CT and MR imaging of retroperitoneal schwannoma

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Abstract
The purpose of this presentation is to allow the radiologist to discuss the diagnosis of retroperitoneal schwannoma, involving mostly a fortuitous discovery and a misleading clinical presentation. We present 4 cases of retroperitoneal schwannoma, two having benefited from a surgery and two others of a therapeutic abstention. The retroperitoneal localization and the imaging are good indicator elements of this pathology. The constant improvement of the CT and MR imaging allows a better approach of this entity also by specifying its localization and its anatomical relationships to guide the therapeutic attitude which must be remain multidisciplinary.

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Primary retroperitoneal tumours are rare with a prevalence of less than 1%. Eighty percent of the cases are malignant, half of which involve sarcoma. The compliance of the retroperitoneal space delays their discovery. This space is currently well analysed by computed tomography (CT) and magnetic resonance imaging (MRI). Their resolution in density or signal provides a spatial, morphological as well as tissue analysis that helps determine the type of lesion and plan the surgery.

Among the benign lesions, schwannoma are rare. The clinical presentation is often misleading or even absent although the radiological appearance indicates the diagnosis. Diagnostic difficulties are often encountered.

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We present four cases where the pre-surgical imaging strongly indicates the diagnosis of retroperitoneal schwannoma. This is confirmed by the surgery in the first two cases. The latter two cases were not treated and were monitored for 10 years and 30 months respectively.

Case 1

Mr M., 40 years old, without any noteworthy antecedents, consulted the emergency unit for pain in the side and left iliac fossa that increased over several days, associated with constipation without bowel obstruction and dysuria since the previous evening.

He reported low-grade fever at home that responded well to paracetamol.

The clinical examination detected a sensitive abdomen, especially on the left, without defence, fever or source of infection. Only the CRP was high, the leukocytes were normal and no bacteriological sample was found to be positive. A CT-scan with injection was carried out, followed by MRI.

The CT-scan revealed a large (height: 12.7 cm, anterior posterior diameter: 10 cm, transverse diameter: 11.5 cm), heterogeneous pelvic tumour comprising calcifications and fluid without visible fat with enhancement of its tissue (Fig. 1).

It pushes the bladder forward and the rectum to the right. The MRI revealed haemorrhagic modifications with liquid levels at the right side of the lesion (Fig. 2).

The MRI and CT analysis confirmed the retroperitoneal location with a fully visible elevation of the peritoneal layer (Fig. 3).

The radiological appearance and the anamnesis suggest a slowly evolving lesion.

The radiologist concluded that it was a neurogenic tumour and first indicated a schwannoma or a paraganglioma.

A biopsy by anterior lateral route with marking of the path of the needle with methylene blue on the fleshy portions confirmed the hypothesis of schwannoma.

The patient presented intermittent episodes of fever attributed to the tumoral necrosis.

Intense hyperfixation was found on the tissue part of the lesion by PET-scan (a maximum of 13 SUV at the anterior superior part of the tumour and a maximum ranging from 5 to 9 SUV on the other tissue regions), without any other suspect metabolic site (Fig. 4).

The full excision of the tumour was carried out outside of the connecting hole of the first right sacral vertebra with definitive histological confirmation of benign cystic schwannoma.
Figure 4. PET (first line), CT (second line) images and PET-scan fusion (third line) objectifying the fixation of 18-FDG within the tissue zones of the mass.

Case 2

Ms L., 30 years old, consulted for intense abdominal pain on the left side without fever that persisted for several weeks. The biological investigation was normal. She also reported antecedents of nephritic colitis associated with weight loss and anorexia.

The injected abdominopelvic CT-scan (Fig. 5a and b) detected a solitary, well-defined, left lateroaortic tumour, located between the lumbar vertebrae and the left psoas muscle, in contact with the primary iliac vessels. This oblong lesion measures $3 \times 3.4 \times 7.7$ cm and presents a cystic centre (density: 17 Hounsfield units), low peripheral enhancement and several rare calcifications. The retroperitoneal location

Figure 5. CT injected at portal time revealing this mass along the retroperitoneal vessels and the independent and slightly retaining aspect of the intestinal loops. Presence of a central necrosis and several microcalcifications. a: oblique coronal format; b: axial section.
of this tumour may be suggested due to the close contact with the aorta and the left primary iliac vessels.

The full excision of this lesion confirmed mainly Antoni type A schwannoma (Fig. 6) with PS 100 positivity in immunohistochemistry (Fig. 7).

Case 3

Mr B., 74 years old, consulted in rheumatology for pelvic bearing down lateralised on the right, which he claimed has been increasing over the last few years. He did not present any changes in his general condition or fever or inflammatory syndrome.

The patient mentioned an old tumour that was detected at least 10 years ago, seen in an old CT-scan for which he had a few not very useful images. An MRI was requested (Fig. 8a and b).

It revealed a tissue tumour, oval in hyposignal T1, heterogeneous hypersignal T2 with well-defined peripheral ring in hyposignal which was 7.1 cm thick, 4.6 cm wide and 7.5 cm high. This lesion was located outside of the inner iliac vessels, at the anterior right part of the sacrum and appearing in close relationship with the right L5 root. The anamnesis and the imaging indicated schwannoma.

Case 4

Ms C., 24 years old, was referred to the emergency unit for left colic nephritis. The biological investigation was normal. A CT-scan was requested (Fig. 9a and b). It did not find lithiasis or urinary tract disease but, by chance, detected a well-defined, unilocular, cystic image (3.8 cm by 4.3 cm in the axial plane and 4.9 cm high), with enhancement of only one wall. This lesion, located in the retroperitoneum, under the cauda pancreatis, seemed to be extrapancreatic and indicated schwannoma. The MRI also pointed to a unilocular cystic lesion of the retroperitoneum suggesting cystic schwannoma (Fig. 9c). No treatment with monitoring was chosen. No change in the radiological appearance was noted after 30 months.

Discussion

What may orient us towards the diagnosis of retroperitoneal schwannoma?

When faced with the discovery of a retroperitoneal tumour, the absence of signs of clinical gravity is an argument pointing to a possible schwannoma.

It most often involves an asymptomatic tumour discovered by chance. These tumours may become symptomatic by the compression they exert on the adjacent structures according to their size and location. Now, the retroperitoneum is a compliant space that allows lesions, especially if their evolution is slow, to reach a large size. The clinical aspects, when they exist, are varied and non-specific: urinary disorders (haematuria, hydronephrosis by urethral compression with nephritic colitis) [1], lumbago, pain or sensation of bearing down, renovascular hypertension, weight loss, unexplained fever [2], venous thromboses [3].

Schwannoma or neurinoma are the most common tumours of the peripheral nervous system. The location is ubiquitous except for cranial nerves I and II, preferentially in the head, neck and limbs. Impairment of the retroperitoneum is rare and is estimated at between 0.3% and 3.2% of all schwannomas [4,5]. Of all tumours of the retroperitoneum, the percentage of the schwannomas varies according to the series: 0.5 to 3% [6]. In the retroperitoneum, they are generally paravertebral, near the kidneys or pre-sacral. Occasionally, they may occur in the abdominal wall, bladder or intestinal lining. It is often difficult to determine the carrying nerve.

Multiple impairment is possible in 5 to 18% of the cases and integrates with type II neurofibromatosis. It occurs in the thirties and forties with controversial impairment in the literature as to the gender [7,8].

With imaging in slices, the retroperitoneal location no longer presents the radiologist with a real problem due to the assimilation of the abdominal compartments and the inherent communications as well as the radiological semiology described (the “beak sign”, “phantom organ sign”, position bascule…) [9,10].
Certain radiological aspects may indicate schwannoma in spite of the rareness in this space:

First of all the size, even though variable, may be 10 or even 20 cm in the extraperitoneal space. This large size is due to the compliance of the space and the slowly evolving nature of this lesion [11,12].

The lesion is solitary and oval or sphere-shaped.

The sonograph may reveal a well-defined mass, with regular contours that is hypoechogetic or of mixed echogenicity.

The CT-scan offers an optimum spatial analysis with a wide field of exploration, enabling pre-surgical mapping, in particular with the adjacent vital organs. Although the CT-scan provides a better analysis of the bone extension, the tissue analysis is better with MRI. The CT-scan finds a well-defined lesion, whose density depends on the lesional rearrangements. Whether cystic, necrotic, haemorrhagic or in the form of calcification, the larger the tumour the more marked the rearrangements [13,14]. The CT-scan guided biopsy may not contribute a great deal in view of these rearrangements and remains debatable [4]. Pure cystic forms are described and should not automatically eliminate the diagnosis [15–17]. Califications are possible in all types of neurogenic tumours [18].

In MRI, the association of a T1 isosignal at the adjacent skeletal muscle, with a T2 hypersignal (or even an isosignal to the muscle) is classically described but remains non-specific. The intensity of the signal in T2 is inversely proportional to the cellularity of the tumour, as opposed to the necrotic zones that reinforce the T2 hypersignal. Therefore, Antoni type B is associated with a more intense T2 hypersignal. The contrast enhancement will involve the tissue portions as well as the partitions, septa and walls of the lesion and the more the rearrangements, the more heterogeneous the contrast.

The locations of the duodeno-pancreatic sphere offer a good and efficient alternative, especially with the small lesions (under 3 cm) reported in the studies that emphasise...
the contribution of endoscopic fine-needle guided aspiration biopsy [19,20].

Certain authors propose an angiography in order to assess the vascularisation of the tumour and/or consider pre-surgical embolisation [11].

The hyperfixing nature of the lesion by CT-scan, as in patient 1, is not a proven criterion of malignancy in view of the low specificity of the CT-scan [21].

Its malignant transformation is exceptional and the most common site is on the large roots of limbs [22]. It is indicated by the term MPNST (Malignant Peripheral Nerve Sheath Tumour) [23,24].

The main differential diagnoses are for malignant lesions, sacromateous adenopathy and, for the benign lesions, cystic lymphangioma and GIST.

The histological aspect

A schwannoma or neurinoma is a benign tumour exclusively consisting of Schwann cells, without axon, without myelin and without participation of the endoneurium, as opposed to neurofibromes that are formed by the proliferation of all of the elements comprising the normal nerve, including myelinated and unmyelinated axons, perineural cells and fibroblasts with dissociation of nerve fibres.

As opposed to the neurofibroma, the schwannoma is well defined, most often unique, and has an exceptional risk of degeneration, as opposed to each of the elements noted for the neurofibroma. This histological difference results in the macroscopic appearance and the surgical sanction with a difficult resection of the neurofibroma due to the anergic interlinking of the nerve elements without a border between healthy tissue and lesion.

Macroscopically, it consists of a most often pearly white tumour with haemorrhagic or cystic rearrangements, establishing on a nerve bundle or at the meeting point of several bundles. These rearranged schwannoma are qualified as "ancient schwannoma". As in imaging, this tumour appears to be well defined and oval or round, always with a split outside of inflammatory adhesions [25].

This tumoral proliferation of Schwann cells is limited by a capsule, separating the tumour from the rest of the nerve and adjacent structures. Mitoses are absent and the nerve fibres are pushed back and incorporated in the capsule. Two distinct architectural aspects are described. Antoni type A corresponds to a very cellular appearance where fusiform cells and fibres form frameworks that, at places, are erected like fences. The juxtaposition of these fences is described under the term of Verocay body. Antoni type B corresponds to looser tissue characterised by myxoid degeneration.

The prognosis of a pigmented variety, called melanotic schwannoma, is more variable.

In immunohistochemistry, the PS100 is positive.

What treatment can be proposed?

The therapeutic attitude remains debatable for these retroperitoneal schwannoma.

Since the risk of recurrence is low and malignant transformation even lower, certain authors prefer enucleation and the results are good [21]. The lack of a certain diagnosis in imaging and the, exceptional although described, risk of degeneration encourages other authors to perform more widespread and radical surgery with, for optimum treatment, the most complete excision possible, including wider margins of safety [11].

Partial excision may be considered if there is invasion of a root or motor trunk whose impairment may result in prejudicial functional sequelae, making certain authors remember the importance of identifying the carrying nerve [2].

The radiologist should mention the anatomic relationship of the lesion with the adjacent organs, try to identify the carrying nerve and also give warning signals about possible degeneration and/or the uncertainty as to the possibility of another malignant disease, as the certain diagnosis of schwannoma remains histological.

Several approaches have been described: transperitoneal by anterior route or anterolateral and retroperitoneal by lateral or paravertebral route.

When there is a major intra-spinal extension such as hourglass schwannoma, a combined approach (laminectomy then anterior transperitoneal) has been described by Pollo [2]. An anterior extraperitoneal approach was also reported for a pre-sacral lesion [26]. Resection by laparoscopy has also been carried out by several teams [27—29].

In a study where the histological diagnosis was based on ultrasound endoscopic biopsies, Kudo et al. proposed monitoring in view of the benefits/risks of surgery [19].

Conclusion

Consisting of a benign lesion with a high surgical risk due to the location, the procedure when confronted with a suspicion of retroperitoneal schwannoma should be based on a radio-surgical consultation. If the imagining is highly indicative and the diagnosis is backed up by a biopsy, monitoring in an asymptomatic patient appears to be reasonable [12]. As for symptomatic patients, surgical resection should follow good pre-surgical mapping and, if possible, a biopsy should be directed on the tissue zones. Pre-surgical embolisation for large tumours should also be considered. Monitoring is recommended after partial excision.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


