

CLINICAL CASE - TEST YOURSELF Abdominal Imaging

Incidental left flank mass

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PARTA

A 71-year-old female patient, with an unremarkable past medical history, presented at the radiology department for a scheduled kidney-ureter-urinary bladder ultrasound (KUB US) for further investigation of dysuria. Laboratory studies were normal. An incidental mass was discovered in the left flank on US (**Fig. 1**). A Computed Tomography (CT) of the abdomen and pelvis (**Fig. 2**) followed by a Magnetic Resonance Imaging (MRI) examination of the retroperitoneum (**Figs. 3-5**) were performed.



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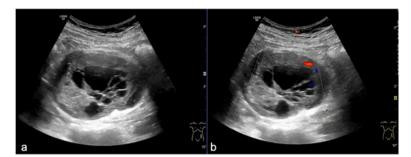


Fig 1. Gray scale US (a) and colour Doppler over the left flank area (b).



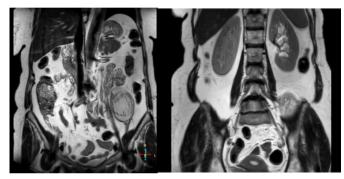


Fig 3. Coronal T2 weighted images

Fig 2. Portal venous coronal CT image

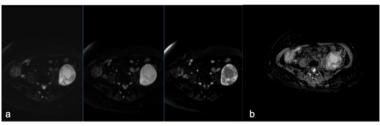


Fig 4. DWI images (b0,400,800) (a) and corresponding ADC map (b)

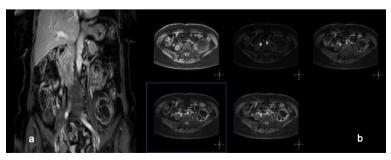


Fig 5. Portal venous coronal (a) and dynamic contrast enhanced (DCE) axial MR images (b)

PART B

Diagnosis: Ancient schwannoma of the left femoral nerve

Gray scale US (Fig 1a) depicts a circumscribed, oval mass of mixed echogenicity in the left flank, predominantly solid in the periphery with central cystic areas and septations. The mass shows a hypoechoic thick halo with minimal blood flow on colour Doppler US (Fig 1b).

On coronal CT images after intravenous contrast injection (Fig 2), a well-defined, ovoid, heterogeneous, hypodense, retroperitoneal mass is identified, located in the left paravertebral region, below the left kidney and in contact with the left psoas muscle, without signs of invasion. The lesion shows mild peripheral enhancement as well as intralesional enhancing septa. No associated free fluid, perilesional fat stranding or regional enlarged lymph nodes are demonstrated. Apart from the presence of pelvicalyceal cysts identified in the left kidney the rest of the examination is unremarkable.

Coronal T2 weighted images reveal an ovoid mass at the level of L4-S1 with a tail-like appendage extending from the proximal and distal aspect of the mass indicating the tumour's location along the left femoral nerve (Fig. 3). The mass demonstrates a thin peripheral hypointense capsule and internal heterogeneity with high T2 weighted signal intensity, probably corresponding to cystic areas, and low T2 weighted signal intensity thick, nodular septa. Diffusion Weighted Imaging (DWI) sequence shows restricted diffusion at the periphery and along the septa (Fig. 4). After gadolinium injection, a gradual enhancement of the circumference of the cystic areas and the tumour capsule is observed (Fig. 5).

The main differential diagnoses are of benign retroperitoneal solid lesions with cystic components and neurogenic origin including schwannomas, neurofibromas, paragangliomas, and ganglioneuromas.

Primary retroperitoneal tumours are rare with a prevalence of less than 1%. 80% to 90% of the cases are malignant, half of which are attributed to sarcoma [1]. Among benign lesions, schwannomas are rare with an estimated incidence between 0.5 to 3% [2]. When faced with the discovery of a retroperitoneal tumour, the absence of clinically significant signs and symptoms points towards the diagnosis of schwannoma which presents in most cases as an asymptomatic tumour discovered incidentally. Since their evolution is slow, they can reach large sizes, often measuring between 10 and 20 cm, and when symptomatic, this is mainly due to compression of adjacent structures. Laboratory data are generally not remarkable or contributory to diagnosis.

Due to their rare occurrence in the retroperitoneum (0.3%-3.2%), imaging findings of retroperitoneal schwannomas have not been extensively reported [3]. An ancient schwannoma is a rare variant, was first described by Ackerman and Taylor [4] and is usually located deep in the head and neck region, thorax, retroperitoneum, pelvis, and extremities of elderly patients [5]. It is characterized by degenerative changes typified by perivascular hyalinization, calcification, cystic necrosis, hemorrhage and hypocellularity with myxomatous and cystic changes. Sonography may reveal a well-defined mass of mixed echogenicity. An ancient schwannoma is depicted as a well-defined, ovoid or spherical, inhomogeneous low-density mass on CT [6]. Its heterogeneity reflects the varied histologic components of Antoni A (very cellular areas consisting of Schwann cell nuclei) and B (less cellular with myxomatous and cystic changes) areas and cystic degeneration [7]. MRI is the most valuable diagnostic radiological examination for its evaluation [8]. MRI is able to demonstrate the tumour's location along a nerve and eccentric to the nerve axis as well as its relationship to surrounding structures.

The treatment of choice for ancient schwannomas is complete surgical excision. Tumour enucleation, with preservation of nearby vital structures, is the preferred surgical approach, since these tumours are rarely malignant and do not tend to recur following excision [9].

As for our patient, since she was diagnosed right before the Covid-19 pandemic, she chose to defer surgery and have a follow-up examination, especially due to the fact that she did not have any symptomatology. An MRI performed 4 months later did not demonstrate any changes regarding size and morphological characteristics of the lesion and the patient remained asymptomatic as well.

In general, an ancient schwannoma has a smooth, regular margin, without signs of invasion or obstruction of surrounding structures. Ancient schwannomas are isointense on T1-weighted images relative to surrounding muscle and hyperintense on T2-weighted images [8, 10]. The intensity of the signal on T2 weighted images is inversely

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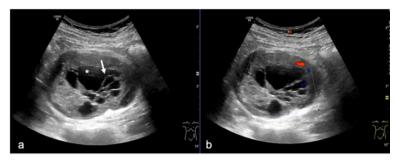


Fig 1. Gray scale US (a) and colour Doppler over the left flank area (b). A circumscribed, ovoid mass of mixed echogenicity, predominantly solid in the periphery with central cystic areas (*) and presence of septation (arrow) is identified. The mass shows a hypoechoic thick halo with minimal blood flow on colour Doppler US.



Fig 2. Portal venous coronal CT image demonstrates a well-defined, heterogeneous, low-density, retroperitoneal mass (arrowhead) with mild peripheral enhancement and intralesional enhancing septa (arrow) in the left paravertebral region, in contact with the left psoas muscle (p).

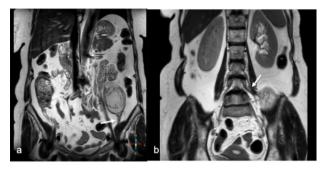


Fig 3. Coronal T2 weighted images reveal an ovoid mass at the level of L4-S1 with a tail-like appendage identified both at the distal (a, arrow) and proximal aspect of the mass (b, arrow). The mass demonstrates a thin peripheral hypointense capsule and internal heterogeneity with high T2 weighted signal intensity, probably cystic areas, and low T2 weighted signal intensity thick, nodular septa.

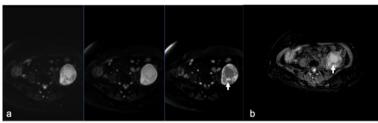


Fig 4. DWI images (b0,400,800) (a) and corresponding ADC map (b) show restricted diffusion at the periphery and along the septa (arrows).

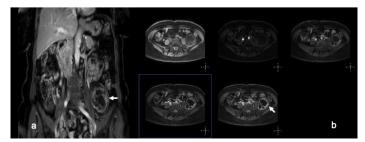


Fig 5. Portal venous coronal (a) and dynamic contrast enhanced (DCE) axial MR images (b) show a gradual enhancement of the circumference of the cystic areas and tumour capsule (arrows). 55

proportional to the cellularity of the tumour, as opposed to the noncellular and necrotic zones that accentuate the T2 high signal. Because it contains cystic areas, ancient schwannoma has been radiologically misdiagnosed as other malignant tumours, but the smooth, regular border of schwannomas due to their fibrous capsule allows them to be recognized for their benign nature. According to the findings of Isobe et al. [8], the enhancement of the circumference of the degenerated area as well as of the septa and fibrous tumour capsule on MR images are important imaging features for the discrimination of an ancient schwannoma from other tumours. The radiologic findings for ancient schwannoma are similar to those of ordinary schwannoma and the differential between them is difficult based only on imaging, although internal heterogeneity and the presence of calcifications are more frequently observed in ancient schwannomas. However, calcification may not be detected on imaging examinations.

To conclude, an ancient schwannoma should be considered when a large, asymptomatic, well-circumscribed heterogeneous mass with cystic changes following the route of a nerve is identified as isointense on T1 –weighted images and hyperintense on T2 -weighted MR images and demonstrates an enhancing fibrous capsule and degenerative areas with enhancing circumference. \mathbf{R}

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Key words

ancient schwannoma, femoral nerve, retroperitoneal schwannoma

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