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Retroperitoneal Schwannoma Simulating An Ovarian Tumor (*Case Report)*

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***Abstract**: Schwannoma is a tumor that develops on the Schwann cells. It is a very rare tumor both in terms of its frequency and its retroperitoneal location. Its presentation is varied and its preoperative diagnosis is difficult. Although the vast majority of schwannomas are benign, there are malignant forms frequently associated with Von Recklinghausen syndrome. Its diagnosis is histological and its surgical treatment consists in a complete removal of the mass because of a tumor degenerative risk.We report a case of a 64 year old female patient, followed for chronic pelvic pain associated with digestive signs, ultrasound and CT scan showed a pelvic retroperitoneal mass of 8x 6 cm of great axis. The removal was performed by laparotomy. The histological and immunological study concluded to a benign schwannoma. Through a review of the literature we recall the frequency, diagnosis, imaging data, treatment and evolution of this rare tumor.*

Keywords—Retroperitoneal tumor; Benign schwannoma; Surgery

1. INTRODUCTION

Schwannoma is a rare nerve tumor, developing from cells of the Schwann nerve sheath (1, 2, 4). It is usually benign but with a risk of malignant transformation (1, 2, 3). It develops in most cases in the cranial nerves (especially the eighth pair) or in the peripheral nerves but extremely rarely in the retroperitoneum (1, 4, 5). It may be part of a phacomatosis (2).

This tumor poses the problem of preoperative diagnosis because it is only confirmed on histological examination of the adnexectomy specimen (1, 3, 4, 5).

We report a new case of pelvic retroperitoneal schwannoma and through a review of the literature we will give an update on this type of tumor.

2. CASE REPORT:

The patient was 64 years old, multiparous, with no particular pathological history, admitted for chronic pelvic pain of the lumbosacral region, radiating to the thighs and external genitalia, associated with constipation, without urinary signs, and evolving in a context of apyrexia and conservation of the general state. The gynecological examination noted the presence of an abdominopelvic mass reaching the umbilicus without other associated signs.

Abdominopelvic ultrasound showed an echogenic, multiclonal image with a well-limited supra- and latero-uterine double component.

The abdominopelvic CT scan showed a large retroperitoneal mass, suprauterine, multicompartmental, with a double component of fluid and fat containing calcifications with an enhanced wall and partitions after injection of the contrast medium (Figure 1).

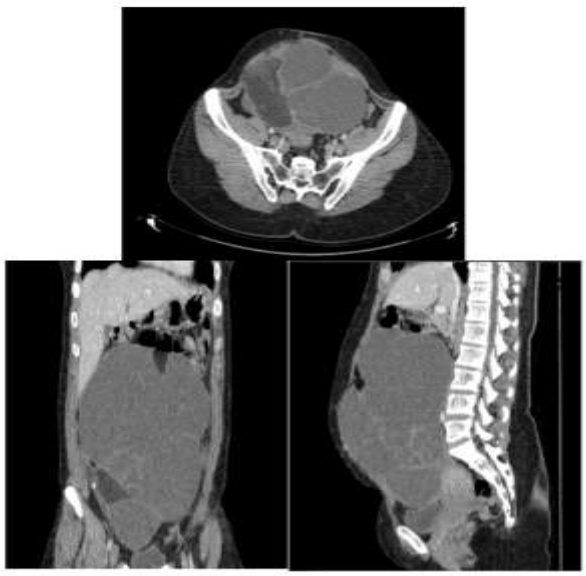

Figure 1 : The abdominopelvic CT scan showed a large retroperitoneal mass, suprauterine, multicompartmental, with a double component of fluid and fat containing calcifications with an enhanced wall and partitions after injection of the contrast medium

The surgical exploration performed by laparotomy found a retroperitoneal tumor, the direct approach allows easy removal of the tumor, the postoperative course was simple. Histological examination of the surgical specimen confirmed the diagnosis of benign schwannoma .

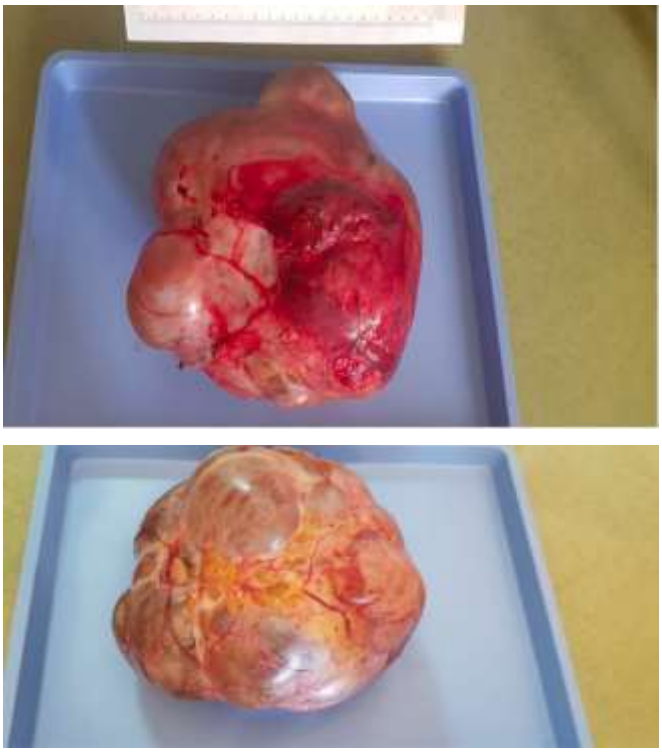

Figure 3 : The surgical exploration performed by laparotomy found a retroperitoneal tumor, with a double component and weak vascularization in the periphery without ascites or carcinosis

Histological examination of the surgical specimen confirmed the diagnosis of benign schwannoma .

3. DISCUSSION

Schwannoma is a solid tumor that develops at the expense of Schwann cells (1, 2, 3). In 1910, Verocay described the first observation (6, 7). The term schwannoma was coined by Masson in 1932 (1, 2, 7). A schwannoma is very rarely retroperitoneal (1, 2). It represents 3% of schwannomas and 4% of primary retroperitoneal tumors (1, 2, 8). This retroperitoneal location represents only 0.7% of benign schwannomas and 1.7% of malignant schwannomas (1, 2). Both sexes are equally affected and the age of the patients is most often between 30 and 60 years (1, 2, 4). Association with phacomatosis is usual (1,2). Diagnosis is most often incidental and late, given the latency of the tumor's evolution. Sometimes during exploration of abdominal or pelvic pain or due to compression of neighbouring organs: gastric pain and dyspepsia, but alsoportal thrombosis or ureteral compression (1,2,3,4,6,7,8). This lack of specificity of clinical signs makes its diagnosis difficult and often delayed. Ultrasound and CT scan, if they make the diagnosis of retroperitoneal mass, do not show pathognomonic images, however schwannoma is classically described as a well encapsulated and solid tumor when it is small. It may become hemorrhagic and necrotic when larger in size (2, 3, 4, 9). Another imaging argument is the presence of intra-tumoral cysts which are found in 63% of cases for benign schwannomas and in 75% of cases for malignant schwannomas, this is noteworthy because retroperitoneal tumors rarely form cysts (2, 4, 9, 10). Calcifications have also been observed, which would be an argument in favor of degeneration (2, 3, 4). Nuclear magnetic resonance provides the same information. It shows a well encapsulated tumor with a hyposignal in T1 and a heterogeneous hypersignal in T2 (4, 9, 10, 11).

Preoperatively, the diagnosis of schwannoma will be evoked in one third of cases (1,2,4,6,7,8). The diagnosis of certainty can therefore only be made on histology, the samples being obtained by biopsies or on the operating room. Percutaneous aspiration biopsy is not recommended by most authors because of the difficulties of interpretation, the risk of neoplastic dissemination in case of malignant tumor and the peritumoral hypervascularization (1, 2, 4, 6, 12).

Surgical excision is recommended in view of the possible histological heterogeneity of retroperitoneal tumours (1,2,4,5,6,12,13,14,15).

Macroscopically, the tumor is nodular, well-limited, encapsulated and has cystic areas. Microscopically, the tumor is characterized by the presence of Verocay nodules (1).

There are two histological types of schwannomas (Antoni's types A and B):

- type A: spindle cells, arranged in bundles. The nucleus is oval. The cytoplasm is not very abundant;

- type B: the arrangement of the spindle cells is random. The cells are separated by a matrix stained heterogeneously with hematoxylin eosin and alcian blue. The expression of the S100 protein in immunohistochemical study is the witness of a neurectodermal differentiation, its positivity thus directs towards a schwannoma (1,2).

Treatment consists of complete surgical enucleation of the tumor, which is generally easy because it is encapsulated.

The approach depends on the location and volume of the tumor (1,2,4,5,7,13,15).

This excision can be difficult and sometimes incomplete because of intimate contact with large vessels or noble organs (2,4,5,13,14). Recurrence is rare if the removal has been complete. A case of heterotopic recurrence has been described, appearance of a retroperitoneal schwannoma following removal of a spinal schwannoma (16). On the other hand, benign schwannoma degenerates only exceptionally: only one case has been described where a malignant schwannoma appeared remotely on a site of excision of a benign schwannoma (2, 4, 16). This low risk of

transformation and recurrence makes annual postoperative CT surveillance necessary (1,2,5,14,15). Although the vast majority of schwannomas are benign and have a good prognosis, there are malignant forms whose histological nature is controversial because of their frequent association with Von Recklinghausen syndrome (4% of cases) and other neurofibromatoses (2).

The treatment of malignant schwannoma is surgical, as these tumors respond poorly to radiotherapy or chemotherapy. Metastases are preferentially hepatic, pulmonary, bone and subcutaneous tissue (17). Lymphatic dissemination is very rare. The association of malignant schwannoma with neurofibromatosis is a negative factor since the 5-year survival rate drops from 47% to 23% (18).

4. CONCLUSION

Retroperitoneal schwannoma is a rare tumor with a good prognosis in its benign form. Its clinical diagnosis is often delayed because of a borrowed symptomatology. The paraclinical examinations specify its retroperitoneal origin and evaluate the possibilities of removal. Radical surgery is the reference treatment. Because of recurrence or even malignant transformation, further surveillance is necessary.

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