RADIOLOGIC PATHOLOGIC CORRELATION / Gastrointestinal imaging

Retroperitoneal ganglioneuroma revealed as an ‘‘incidentaloma’’ in a healthy volunteer

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Case study

A 32-year-old male patient who was asymptomatic and had no medical history of note was initially a healthy volunteer in the research laboratory for a study using MRI. During this examination, a left retroperitoneal mass was revealed. A multiphase computed tomography (CT) acquisition of the chest, abdomen, and pelvis and a contrast-enhanced abdominal MRI were then subsequently carried out in order to pinpoint the location and identify the nature of this lesion.

The CT acquisitions confirmed the presence of a voluminous retroperitoneal mass measuring 96 × 84 × 55 mm situated against the medial border of the left kidney with no beak sign, excluding a potential renal origin (Fig. 1). The posterior surface of the lesion was directly adjacent to the left anterolateral border of the vertebral body and the left psoas muscle; it caused displacement towards the front and the top of the left renal hilum, which remained unobstructed. The medial border of the lesion was in contact with the aorta, with no displacement or infiltration. The left adrenal gland was of normal morphology. There were no further abnormalities within the abdomen, pelvis, or chest. Prior to contrast material administration, the lesion was hypodense, with no fatty contingent or calcifications. There was minimal uptake of contrast material.

The MRI confirmed the minimal uptake of contrast material after administration. On T1-weighted sequences, the mass showed homogeneous low signal intensity. On T2-weighted sequences, there was a component with high signal intensity (lateral part), and another component with isosignal intensity (medial part) (Fig. 2).

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The diagnoses of ganglioneuroma, paraganglioma, and seminoma were all suggested. The testicular sonogram was unremarkable. An FDG-PET scan showed the lesion to be moderately hypermetabolic. Germ cell tumour markers were negative, as was the catecholamine urine test.

We decided upon surgical excision, achieved by robot-assisted peritoneoscopy. Macroscopic examination of the mass confirmed that it was well delineated, and that it did have two components, one containing larger numbers of adipocytes than the other (the medial part). Microscopic examination demonstrated mature ganglion cells in both components within a stroma of Schwann cells and a few scattered microcalcifications, no necrosis or mitosis, pointing to a diagnosis of mature ganglioneuroma (Fig. 3).

**Discussion**

Ganglioneuroma is a rare benign tumour arising from cells of ectodermal origin that develops in the sympathetic nervous system. It is composed of mature ganglion cells (in contrast to neuroblastoma and ganglioneuroblastoma which are made up of immature ganglion cells) and a stroma made up of neural cells together with a Schwann cell component. Ganglioneuromas are most commonly seen in young adults [1]. They develop along the sympathetic chains, and this explains why their location is cervical, mediastinal, retroperitoneal, or pelvic. The retroperitoneum is a common site for ganglioneuroma (32–52%) [2,3], and one in two is extra-adrenal. They are often found incidentally because, in spite of their large size, retroperitoneal ganglioneuromas are generally asymptomatic. Sometimes abdominal pain, palpation of an abdominal mass, or compression of the neighbouring organs leads to diagnosis [1].

Ganglioneuroma has quite specific features on imaging: it is a well-circumscribed oval-shaped tumour with regular borders, highly homogeneous content, and minimal vascularisation; it does not invade the adjacent vascular structures, and is situated along the sympathetic chains; symptoms are minimal or absent, and it occurs in young or middle-aged patients. CT can demonstrate calcifications (seen in 50% of cases). There is no fatty or cystic component. There is minimal contrast material uptake. On MRI, ganglioneuroma shows homogeneous low signal intensity on T1-weighted images and high or isosignal intensity on T2-weighted images, depending on the amount of stroma contained in the lesion [4]. In the vast majority of cases
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Figure 2. Abdominal MRI. a, b: coronal and axial T2-weighted views: lateral part of the left retroperitoneal mass shows high signal intensity, medial part shows isosignal intensity; c: T1-weighted sequences with fat-saturation: homogeneous low signal intensity; d: minimal contrast uptake after gadolinium administration.

Figure 3. Macroscopy of excised lesion (a, b) and microscopic examination (haematoxylin eosin saffron stain) (c) (×10) and (d) (×40) of the lesion. a, b: voluminous well circumscribed nodule dissected in two, measuring 9 × 4 × 6 cm, homogeneous, yellowish beige, firm, non-lobulated. Adipose tissue appended to the lesion; c, d: mature ganglion cells (white arrow), within a stroma of Schwann cells (black arrow). No necrosis or mitosis.
hormonal assessment is normal (with, in rare cases, secretion of catecholamines or VIP being reported [3]). The treatment options are either monitoring or surgical excision.

There are numerous differential diagnoses for a large retroperitoneal tissue lesion developing between normal structures [5], but when the semiological nuances are taken into account together with the context in which the lesion was discovered, this will assist in narrowing down the list to consider. This means that when a solid and homogeneous mass is seen in an asymptomatic patient, the only plausible diagnoses are unicentric hyaline-vascular type Castleman disease or non-secreting paraganglioma if the tumour is hypervascularised [6], and ganglioneuroma if the lesion has minimal vasculature, which was the case here.

Other diagnoses must of course be suggested, but they will immediately be excluded for semiological or epidemiological reasons. Among the benign tumours to exclude, teratomas and lipomas have a fatty component, schwannomas often have a cystic component, desmoid tumours are poorly circumscribed, cystic lymphangiomata contain fluid, and retroperitoneal seminoma (burned-out tumour) is accompanied by deterioration of the patient’s general condition. Malignant lesions (primary, secondary, lymphomatous) are multiple, often necrotic, and invade the adjacent structures.

Diagnostic investigations must therefore on principle include all of the following: PET-CT to demonstrate the unifocal nature of the lesion, an essential step in the path to diagnosis, a testicular sonogram, hormonal assessment (catecholamine test), and tumour marker tests.

Finally, a sound assessment bringing together the clinical context and the imaging features in a single, homogeneous, retroperitoneal mass with minimal vasculature can only lead to a diagnosis of mature ganglioneuroma. It would therefore be possible to refrain from any treatment, and simple monitoring would be sufficient. If there is any doubt, a CT-guided biopsy will have its place; in our case, however, the surgical excision carried out from the outset was ultimately able to back up the radiologist’s diagnosis because optimum histological analysis was possible.

Disclosure of interest

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

References