CASE NOTE

Retroperitoneal nonfunctioning paraganglioma

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Paragangliomas are extra-adrenal chromaffin tumours[1] that develop at the expense of neuroectodermal cells of the autonomous nervous system.[2] Retroperitoneal and nonfunctioning forms are very rare.[3] They are often asymptomatic and can reach a substantial size. Treatment usually involves surgery with the goal of total excision. We report the case of a patient who presented with indistinct abdominal pain. This case demonstrates how the diagnosis of retroperitoneal nonfunctioning paragangliomas relies mostly on histological results.

Case report

A 55-year-old woman presented to us with indistinct abdominal pain and a sensation of heaviness of 2 months' duration. She reported no digestive difficulty. She had been postmenopausal for 10 years, was being monitored for hypertension (2 years) and had undergone a cholecystectomy 3 months earlier. On clinical examination, we found a mass on her left side that was firm and slightly tender. An abdominal computed tomography scan showed a well-defined round mass of heterogeneous density that was touching the aorta and the left kidney pedicle (Fig. 1). The results of tests for methoxyl derivatives were negative. During surgery with a median approach, we found a retroperitoneal tumour of about 6 cm diameter that was encapsulated and soft. It was adhered to the aorta, to the left renal vein at its base and to the body and tail of the pancreas at the top. We conducted a complete excision of the tumour (Fig. 2).

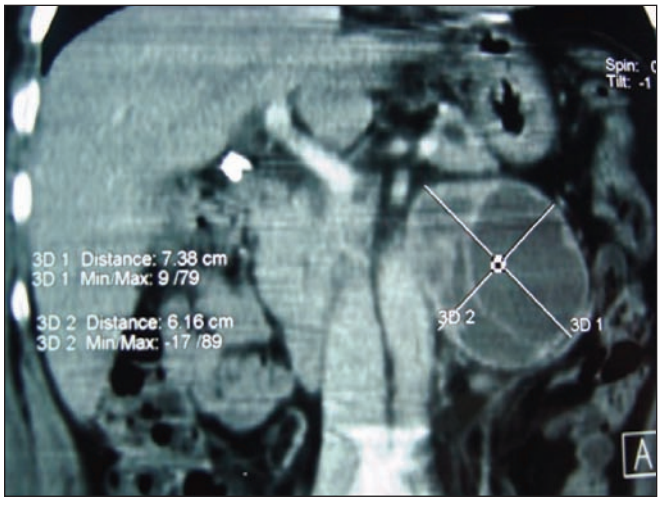

Fig. 1. An abdominal computed tomography scan in the transverse plane shows a round mass that is heterogeneously dense and well-defined, touching both the aorta and left kidney pedicle.

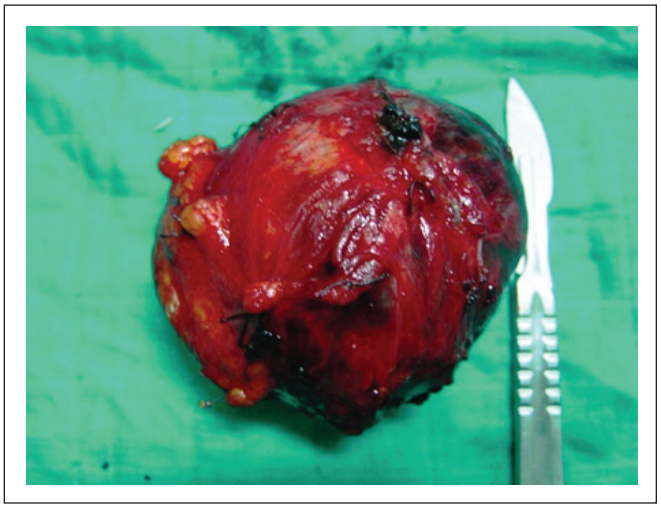

Fig. 2. The excised tumour.

Discussion

Retroperitoneal paragangliomas are rare tumours. In 60% of cases, they secrete hormones and are classified as pheochromocytomas. Cancerous forms represent 20%–50% of cases and are characterized by the occurrence of ganglionic or distant metastases in 30% of cases.[3] Nonfunctioning retroperitoneal forms are even more rare and are most often isolated.[4] They are characterized by their asymptomatic profile and normal levels catecholamines in the urine and blood. Patient age at diagnosis is usually about 40 years, with a sex ratio close to 1.[3]

Owing to their location, nonsecreting character and slow growth, retroperitoneal nonfunctioning paragangliomas are clinically latent tumours. Nevertheless, patients will sometimes present with nonspecific symptoms such as lower back pain, abdominal heaviness (as in our patient), urinary symptoms or changes in their general condition.[4]

Ultrasound scans show a well-defined round or oval mass with central zones of cystic appearance. Computed tomography scans typically show a solid round or oval mass that is homogenous but that may have a cystic or necrotic appearance at its centre, or it may appear calcified.[3] Angiography results are nonspecific, showing an arterial honeycomb appearance and tardive arteriovenous shunts.

A definitive diagnosis can be reached only by histology,[4] and there are no histologic criteria to distinguish between benign and cancerous tumours. Only the appearance of distant metastases, metastases at a site where paraganglionic tissue is not usually found or local recurrence can confirm that the tumour is cancerous. However, total excision is the basis of curative treatment because these tumours are potentially malignant.[5]

Radiotherapy can be used as an analgesic in spinal metastases or in an effort to sterilize tumour remnants after surgery. Chemotherapy may be considered for metastatic forms, but these therapies do not influence overall prognosis.[3]

Follow-up using scintigraphy with metaiodobenzylguanidine enables the detection of metastases or recurrence.

Competing interests: None declared.

References

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