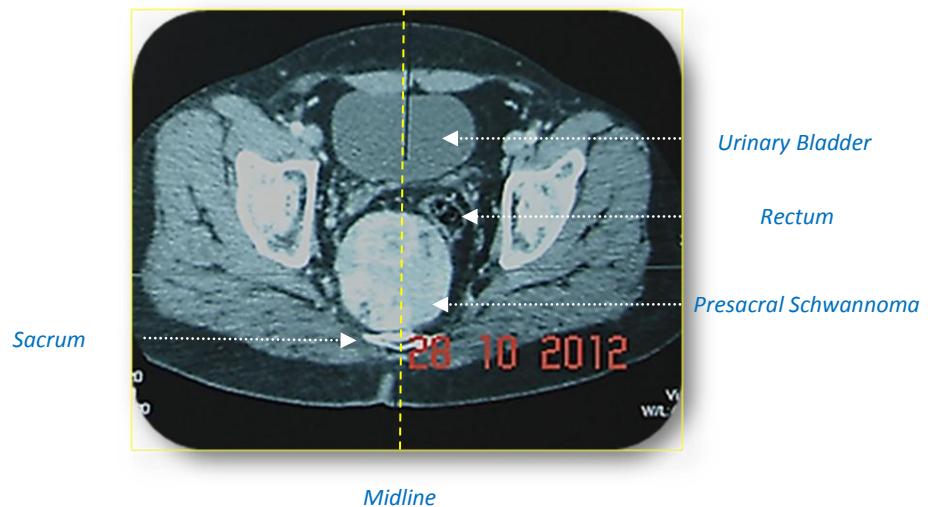


Figure (2)
CT- Scan with Contrast Infusion
Axial Section

The mass is inhomogeneous in taking the contrast. Note how the mass is deviated from the midline of the sacrum to the right. The rectum is pushed away to the left- anterior side.

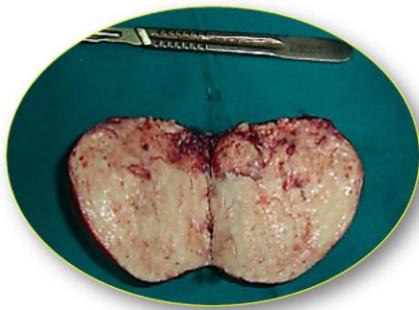


Figure (3- b)
Per- Operative View

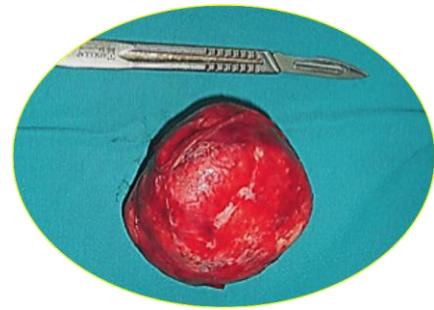


Figure (3- a)
Per- Operative View

Figure (3)
The Resected Mass, Macroscopic View
Per- Operative Views

Figure (3- a): The tumor has spherical shape with two polar elongations. It has elastic rather than hard nature.

Figure (3- b): Cut surface of the resected mass shows degenerated cystic structures and hemorrhagic foci.

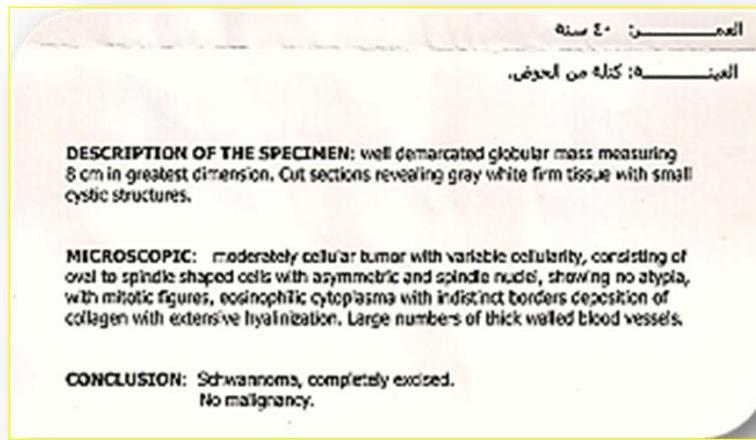


Figure (4)
Pathology Report

Benign schwannoma totally excised.
There also were cystic changes revealing the old age of the tumor.

Discussion:

Schwannomas are rare benign neoplasms arising from the nerve sheath. Some cases of malignant transformation have been reported. In fact, these were a transformation from plexiform neurofibromatosis rather than a malignant degeneration of previously exist benign schwannomas.

Schwannomas mostly occur in the head, neck, and the upper extremities. Acoustic schwannoma is the most popular member of the family. Retroperitoneal schwannomas are very rare; no more than 133 reported case in the english medical literature, Takatera et al. Retroperitoneal schwannomas consist 1-5% of all primary retroperitoneal tumors. Nevertheless, just 21 reported cases, mostly in women, of presacral schwannomas are described in the medical literature.*

Pelvic retroperitoneal schwannomas are classified as sacral, dumb-bell, and presacral schwannoma. Sacral schwannomas are completely confined to the sacrum. Dumb-bell schwannomas have two components; one intra-sacral whereas the other extra- sacral. Presacral schwannomas are very rare tumor entirely located in the presacral space.

Presacral schwannoma gets very large size before being detectable. In spite of its origin from the nerve sheath, it seldom causes functional disorders. The slow growth of the tumor gives the neighboring nerves the opportunity to compensate the functional loss caused by the tumor impaction on the mother nerve.

There are no specific symptoms. The most presenting symptoms are those related to the rectum, bladder, nerves, compressed by the tumor mass. Most often, the tumor is discovered incidentally with rectal examination or while doing radio investigations to the pelvis for another reason.

CT, MRI, are the radio of choice. MRI has the privilege of better showing the texture, internal cystic degenerations, location, size, and the relationship of the tumor with the soft neighborhoods. CT scan superimposes in delineating the bone- tumor relationship. The presence of calcifications and cystic degenerations within the tumor are synonym to old age. Finally, the tumor location outside the midline is considered good radio sign to differentiate it from chordoma which quite respect the midline of the sacrum.

Since benign schwannomas are encapsulated tumor, total excision of the tumor is the treatment. However, piecemeal excision of the tumor is another surgical choice whenever total excision of the tumor is technically difficult, and harbors great risk of bleeding or nervous compromise. Such cases are expected each time the tumor invades the sacrum. There is no adjuvant therapy even with partial excision. Reoccurrence rate is 11% in total excision, over 50% in partial excision.

* lin CM, Kao CC, Lin TC, Cha TL, Wu ST. Giant presacral schwannoma mimicking malignancy in a man. *Acta Chir Belg* 2010;110:387-389.

In another context, one can also read:

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