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clinical practice

Unusual presentation of retroperitoneal Schwannoma: case report

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SUMMARY: **Unusual presentation of retroperitoneal Schwannoma: case report.**

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Schwannoma is a rare tumor that develops from the Schwann cells in the nerve sheath. A 42 years old woman was found incidentally to have a bulky mass in epigastric region. Abdominal ultrasonography CT and MRI have been of aid to know the position and size of the tumor. A massive capsulated retroperitoneal lesion was identified. It moved forward the hepatoduodenal ligament, inferior vena cava laterally and aorta medially. The mass is exte-riorized and detached from adhesions. There were no complications after the operation and the patient was discharged on the fourth post-operative day. The microscopically examina-tion showed features suggestive of Cellular Schwannoma. After 8 months during follow-up, the patient did not report any neurological deficit and control CT did not suggest the presence of recurrent disease.

KEY WORDS: Retroperitoneal Schwannoma - Incidentaloma - Benign tumors.

Introduction

Schwannomas are rare tumor originating from Schwann sheath of the peripheral nerves or cranial nerves (1). Generally are solitary and encapsulated benign tumors, which are characterized by slow growth developed mainly in females (male/female ratio of 2:3) between the third and the fifth decade of life (2, 3). Schwannomas are usually located intracranially, in association with the eight cranial nerve while retroperitoneal location is rare, accounting for 0.3 % to 3.2%, often incidentally detected (4, 5). Retroperitoneal locations produce vague symptoms like abdominal pain and distension (6). CT and MRI can be define size and position but they do not allow to make histological diagnosis (6, 7).

Case presentation

A 42-year old woman came to our observation for nodular thyroid disease and an aspecific symptomatology characterized by palpitations and high blood pressure. *Cardiac ultrasonography* showed incidental presence of a mass in epigastric region.

Abdominal ultrasonography demonstrated the presence of a 113 x 93 x 87 mm bulky slightly vascularized hypoechoic mass with hyperechoic outline localized in the right hypochondriac region.

Upper abdominal MRI, confirmed the presence of a rounded mass of 89 x 78 x 110 mm with regular margins that displaces all surrounding parenchymatous structures.

CT scan reveals a more than 9 cm diameter solid mass with a small peripheral calcification and a necrotic area causing aorta dislocation to the left, interposed between pancreas and inferior cava, apparently originated from the apex of right psoas compartment (Figures 1, 2).

At surgery, a massive capsulated retroperitoneal lesion, forward the hepatoduodenal ligament, inferior vena cava laterally and aorta medially, was found (Figure 3).

The tumor was extended above and behind the pancreas and cranially as far as the right diaphragmatic pillar and diaphragmatic dome.

Enlarged lymph nodes along the left gastric ar-

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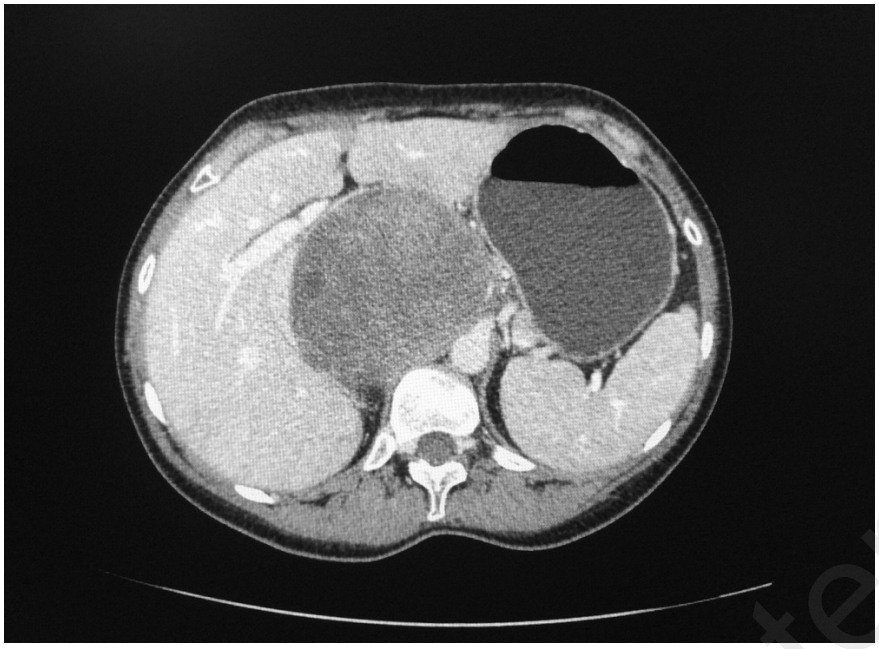

Figure 1 - CT scan shows a 9 cm solid formation.

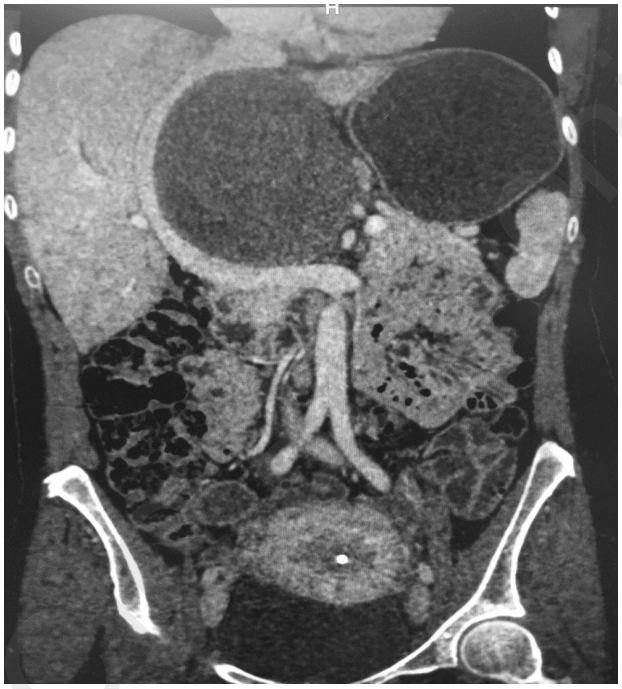

Figure 2 - CT scan. The bulky mass causes blood vessels dislocation.

Figure 3 - Operative picture.

tery were observed and extemporaneous examination was carried out. Histological finding demonstrated a reactive lymphadenitis so the mass was removed.

No postoperative complications were observed and patient was discharged 4 days after surgery.

A 12 x 9 x 6 cm solid and partly cystic (the largest cysts 3,5 cm) mass was found. Histological findings showed features suggestive of cellular Schwannoma. The immunophenotype was positive for S100, HMB45, desmin, smooth muscle actin

specific and EMA negative.

24 months after surgery, patient is asymptomatic, without cardiac or neurological disorders without recurrence.

Discussion

Benign schwannomas are usually slow growing tumors (diameter 5-15 cm) that predominantly occurs in females (male/female ratio of 2:3) between the third and the fifth decade of life (2, 4-6, 8).

Because retroperitoneal schwannomas are quite rare and lack specific symptoms, a correct preoperative diagnosis is often missed (9).

Symptoms are generally due to compression of adjacent structures (2). Most commonly symptoms are vagal, abdominal distension and pain while, in our case, we observed palpitations and high blood pressure (6, 8, 10).

Schwannomas have been associated with Recklinghausen disease which is characterized by the presence of coffee-milk stains, neurofibromas and multiple hamartomas (1, 2, 6, 11). None of those stigmata presented in our patient.

Schwannomas show cellular heterogeneity and cystic degeneration and the histological examination shows its benign features (2).

Due to the absence of any pathognomonic imaging features, erroneous imaging diagnosis is not uncommon (5): CT-guided fine needle aspiration (FNA) and endoscopic ultrasound (EUS); FNA are unrealiable in large schwannomas due to cellular pleomorphism in areas of degeneration (12). So, definitive diagnosis is based on definitive histological examination of the surgical specimen (5-7, 12).

Neoplasia presents in three specific histomorphological variations:

- conventional Schwannoma
- cellular Schwannoma
- melanotic Schwannoma

Conventional schwannoma is manifested anatomically and clinically as a single lesion, plexiform or by neoplastic foci. The histopathological exam classifies the conventional Schwannoma in two histotypes: type A is made by a cellular hyperdense population while type B is a poor of cell area, hypodense with micropseudocystic excavations and areas of necrosis (6, 7).

Cellular Schwannoma at the macroscopic examination has a globular shape, bordered by a capsul, well-defined. It has compact and uniform surfaces with parenchymatous consistency and grey and yellowish shades. Microscopic examination reveals a high density and compact cell population. Immunohistochemistry confirms the diagnosis by S-100 protein positive and negative for desmin and muscle-specific actin (5-10).

Melanotic Schwannoma presents irregular, globoid shape, surrounded by a fibrous capsule and its colour varies from blue to black. Its charactheristic is more or less melanin granules found in tumor cells (13). Signs of malignancy are represented by mitotic figures, pleomorfism and infiltration of blood vessels.

Malignant Schwannomas are much more aggressive, generally associated with Recklinghausen's disease. They also have a high incidence of local recurrence after excision and may develop metastasis (6).

According to the literature, gold treatment for benign Schwannoma is radical surgical excision which allows an accurate pathological evaluation and correct diagnosis (4).

Competing interests

The Authors declare that there is no conflict of interest regarding the publication of this paper.

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