

CASE REPORT

Retrorectal Schwannoma

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ABSTRACT

Schwannoma is a benign encapsulated nerve sheath tumor. These tumors are more frequently located in the head, neck, extremities, and trunk. Retroperitoneal pelvic localization of schwannoma accounts for 0.5 to 5% of all cases, while the incidence of retrorectal tumors is estimated at 1 in 40,000 to 63,000 cases in the general population, which we report here.

Keywords: Retrorectal tumors, Schwannoma, Surgery.

How to cite this article: Alhomoud H. Retrorectal Schwannoma. *World J Lap Surg* 2016;9(1):44-46.

Source of support: Nil

Conflict of interest: None

CASE REPORT

A 50-year-old male presented with a feeling of heaviness in the pelvic floor associated with change in bowel habits and tenesmus for the last 5 years.

On digital rectal examination, an irreducible firm mass posteriorly on the right side was felt. Mucosa was intact. Rigid sigmoid scope up to 20 cm was normal. Routine laboratory tests and tumor markers were within normal limits. Computed tomography (CT) of the chest, abdomen, and pelvis showed a large right lower pelvic mass, whose nature was not clear (Fig. 1). Magnetic resonance imaging (MRI) of the abdomen and pelvis showed a large right presacral mass, whose appearance suggested a neurogenic tumor (Fig. 2). Endorectal ultrasound was done which showed a mass suggestive of sarcoma or a duplicated cyst (Fig. 3).

The patient was operated on April 2, 2012, through an arcuate incision in the right buttock. The patient was placed in the prone jack-knife position. An oval well-circumscribed encapsulated mass 10 × 8 cm was removed.

Histology revealed the presence of compact spindle cells arranged in short bundles and a peripheral lymphoid cuff with some germinal centers (Fig. 4). At



Fig. 1: Computed tomography of the chest, abdomen, and pelvis showing a large right lower pelvic mass, whose nature is not clear

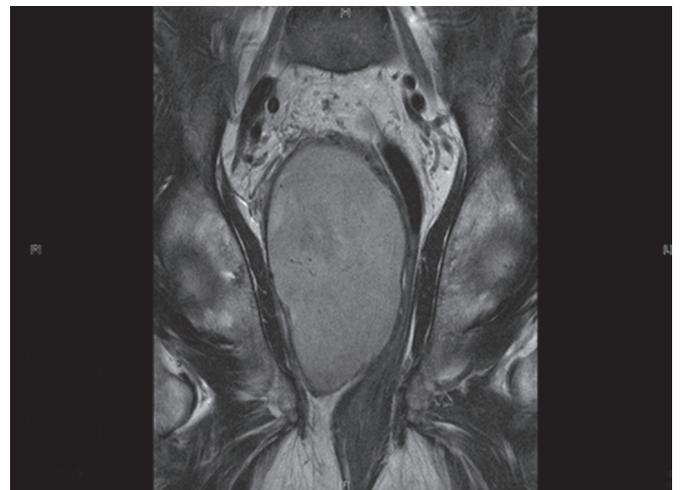


Fig. 2: Magnetic resonance imaging of the abdomen and pelvis showing a large right presacral mass, whose appearance could suggest a neurogenic tumor

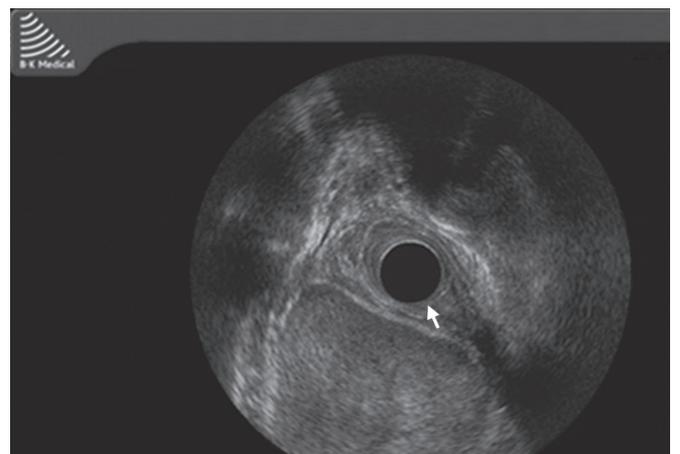


Fig. 3: Endorectal ultrasound (showing a mass suggestive of sarcoma or a duplicated cyst)

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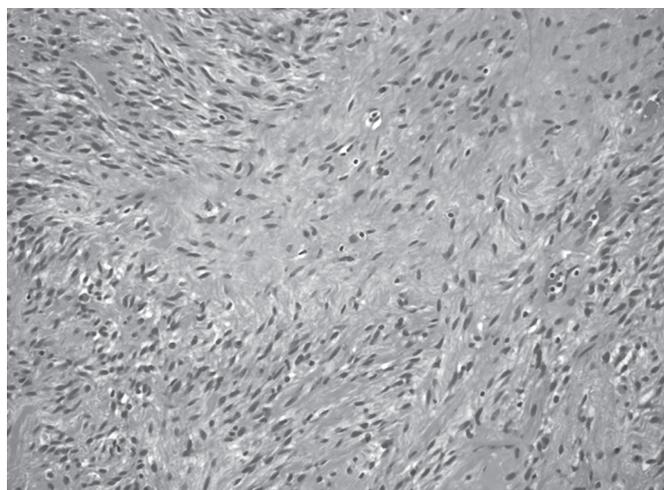


Fig. 4: Histology revealing the presence of compact spindle cells arranged in short bundles and a peripheral lymphoid cuff with some germinal centers

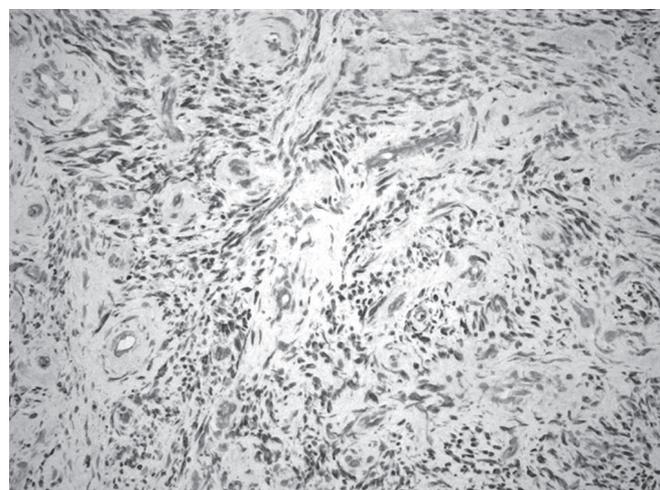


Fig. 5: At immunohistochemistry, the spindle cells were positive for S-100 protein and negative for α -smooth muscle actin and CD34

immunohistochemistry, the spindle cells were positive for S-100 protein and negative for α -smooth muscle actin and CD34 (Fig. 5).

The histological and immunohistochemical features were compatible with a diagnosis of schwannoma.

The patient's recovery was uneventful and he was discharged on postoperative day 4.

DISCUSSION

Schwannoma is a benign encapsulated nerve sheath tumor arising from Schwann cells. These tumors are more frequently located in the head, neck, extremities, and trunk.¹ Retroperitoneal pelvic localization of schwannoma accounts for 0.5–5% of all cases.² The incidence of retrorectal tumors is estimated at 1 in 40,000 to 63,000 cases in the general population.³ A few large series provide an estimate of the overall incidence of retrorectal tumors in the general population and retrorectal schwannoma (Table 1). Schwannomas are slow-growing lesions that can reach a large volume without any symptoms for years if they are located in a place with large capacity like the presacral region.¹¹

Presacral or retrorectal virtual space is limited from behind by the presacral fascia and in the front by the fascia propria of the rectum. Reflection of the pelvic

peritoneum is its upper limit. Waldeyer fascia is the lower space which separates it from the supralelevator space. Ureters, iliac vessels, and sacral nerve roots constitute its lateral limits.⁶

Retrorectal schwannoma may present with perirectal pain, change in defecation habits and sensation of incomplete evacuation, obstructed defecation, and tenesmus.

Preoperative diagnosis of retrorectal schwannoma is challenging. In a study published in January 2012 in *ColoRectal Disease Journal* by Macafee, 56 patients underwent excision of retrorectal tumors between 2002 and 2010 (11 cases were schwannomas), with MRI and CT done on all patients. Results showed that preoperative MRI is vital to make the correct diagnosis between benign disease and malignancy and the feasibility of tumor resection.¹⁰

Histological examination including immunohistochemistry can give the exact diagnosis of schwannoma. The two histological growth patterns are Antoni A and Antoni B. In Antoni A type, there is dense growth of fusiform cells, compactly arranged in palisades to form Verocay bodies. In Antoni B, the fusiform cells are more loosely distributed with rounded or elongated nuclei, with a greater quantity of myxoid stroma and xanthomatous histiocytes.¹²

Table 1: Incidence of retrorectal masses and retrorectal schwannoma

Authors	Institution	Length of study (years)	No. of cases	Schwannoma
Jao et al ⁴	Mayo Clinic	19	120	7
Sean et al ³	Washington University Hospital	22	34	5
Buchs et al ⁵	University Hospital of Geneva	9	16	1
Canelles et al ⁶	University Hospital Spain	13	20	3
Chang et al ⁷	Memorial Hospital Taiwan	13	14	3
Kye et al ⁸	Catholic University of Korea School of Medicine	9	15	4
Strupas et al ⁹	University Medical School Lithuania	13	14	1
Macafee et al ¹⁰	General Hospital, Leeds, UK	9	56	

The use of immunohistochemical panels plays a fundamental role in the diagnosis of schwannomas and in ruling out other neoplasms of mesenchymal origin. Using anti-CD34 antibodies, desmin, cytokeratins (AE1/AE3), cKit, chromogranin, and S-100 protein.

Total resection is the main therapeutic treatment of retrorectal schwannoma, using transabdominal or retrorectal approach, open or laparoscopy.¹³ All lesions below the middle of S3 without sacral, pelvic sidewall or visceral involvement were excised using the perineal approach. All lesions above S3 were excised by means of an abdominal approach.¹⁴

The rate of recurrence after complete resection is rare,¹⁵ but it may reach 10 to 54% in incomplete resection.

CONCLUSION

In conclusion, schwannoma can occur anywhere on the peripheral nerve. Retrorectal schwannoma is quite rare, preoperative MRI is vital to make the correct diagnosis, and use of immunohistochemical panel is important to achieve a definitive histopathological diagnosis. Surgical resection with free margins is the best treatment.

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