Clinical Case Report

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A giant ancient schwannoma mimicking an adnexal mass

Case report

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Abstract

Introduction: Ancient schwannoma is a rare tumor of the peripheral nerve sheath. As degenerative properties are defined histologically, it can be wrongly interpreted as malignant.

Case presentation: The case presented here is of a giant ancient schwannoma with a pelvic retroperitoneal location, which was mimicking an adnexal mass.

Conclusion: In the rarely seen cases in the retroperitoneum, it may reach very large dimensions.

Abbreviation: MRI = magnetic resonance imaging.

Keywords: adnexal mass, ancient, giant schwannoma, retroperitoneal region

1. Introduction

Ancient schwannoma is a rare variation of peripheral nerve sheath tumors. The term "ancient" is used because of the degenerative features that cause a reduction in blood supply over time as the tumor grows, which are defined histologically and often lead to an incorrect diagnosis of malignancy.[1] These tumors, which grow extremely slowly, are generally diagnosed incidentally. They are seen more often in females between the 2nd and the 5th decades of life.[2] Schwannomas generally have an intracranial location and are rarely seen in the retroperitoneal region. In cases of pelvic retroperitoneal location, they can mimic an adnexal mass.[3,4] The case presented here is of a giant ancient schwannoma with a pelvic retroperitoneal location, which was mimicking an adnexal mass.

1.1. Case report

A 36-year-old female presented at the polyclinic with complaints of right lower quadrant pain and extended menstruation periods which had been ongoing for 1 year and a noticeable hardness in the same place in the abdomen in the last month. The physical examination revealed a fixed, hard mass of 10 cm diameter which was palpated in the right lower quadrant of the abdomen. In the abdominal ultrasonography, a multilocular cystic mass of 14×12×9 cm size, containing many septa, and with thick walls was observed in the right half of the pelvis (Fig. 1). On the Doppler examination, the cyst walls and septa were seen to be vascular. On the abdominal magnetic resonance imaging (MRI), a cystic mass of 15×10×9 cm with regular borders and containing internal septations was determined, located between the iliopsoas muscle and the abdominal anterior wall islets on the pelvis upper right side extending as far as the level of the right kidney lower pole. Heterogeneous contrast substance involvement was seen in the wall and septa of the mass. A clear evaluation could not be made of the relationship with the adnexal structures. In the laboratory examination, CEA, CA-125, and CA19–9 tumor markers were found to be within normal limits.

The patient was admitted for surgery with initial diagnoses of cystic tumors originating from right adnexal structures, hydatid cyst, and appendix mucinous cystadenoma, and in the exploration, a capsular mass 15×12×10 cm was determined in the pelvic retroperitoneal area, unrelated to the adnexal structures. The mass, which was smooth-surfaced and showed fluctuation with palpation was excised en bloc (Fig. 2). The patient was discharged problem-free on the postoperative 4th day. In the histopathological evaluation, it was macroscopically reported that when the mass was opened, hemorrhagic fluid was emptied, and the greater part of it was multilocular containing a solid area of cartilage hardness, dirty white in color and of 5×2×5 cm size. Microscopically, on a myxoid stroma base of observed intense hyalinization, a lesion was observed formed of cells with a quite large pleomorphic nucleus located in hemorrhagic areas and pigment-laden histiocytes in focal areas containing scattered cystic cavities (Fig. 3). In the immunohistochemical examination, diffuse positive staining with S-100 protein was observed (Fig. 4). It was reported as an ancient schwannoma which was showing findings of degeneration. At the 1-year follow-up examination, no lesion was observed on computed tomography. Patient informed consent was obtained.

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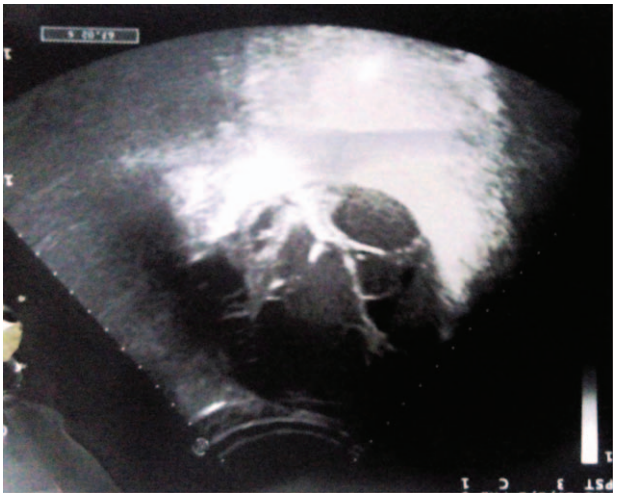

Figure 1. Ultrasonographic appearance of the multilocular cystic mass.

2. Discussion

Schwannomas are benign neurogenic soft tissue tumors originating from peripheral nerve sheaths. They are seen most often in the head and neck region and in the extremities.[1] Although retroperitoneal schwannomas comprise 0.7% to 2.6% of all schwannomas, those with a pelvic location comprise <1%. The majority of pelvic schwannomas originate from the sacral nerve or the hypogastric plexus.[5] Although schwannomas are generally <5 cm in diameter, those with a retroperitoneal location may reach a much greater size with slow growth over a long period.[2,6]

Ancient schwannomas are a rare variant of schwannomas, forming 0.8% of soft tissue tumors.[1,7] They are characterized by distinguishing degenerative tumor properties including cystic necrosis, stromal edema, xanthomatous change, fibrosis, perivascular hyalinization, calcification and pleomorphism in the nucleus, lobulation, and hyperchromasia.[8] Areas of degeneration are caused by vascular insufficiency which forms over time as the tumor grows.[1,2,8] Despite these degenerative features, the behavior of ancient schwannoma does not change. It is a slow-growing benign tumor which rarely shows malignant transformation.[1]

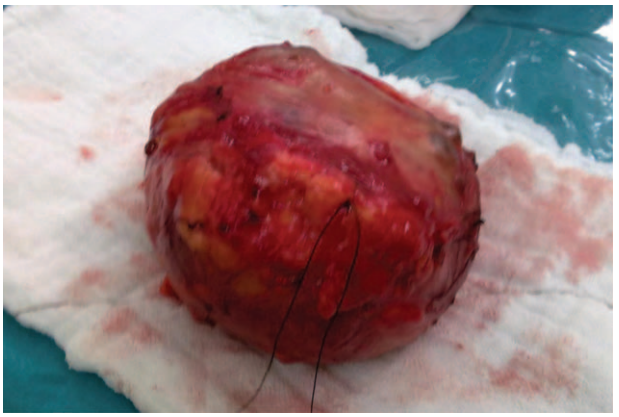

Figure 2. Giant ancient schwannoma.

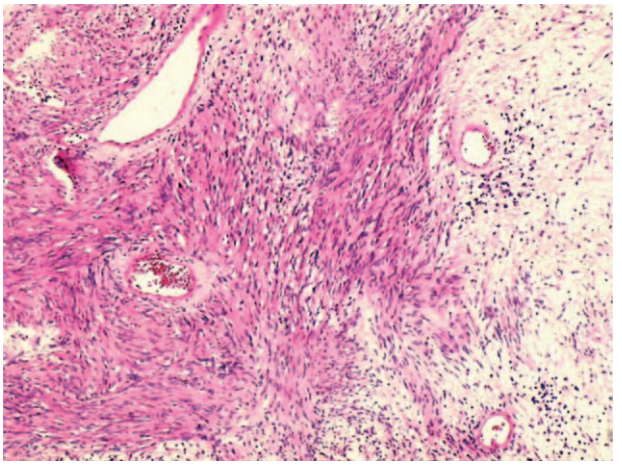

Figure 3. Classical pattern of schwannoma with spindle cells on the left side along with myxoid change with lymphocytic infiltration on the right side (H&E ×50).

The symptoms associated with retroperitoneal schwannomas are nonspecific and are generally related to pressure on adjacent structures. The most common symptom is blunt abdominal pain accompanied by abdominal distension.[9] Although ultrasonography, CT, and MRI reveal the features of schwannoma, they are not sufficient for a definitive preoperative diagnosis. In literature, schwannomas have been reported to have been determined with these tests at the rate of 15.9%.[5] Because of areas of cellular pleomorphism and degeneration in the histopathological evaluation of biopsies taken from the mass with fine needle aspiration, it can be incorrectly interpreted as malignant. Therefore, an accurate diagnosis can only be made from the evaluation of operative material.[1,5,9] S-100 protein positivity in the immunohistochemical evaluation confirms the diagnosis. As radiographic findings are nonspecific and in females masses are often observed of gynecological origin, diagnosis is made more difficult.[9] Because of the possibility of malignant degeneration, complete surgical resection is the best form of treatment and recurrence will not develop afterward just like all the gastrointestinal pathologies.[5,6,9–12] In cases of preoperative diagnosis, laparoscopic excision can be made.[5]

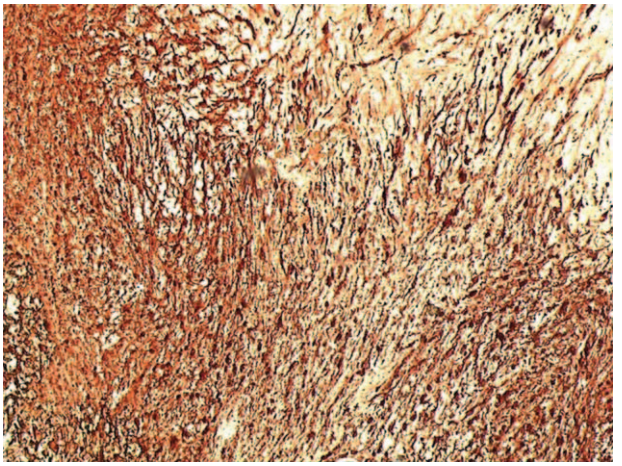

Figure 4. Tumor cells demonstrating diffuse S-100 positivity (IHC ×50). IHC = immunohistochemical.

The patient presented here was of reproductive age and had complaints of lengthy menstruation, abdominal distension, and intra-abdominal cyst. The initial diagnosis after ultrasonography and MRI was of a cyst originating from the adnexal structures. As the cyst was in the retroperitoneal area, it had reached a diameter of 15 cm and was totally excised. The diagnosis of ancient schwannoma was made with histopathological examination, which was then confirmed with S-100 protein diffuse-positive staining in immunohistochemical examinations. Throughout the follow-up period, no recurrence was seen.

3. Conclusion

Ancient schwannomas are capsular benign tumors with degenerative properties which can reach large diameters in the retroperitoneum. In females of reproductive age, it should be kept in mind as a rare event in the differential diagnosis of adnexal masses.

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