CASE REPORT OF A RARE RETROPERITONEAL PELVIC SCHWANNOMA

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Summary

Schwannoma is a tumor arising from peripheral nerve sheath, found very rarely in the retroperitoneal part of the pelvis. It can reach large proportions before causing symptoms due to mass effect and is mostly diagnosed incidentaly.

We describe a pelvic retroperitoneal Schwannoma in a 67-year-old man presented with progressive pain in lower left abdominal quadrant with digestive discomforts, which lasted for three months. Patohistological examination after complete surgical excision revealed a Schwannoma.

KEY WORDS: *schwannoma, retroperitoneal pelvic schwannoma, symptoms, diagnosis, treatment*

PRIKAZ SLUČAJA RIJETKOG RETROPERITONEALNOG SCHWANNOMA

Sažetak

Schwannom je tumor koji potječe iz ovojnice perifernog živca. Retroperitonealni Schwanom smješten u zdjelici je izuzetno rijedak. Doseže velike dimenzije prije uzrokovanja simptoma zbog čega se obično slučajno dijagnosticira.

Opisan je prikaz slučaja retroperitonealnog Schwannoma zdjelice u 67-godišnjeg muškarca s progresivnim bolovima u donjem lijevom kvadrantu abdomena i probavnim smetnjama, u trajanju od tri mjeseca. Kirurški je tumor potpuno odstranjen i patohistološka dijagnoza je Schwannom.

KLJUČNE RIJEČI: *schwannom, retropertionealni schwannom u zdjelici, simptomi, dijagnostika, terapija*

INTRODUCTION

Schwannoma is a benign nerve sheath tumor composed entirely of well-differentiated Schwann cells. It arises as solitary tumor from the peripheral nerves of the face, neck, body, extremities and the retroperitoneal region. Malignant form is very uncommon.

Retroperitoneal location is exceedingly rare and it's incidence is only 0.7 - 2.7% (1). Retroperitoneal Schwannoma is usually located in abdomen, while less than 30% of cases are located in the pelvis (2).

Schwannoma is, according to WHO (World Health Organization) Classification of tumors graded as conventional, non-melanotic, cellular,

plexiform and melanotic Schwannoma. It mostly presents itself as a fusiform, round or oval mass that is normally sharply circumscribed and encapsulated, and causes symptoms by compressing adjacent structures without invading them.

Peak of incidence is between fourth and sixth decade of life with a slight gender predilection towards women (3:2) (3). The delay between onset of clinical manifestations and the diagnosis could be up to two years. The size of retroperitoneal Schwannoma at the time of diagnosis is usually more than 8 cm in diameter (2).

Main symptoms are abdominal distension (30.5%) and abdominal pain (20.7%), lower back pain (6.1%) with/out digestive problems (6.1%).

Most of these tumors are found incidentally during diagnostic imaging (sonography, CT of MRI) indicated for other health reasons. Only in 15.9% of cases a correct preoperative diagnosis was made by either ultrasound-guided biopsy, computed tomography scanning or magnetic resonance imaging (2).

Lack of specific symptoms and their overlapping makes it hard to differentiate retroperitoneal Schwannoma from other retroperitoneal tumors. Moreover, retroperitoneal Schwannomas are rare and vary in presentation. Therefore, misdiagnoses are common. Preoperatively, they are often mistaken for genital or abdominal mesenchymal tumors (e.g. malignant fibrous histiocytoma, malignant peripheral nerve sheath tumor, liposarcoma, synovial sarcoma, hemangiopericytoma or leiomyoma), or intestinal tumors.

The proposed treatment is complete surgical resection with sparing of the nerve and blood supply, or observation in asymptomatic patients.

Hereafter we report a case of retroperitoneal pelvic Schwannoma with typical lack of specific symptoms.

CASE REPORT

A 67-year-old man was admitted for examination because of progressive pain in lower left abdomen accompanied with digestive discomforts, lasting for three months. He had no comorbidities and no significant history of diseases in family.

Colonoscopy revealed a deformation of rectal lumen seeming to be caused by external compression. Subsequently, MRI depicted a multilocular solid cystic mass with uniform smooth wall (Figure 1), measuring 8,6 x 7 cm situated in lower pelvis, compressing the left bladder contour and rectosigmoid colon, in close relation to left internal obturator muscle and left internal iliac artery and vein, but without signs of nearby infiltration.

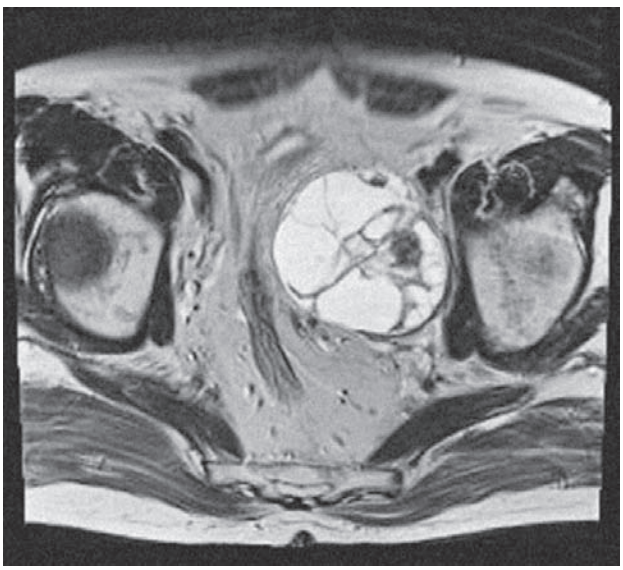

Figure 1. MRI (T2-weighted image) Multilocular cystic mass with uniform smooth wall on the left side of pelvis under the left common iliac artery.

Digital subtraction angiography of pelvis showed no pathological vascularization of the lesion.

Preoperative differential diagnosis was well-circumscribed solid cystic lesion of benign origin, less likely central necrotic mesenchymal tumor with lack of significant pathologic vascularization.

Due to the progression of symptoms and the need to determine the identity of the pelvic mass, surgical treatment was indicated.

After midline abdominal incision, the abdomen was explored and pelvic tumor mass visualized. Tumor was adjaced to left distal common iliac and proximal internal iliac artery and vein, looking like dilatation of common iliac artery, measuring about 8 x 7 cm. Vascular surgeon resected part of distal common iliac and proximal internal iliac artery and isolated the tumor just proximate to capsule, showing intact internal vein,

left genitofemoral and obturator nerves and urether.

Macroscopically the specimen was a gray and yellowish tumor with a capsule (Figure 2), and when cut in half there were cystic cavities and hemorrhage (Figure 3).

The tumor was pathohystologically classified as classical benign Schwannoma composed of Schwann cells with occasional nuclear palisading (Antony A pattern) and areas of Schwann cells in loose connective tissue (Antony B pattern). Blood vessels were thick-walled and hyalinized surrounded by hemorrhage and hemosiderin. Immunohistochemically tumor cells were well positive to S 100 and NSE.

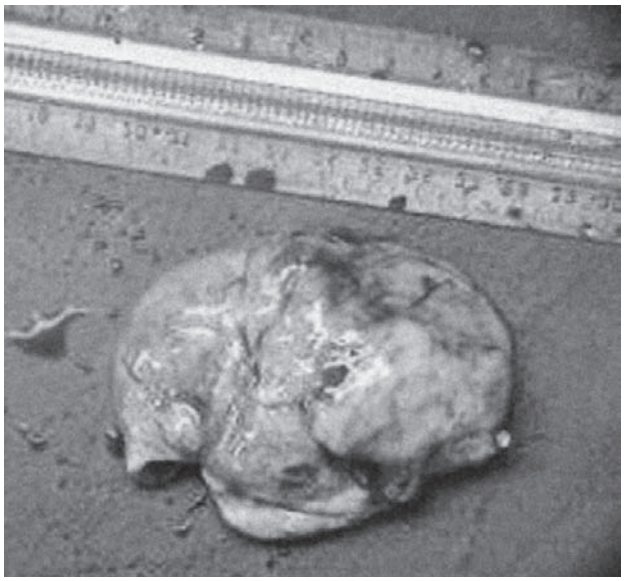

Figure 2. Resected encapsulated tumor, measuring 8.5 x 6.5 x 5.3 cm

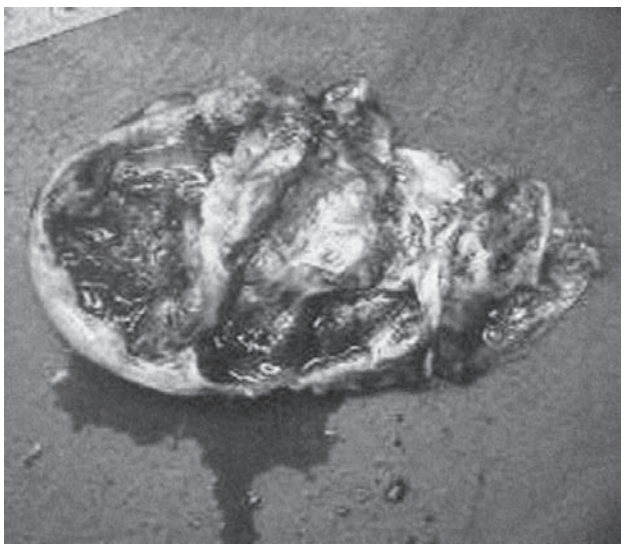

Figure 3. Tumor cut in half. Tissue is partially composed of cystic cavities filled with soft red and brownish tissue tissue.

The patient's postoperative course was uneventful, he reported withdrawal of all symptoms (abdominal pain and digestive discomforts) and was discharged a week after the surgery. Regular follow-up was scheduled 6 months after the operation.

DISCUSSION

A rare case of pelvic retroperitoneal localization of Schwannoma is presented. Since it is a very rare localization (0.7 - 2.7%) and specific symptoms are often missing, the correct preoperative diagnosis is very doubtful. The fact that just a few reports were published about the radiologic features of Schwannoma in this region makes it even more difficult to reach.

Retroperitoneal tumors present with late symptoms, when the tumor grows big enough to produce local mass effect on adjacent structures and compromise their function, by causing neurological symptoms due to nerve compression or abdominal pain due to compression on intestinal or urogenital organs. This was seen in our case, tumor measured 8.5 x 6.5 x 3.5 cm and caused progressive pain in lower left abdomen accompanied with digestive discomfort due to external compression .

Only in 15.9% of cases a correct preoperative diagnosis was possible, set by one of the procedures like: ultrasound-guided biopsy, computer tomography scanning or magnetic resonance imaging. Tumor mass is usually found incidentally during imaging procedures for some other reasons, and it can easily be misinterpreted (2).

Imaging findings of retroperitoneal Schwannoma and soft tissue mesenchymal tumors are sometimes similar and hard to differentiate. Schwannomas with degenerative changes can easily be misdiagnosed as soft-tissue neoplasm. Especially in case of mature Schwannomas, where during long time needed for their development, degenerative and diffuse hypocellular areas and areas of hemorrhage have formed (degenerative

changes are typified by perivascular hyalinization, calcification, cystic necrosis, relative loss of Antoni type A tissue, and degenerative nuclei that may be misinterpreted as sarcomatous pleomorphismus) (4).

MRI is the most useful technique for evaluation of Schwannoma, which appear as a well-defined, complex cystic mass. The cystic component is shown as a low-signal intensity area on T1-weighted images and a high-signal-intensity area on T2-weighted images. A target like pattern consisting of peripheral high signal intensity (representing Antoni B areas) and central low signal intensity (representing Antoni A areas) may also be seen on T2-weighted images. Areas of myxoid and hemorrhagic change may also be seen within the tumor (4, 5, 6).

In our case we had no need for another imaging methods since the MRI was enough to indicate a surgical treatment**.** CT scanning has no advantage over MRI in distinguishing these two entities and is rarely performed in primary soft-tissue tumors. Rare exceptions are patients with a contraindication for MRI. In such patients CT images of Schwannoma show smooth and sharp margins with an enhanced appearance and often with liquefaction, necrosis and hemorrhage within the tumor (7).

Despite strait forward preoperative diagnosis, intraoperative diagnosis was inconclusive. Pelvic mass looked like a dilatation of distal common left iliac artery, was suspicious of a pseudoaneurysm and procedure was carried out in such manner. However, soft tissue neoplasm could not be excluded because on MRI images there was a tendency to confuse Schwannoma with malignant mesenchymal tumor. Aggressive features of a soft-tissue neoplasm included size greater than 5 cm; it's deep location and absence of central enhancement, which was suggestive of necrosis. A pseudocapsule or capsule is commonly seen around a mesenchymal tumor and in malignant Schwannomas.

Heterogeneity and calcifications are seen in Schwannomas and its malignant counterpart. In our case the retroperitoneal mass bigger than 5 cm with focal necrosis and hemorrhage was very suspicious for soft tissue neoplasm (8).

The final diagnosis of Schwannoma was based on pathohistological results, where the cells are strongly positive for S-100 protein.

Histopathologic biopsy is necessary for definite diagnosis and management of intra abdominal and retroperitoneal tumors. Percutaneous ultrasonography or CT guided biopsy could be a method of choice for definite diagnosis. If the tumor is difficult to approach, conventional laparoscopy or explorative laparotomy should be performed.

Benign Shwannoma carries a very good prognosis. The most frequent complication is recurrence, probably due to incomplete excision, with a recurrence rate of 5-10% (9). Regular follow up is necessary, especially after incomplete surgical resection. In case of incidental founding in asymptomatic patient surgical treatment may be postponed, if definite diagnosis was reached. Benign nature and slow growth of the tumor question the necessity of immediate surgical treatment. Obtaining the definitive diagnosis by guided biopsy and regular follow up may be an option in some cases. However, if symptomatic, as in our case, surgery remains the preferred treatment of choice.

CONCLUSION

A retroperitoneal Schwannoma is rare and correct preoperative diagnostic is difficult to make. It is slow growing tumor, may produce vague local symptoms, and is usually diagnosed incidentally. Increased incidence of Schwannomas is expected due to more precise diagnostic imaging and its availability. MRI has an important role in diagnosing of retroperitoneal Schwannoma. When causing symptoms retroperitoneal Shwanoma has to be resected. In all other patients regular follow up may be an alternative.

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