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Case Report Open Access

Retroperitoneal Schwannoma: A Case Report

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Abstract

Schwannomas are rare tumors originating from Schwann cells, producing a nonspecific clinical picture that depends on the location of the tumor. The retroperitoneal location is infrequent, its incidence varies from 1% to 5% of all neoplasms in this site. The definitive diagnosis is histopathological, but it is supported by imaging studies. The definitive treatment is complete extirpation. We present the case of a 24-year-old man with no relevant pathological history who presented with abdominal pain and paresthesia in the lower limb, treated in the department of oncological surgery. The imaging studies showed a retroperitoneal tumor whose histopathological diagnosis was schwannoma with positive immunohistochemistry for S-100. Exploratory laparotomy and complete tumor resection were performed, with complete posterior recovery. After three years of follow-up, he was asymptomatic.

Keywords: Retroperitoneal schwannoma; Schwann cells; Retroperitoneal tumors

Introduction

Schwannoma, also known as Neurilemoma, is an encapsulated tumor of the peripheral nerve sheath that resembles the differentiated Schwann cell [1] and is considered within the category of soft tissue neoplasia [2]. The first tumors of peripheral nerves were described in 1910 by José Verocay (1867-1926) in his work "The knowledge of neurofibromas" but it was not until 1935 that Arthur Purdy Stout (1885-1967) introduced the term Neurilemoma. [3]

They are benign neoplasia of slow growth, usually encapsulated, that can affect any peripheral nerve. They represent between 0.2% and 0.6% of neoplasia in general [4], being the retroperitoneal location extraordinarily exceptional, representing 1%-3% of all schwannomas and only 1%-5% of all retroperitoneal masses.

The clinical picture is non-specific. The most common symptom being an occupying mass effect; in many cases the diagnosis is usually an incidental finding during an imaging study.

Case Report

Male, 24-years-old, consulted for abdominal pain and paresthesia in the right lower extremity associated with a right mass of the abdomen, with asymptomatic evolution for 3 years until February 2015, when he perceived a dull pain, with 5/10 intensity, no irradiated and associated with paresthesia in the homolateral lower limb. There is no relevant medical history.

In the physical examination, patient with good general condition, normal vital signs, and the right hemi-abdomen was asymmetric due to a palpable mass, solid consistency adhered to deep plane, not painful to palpation and kill to percussion. In the lower limbs, peripheral pulses were palpable, capillary refill in less than 2 seconds, sensitivity and motor skills were normal.

Abdominal ultrasound (US) and CAT (Figure 1) were performed, which reported a retroperitoneal tumor at the level of L4 that measured 8 x 7 cm that displaced the ureter and the ipsilateral vessels. Subsequently, an MRI is performed (Figure 2) that reports a neoplasm of 75 x 60 x 89 mm, located above the iliopsoas muscle, isointense to the root of the mesentery on which it seems to depend, there is no connection with the lumbar nerve roots. Complete blood biometry, blood chemistry and general urine test reported results within normal parameters.

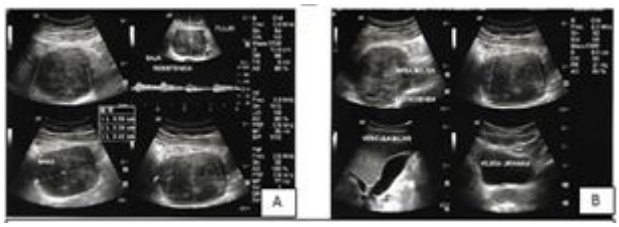

Figure 1: A and B Tumor measuring 84 x 56 x 65 mm, with hypoechoic zones surrounded by an ecgenic capsule, located at the paravertebral level.

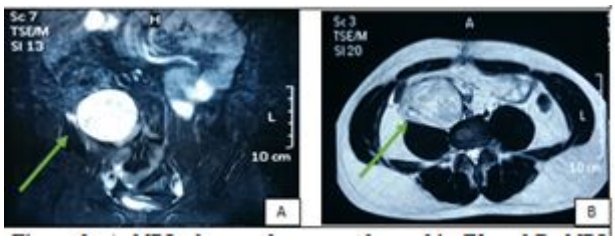

Figure 2: A: MRI of coronal cortex enhanced in T2 and B: MRI of cross-section enhanced in T2, in both cases the arrow indicates a tumor of 75 x 60 x 89 mm, isointense to the root of the mesentery on which it apparently depends.

A first surgical approach is made by performing an exploratory laparotomy with incisional biopsy that showed in the histopathological report fusiform cells, wavy and round that form palisades in two types of areas: Fascicles that form type A and areas type B, laxas with characteristics myxoids (Figure 3(A-3C)). Vascular proliferation and areas of hemorrhage were also reported, in the absence of mitosis and necrosis. The immunohistochemical study showed positivity for the NSE protein, S-100 (Figure 3(D)) and vimentin, with a low cell proliferation index (ki-67 of 8%) accompanied by negativity for CKAE1, CKAE3, CD31, CD 34 and CD 68. The findings are compatible with a schwannoma.

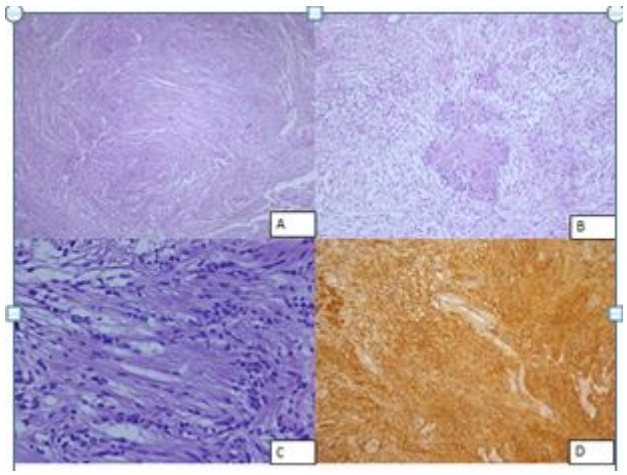

Figure 3: Histological sections of Schwannoma A: Fusiform pattern HE (4X), B: Areas Antoni A (Bodies of Verocay) and Areas Antoni B: HE (10X), C: High power image of nuclear palisades of the Bodies of Verocay HE (40X), D: IHQ: S-100 Strong and diffuse positivity.

In a second surgical approach an exploratory laparotomy was performed for tumor resection, the findings in the transoperative were:

Discussion

Schwannoma, also called neurilemoma, is a benign tumor that originates from the Schwann cells that line the sheaths of the peripheral nerves; these derive embryologically from the cells of the neural crest. These tumors can appear in any nerve of the body, with the exception of the cranial nerves I and II, since they do not possess this type of cells [5,6].

They are usually located in 40%-50% of cases in the skull and 30%-35% in the extremities, studies show that the retroperitoneal location is one of the most infrequent, since it represents between 1% and 5% of retroperitoneal tumors in general and between 1% and 3% of all schwannomas [5].

A study conducted by Theodosios et al. In which they were retrospectively reviewed 15 years found 69 of patients with retroperitoneal tumors and only 5 of them had histopathological diagnosis of retroperitoneal schwannoma [7].

Of the documented cases, very few have been reported with malignant transformation, which is why it is considered infrequent [1]. It has been described that the size of the tumor is not related to its tendency to malignancy [8]. To date, the risk factors for this disease are unknown.

The age of presentation of the tumor varies between the second and fifth decade of life, its distribution according to sex is controversial, since in some studies a greater incidence is observed in women than in men [5]. A female ratio of 2: 3 [9], while other studies claim that there is no predisposition for sex [10].

According to population studies, of schwannomas in general, 90% of cases are sporadic, 3% associated with neurofibromatosis type 2, 2% associated with meningioma and the remaining 5% associated with shwannomatosis [1].

In its retroperitoneal location, an association has been described in 5%-18% with Von Recklinhausen disease or neurofibromatosis type 1, which is inherited in an autosomal dominant manner. In these cases, the location of the schwannomas is usually atypical and malignant [6].

The clinical presentation depends on the location and size of the tumor, which due to its benign nature does not tend to invade neighboring organs and, therefore, is manifested by the symptoms produced by the compression of these. The clinical spectrum in its retroperitoneal location is commonly manifested by undefined abdominal pain, poor localization and nonspecific neurological symptoms in the lower extremities [5].

Diagnostic approach of Retroperitoneal Schwannoma includes imaging evaluation, histopathology and immunohistochemistry. Ultrasound represents a cheap and easily accessible tool but is limited by its low specificity. CT is useful to determine degenerative changes such as calcification, hemorrhage, cystic formations and necrosis. MRI is actually the imaging gold standard for the diagnosis of retroperitoneal tumors since it is capable of determining the origin, extension and internal composition of the lesions. However, despite the benefits of imaging studies benefits this type of tools fail because they cannot differentiate benign from malignant lesions with certainty [5]. It's been suggested that Positron Emission Tomography (PET) can be helpful in differentiating the malignancy of these lesions [5].

Once you have the suggestive evidence of schwannoma through imaging, we proceed to make the definitive diagnosis through histopathology, but it must be complemented with the immunohistochemical study. It's been suggested the usefulness of Fine Needle Aspiration Biopsy (FNAB) guided by CT to the preoperative diagnosis. Several authors have commented that this technique does not have much utility in practice due to the several limitations: first it is possible that the extracted material contains insufficient Schwann cells for the diagnosis; Secondly, if a biopsy of pleomorphic areas with degenerative changes is obtained, they can guide erroneously the diagnosis to a malignant process and thirdly, it can represent a risk for tumor dissemination, hemorrhage and infection. Thick needle biopsy guided by CT seems to have better results than FNAB, but still has substantial risks of morbidity [5].

The macroscopic histopathological study revealed encapsulated tumors, because the nerves are surrounded by a true capsule, the epineuro. The external appearance depends on the size of the tumor, the small tumors resemble a neurofibroma due to its fusiform shape, these usually obliterate the nerve of origin, in the larger tumors, it presents as an eccentric mass on which the nerve fibers are being displaced, these tumors usually do not measure more than 5 cm, except those that are located in the mediastinum and retroperitoneum that tend to be larger and therefore tend to manifest secondary

degenerative changes such as calcifications or cystic formations, the interior is usually pink, white or yellow.

At the microscopic, structural areas known as Antoni A and Antoni B are characteristic. The first pattern is composed of compacted spindle cells with "palisade" nuclei known as Verocay bodies [8]. It also presents cytoplasmic prolongations and occasionally intranuclear vacuoles, all these components arranged as a fascicle. The Antoni B expresses a less marked and ordered cellularity, the oval cells are immersed in a lax and disordered matrix accentuated by microcystic changes, inflammatory cells and collagen fibers [1].

It is considered that malign schwannomas have irregular external surfaces and tendency to invade neighbor structures [11].

The immunohistochemistry panel is typically positive for S-100 Protein, vimentin and Neuro-specific Enolase (NSE), CD56 has also been mentioned by other authors, and is negative for Smooth Muscle Actin (SMA), HHF35, CD34 and CD117 [5].

More recently, SOX10 has been used as a marker for differentiation of the neural crest and has proven to be very useful for schwannomas, especially since leiomyosarcoma does not express it, however the Schwann cells of a neurofibroma are capable of expressing it, but always to a lesser extent [1].

Traditionally S-100 protein is specific for nerve tissue, however it has been reported that in some non-neuronal cells such as Langerhans cells of the skin, fat cells, and chondrocytes may also present some sparse positive reaction. However, in schwannomas the positivity is strong and diffuse [12]

With the finding of a retroperitoneal mass, it is vital to rule out malignancy, mainly sarcomas that appear as hard-surfaced masses with an irregular surface, surrounded by a capsule that is quickly overcome by tumor growth, infiltrating the posterior parietal peritoneum and the intra-abdominal viscera attached to it, contrary to the benign schwannoma that does not infiltrate peritoneum or invade neighboring structures. Also must to be thought of as a differential diagnosis in a neurofibroma whose origin also resides in nerve fibers and behaves similarly; however, it differs from schwannoma because it does not express the S-100 protein, compare to schwannomas that exhibit greater cell differentiation with encapsulated lesions and that they do not contain axons [13].

The definitive treatment of schwannoma is complete exeresis with clean surgical margins. In order to achieve this goal, tissues and organs that are in contact or adhered to the tumor must be sacrificed [14]. Alternative treatments such as chemotherapy and radiotherapy have not proven to be efficient; therefore their use is not recommended [5].

Local recurrences that usually occurs in the first six months after surgery and whose incidence varies from 16% to 54% have been described [9]. They usually occur more frequently in patients who do not undergo complete removal of the tumor, occurring in up to 36.5% of cases. Therefore, periodic follow-up of these patients with control imaging studies is suggested [15-17].

Conclusion

Retroperitoneal schwannomas are benign neoplasia in most of the cases; very infrequent, assuming no more than 5% of retroperitoneal tumors. Clinically they are characterized by their non-specificity; patients are asymptomatic or may have symptoms due to the occupant effect of the tumor. Diagnosis is therefore often complex and based on three essential aspects: imaging studies that provide data such as location, size and commitment of neighboring structures; in histopathology by observing Antoni's area A and B; and finally in immunohistochemistry, whose main marker is the S-100 protein, which is strongly positive and diffuse.

Given the occasional presence of necrosis, cystic areas and hemorrhagic zones, the use of FAC guided by CT does not provide conclusive data to the diagnosis, the biopsy with thick needle guided by CT has not shown such encouraging results either. Currently, the treatment of choice is tumor resection with clean margins.

Very few cases of malignancy have been documented, which is why it is considered infrequent, but not of local recurrence, which in most cases is secondary to an incomplete resection of the tumor.

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