Journal of Kerman University of Medical Sciences
2022; 29(2): 183-187
http://jkmu.kmu.ac.ir/
10.22062/JKMU.2022.91898
Case Report

Retroperitoneal Ancient schwannoma: A Very Rare Case Report

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ABSTRACT

Background: Ancient schwannoma is an uncommon neoplasm originating from the peripheral nerve sheaths. Ancient schwannoma occurs mostly in head and neck region and its occurrence is very rare in retroperitoneal (about 0.7% to 2.6% of all schwannomas), and despite its benign nature, it can show malignant behaviors and it may be misdiagnosed with sarcomas in some cases. There is nonspecific clinical symptoms and radiologic findings. Evaluation of histology and immunochemistry is necessary for diagnosis, and complete surgical resection is the treatment of choice for schwannoma.

Case Report: we report a unique case of retroperitoneal ancient schwannoma in a 50-year-old man with complaint of severe pain in the right flank area for 3 days ago, which was diagnosed with ancient schwannoma after evaluation of the histopathologic and immunohistochemical findings. After surgery, the patient was kept under regular follow-up since last 15 months without any evidence of recurrence.

Keywords: Ancient schwannoma, Retroperitoneal, Nerve sheaths

Citation: Abdal K, Hafezi Ahmadi MR. Retroperitoneal Ancient schwannoma: A Very Rare Case Report. Journal of Kerman University of Medical Sciences 2021; 28(5): 183-187. doi: 10.22062/JKMU.2022.91898

Received: 30.04. 2021
Accepted: 09.08. 2021
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Published by Kerman University of Medical Sciences

Introduction

Schwannoma is usually a benign neurogenic soft tissue tumors arising from schwann cells of the peripheral nerve sheath (1). Although schwannomas are benign tumors but about 5% to 18% of malignant cases has beenassociated with Von Recklinghausen's disease (2). Ancient schwannomas (AS) are a very rare type of schwannomas, forming 0.8% of soft tissue neoplasms, which was first described by Ackerman and Taylor in 1951 (3). Ancient schwannomas are characterized by degenerative changes in tumor such as hemorrhage, hemosiderin deposits, inflammation, fibrosis, calcification, cystic necrosis, and pleomorphism in the nucleus (4). Despite degenerative changes in AS, it is a benign tumor which rarely demonstrates malignant transformation (2). Generally, Schwannoma are seen during the ages of 40-60 years and is more predominant in females than males (5). Schwannoma, also known as neurilemmoma, which is usually seen on the flexor surface of the head, neck, and extremities, are extremely rarely seen in the retroperitoneal region (6). About 0.3-3.2% of all schwannomas, are exhibited in the retroperitoneal area (3).

Here, we reported a patient with retroperitoneal ancient schwannoma.

Case Report

The patient was a 50-year-old man who referred to Imam Khomeini Hospital, Ilam, Iran, with complaint of severe pain in the right flank area for 3 days ago. The patient had no past medical history and trauma. Analysis of the laboratory blood and urine tests showed no abnormal findings. Clinical examination showed a palpable mass in the right flank region. The chest X-ray was normal.

A computed tomography scan (CT) confirmed a round-oval, well-defined mass with cystic and necrotic central region in the right kidney (Figure 1).

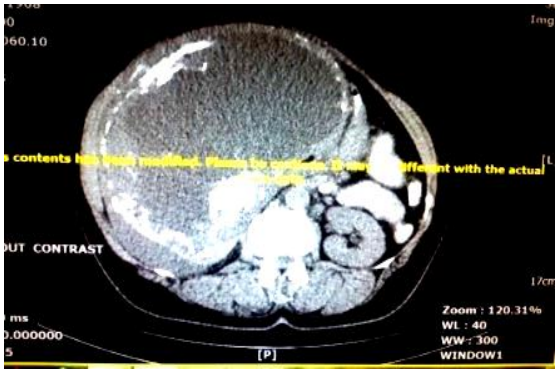

Figure 1. Image of retroperitoneal mass on the computerized tomography showing a heterogeneous mass with cystic spaces and a well-developed capsule

Based on the clinical and radiography evaluation, with initial diagnosis of cysts or tumors in the right kidney, the patient was admitted for surgery. The mass was dissected from the retroperitoneum but was strongly adherent to the right kidney. The right kidney and the mass were excised en bloc. Grossly, the specimen was an encapsulated, well-circumscribed, globular, grey-white to grey-brown mass (6 × 5 × 4 cm). The cut section revealed a variegated appearance with cystic area and region of calcification and ossification with hemorrhage (Figure 2).

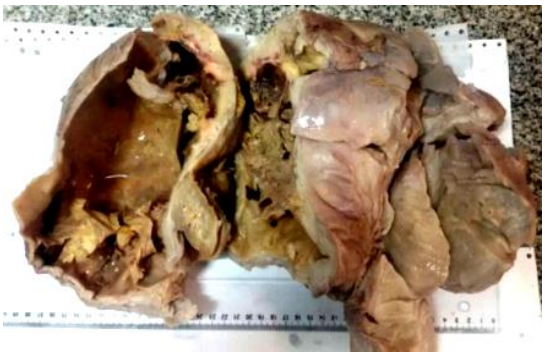

Figure 2. Tumor gross examination indicating a solitary encapsulated, well-circumscribed, globular, grey-brown mass with dimensions of 6 × 5 × 4 cm.

Microscopically, histopathologic section revealed proliferation of spindle cells arranged in fascicles in myxoid matrix. The tumor cells had a wavy figure with a poorly cytoplasm and oval nuclei. Verocay bodies were seen in some area of the tumor stroma. The stroma contains thick-walled dilated vessels. Nuclear polymorphism including bizarre forms with nuclear inclusions and other secondary changes such as calcification and ossification were seen.

Mitotic figure was zero. The accompanying kidney shows some degree of chronic inflammation and atrophy (Figure 3). In the immunohistochemical evaluation, the cells tumor were diffuse and staining positive for S100 protein (Figure 4).

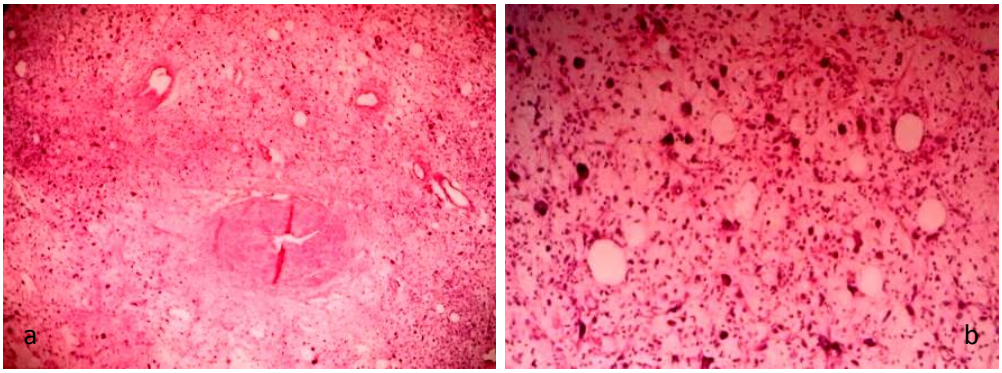

Figure 3. (a) Histopathological slide of the tissue showing proliferation of spindle-wavy figure cells arranged in fascicles in myxoid matrix (H and E, ×10). (b) Vascular sections with thin muscular walls, nuclear polymorphism, calcification, chronic inflammation, and atrophy (H and E, ×20) in the stroma tumor.

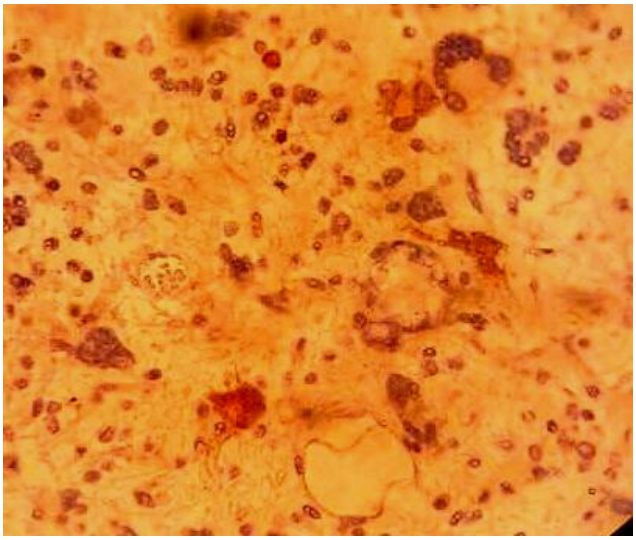

Figure 4. Immunohistochemical staining: Tumor cells reveal a diffuse positive expression for S-100 (×40).

Based on the histopathological and immunohistochemical findings, diagnosis of ancient schwannoma was confirmed.

The patient had been recovered after surgery and under regular follow-up since last 15 months without any evidence of recurrence.

Discussion

Retroperitoneal ancient schwannoma is an extremely rare tumor, which accounts for only about 0.5% to 12% of all retroperitoneal neoplasms (6). Etiology of schwannoma is unknown, but mostly found in peripheral nerve sheath in the limbs, head, and neck (7). Ancient schwannoma, in the retroperitoneal area, occurs more often between 40 and 60 years of age, with a slight tendency with male/female ratio of 2:3 (5). Here, the patient was a 50-year-old man. Because of the retroperitoneal AS usually have no symptoms, preoperative diagnosis of these tumors is often difficult (8).

To the best of our knowledge, only 10 cases of retroperitoneal AS can be found in urological, pathological, and radiological literature. This finding supported that the retroperitoneal AS seems to be extremely rare, therefore, in this paper, a unique case of AS in the retroperitoneal was presented.

Clinically, AS symptoms are nonspecific, such as ambiguous abdominal pain, flank pain, hematuria, headache, hypertension, and recurrent renal colic pain (9). Frozen section procedure could be done during the surgery procedure if it is essential to determine the

negative surgical margin (10). In this case, the tumor was resected as en bloc.

Grossly, schwannomas are usually solitary, well-circumscribed, gelatinous, firm, flat-surfaced with encapsulated or unencapsulated tumor which may be seen in degenerative changes such as necrosis and hyalinized vessel (6,8).

Histopathologically, schwannomas revealed a biphasic pattern, Antoni A and Antoni B. Antoni A area are consisted of fascicles of spindle cell which often form a palisaded arrangement around central acellular, eosinophilic areas known as Verocay bodies (4). Anti-type B area are composed of poor spindle cells arranged in the hypocellular myxoid matrix (1). In schwannoma, many variant types, such as plexiform, cellular, epithelioid, and ancient schwannomas have been explained (10). Ancient schwannoma is a very rare morphological type. It is characterized by degenerative changes, such as nuclear atypia, perivascular hyalinization, hemorrhage, necrosis, and calcification (4,8). Immunohistochemically, schwannoma demonstrated a strong and diffuse positivity for S100 protein in the cytoplasm of the tumor cells (5,6). In our case, Antoni A and B areas, and S100 positivity were seen.

Radiographically, computed tomography (CT) and magnetic resonance imaging (MRI) are useful for diagnostic evaluation although they are nonspecific (2). CT scan of schwannomas typically demonstrated well-defined cystic changes with necrotic central areas, calcification, hemorrhage, and hyalinization (4). Cystic changes in retroperitoneal schwannomas (up to 66%) is more than those in other retroperitoneal neoplasms (3). MRI of schwannomas usually shows masses of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. In general, the signal intensity on T2-weighted images may vary depending on cell density. Hypercellular tumors (Antoni type A) have intermediate signals, and tumors with Antoni type B have a bright signal on T2-weighted images (6,8).

Preoperative, fine needle aspiration biopsy can be performed easily but it is unreliable for diagnosis of retroperitoneal schwannoma (3).

Although retroperitoneal schwannoma is benign but malignant transformations reported, in some cases, have been associated with von Recklinghausen's disease. Malignant schwannomas is extremely rare with poor prognosis and they may show a high-grade sarcomas with local recurrence and distant metastasis (5,8). Malignancy is usually characterized by mitosis, pleomorphism, and blood vessel infiltration. Therefore, accurate monitoring is necessary after removal of retroperitoneal ancient schwannomas (1,3,7).

Complete surgical resection is the treatment of choice for schwannoma and prognosis is excellent (7). Local recurrence is rarely seen and malignant transformation is extremely rare. Laparoscopic resection of retroperitoneal schwannoma was explained (8,9).

In the present case, the patient had been recovered after surgery and under regular follow-up since last 15 months without any evidence of recurrence.

Conclusion

Retroperitoneal ancient schwannoma is an extremely rare tumor which may be misdiagnosed as a malignant tumor. Therefore, pathologist are recommended to consider ancient schwannoma as a rare event in the differential diagnosis of malignant soft tissue tumors in order to avoid aggressive treatment for patient.

Financial support and sponsorship

None.

Conflicts of interests

There are no conflicts of interests.

References

1. Abdal K, Heidari S, Emamverdizadeh P. Schwannoma in gingiva: A Case Report. J Kerman University of Medical Sciences 2019; 26(6): 494-498.

2. Radojkovic M, Mihailovic D, Stojanovic M, et al. Large retroperitoneal schwannoma: a rare cause of chronic back pain. Journal of International Medical Research. 2018;46(8): 3404–3410.

3. Giglio M, Giasotto V, Medica M, Germinale F, Durand F, Queirolo G, et al. Retroperitoneal ancient schwannoma: case report and analysis of clinico-radiological findings. Ann Urol (Paris) 2002; 36(2):104-6. doi: 10.1016/s0003-4401(01)00084-5.

4. Abdal K, Darvishi M, Hafezi Ahmadi M. A case report of malignant peripheral nerve sheath tumor of the breast. Journal of Babol University of Medical Sciences 2019; 21(1):1-5.

5. Al Skaini MS, Haroon H, Sardar A, Bazeed M, Al Zain A, Al Shahrani M, et al. Giant retroperitoneal ancient schwannoma: Is preoperative biopsy always mandatory? Int J Surg Case Rep 2015; 6C:233-6. doi: 10.1016/j.ijscr.2014.08.015.

6. Narasimha A, Kumar MH, Kalyani R, Madan M. Retroperitoneal cystic schwannoma: a case report with review of literature. J Cytol 2010; 27(4):136-9. doi: 10.4103/0970-9371.73299.

7. Wong CS, Chu TY, Tam KF. Retroperitoneal schwannoma: a common tumour in an uncommon site. Hong Kong Med J 2010; 16(1):66-8.

8. Akdemir F, Okulu E, Kayıgil Ö, Yazgan AK. Retroperitoneal ancient schwannoma: a rare case report. J Urol Res 2015; 2(4):1035.

9. Nayyar R, Khattar N, Sood R, Bhardwaj M. Cystic retroperitoneal renal hilar ancient schwannoma: Report of a rare case with atypical presentation masquerading as simple cyst. Indian J Urol 2011; 27(3):404-6. doi: 10.4103/0970-1591.85450.

10. Zhao M, Li X, Wang J, Li W, Huang Z. Retroperitoneal schwannoma treated with percutaneous computed tomography-guided radiofrequency ablation. J Neurosurg Spine 2012; 17(2):173-6. doi: 10.3171/2012.4.SPINE111061.