

LETTER / *Gastrointestinal imaging*

Thrombosis of the superior mesenteric vein revealing an ectopic pheochromocytoma of the organ of Zuckerkandl

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KEYWORDS

Mesenteric venous thrombosis;
Pheochromocytoma;
Organ of Zuckerkandl;
Hypertension

Observation

A 43-year-old man was admitted to a hospital emergency unit for an acute painful abdominal condition with vomiting. His history included hypertension (HT) treated for the previous 5 years with a beta-blocker, excess weight, non insulin-dependent diabetes and active chronic smoking estimated as 30 pack-years. Clinical examination found diffuse abdominal sensitivity without guarding or hyperthermia. Initial laboratory results showed hyperleucocytosis of 14,000/mm³ combined with a rise in CRP to 90 mg/L. Renal function was normal. An abdominopelvic computed tomography (CT) scan was performed without, and then with, injection of iodinated contrast agent in the portal phase. It showed a right lateral-aortic retroperitoneal tumour mass with a regular outline situated anterior to the inferior vena cava at the posterior-lateral part of the inferior duodenal flexure. This lesion measured 61 × 50 × 54 mm with fine intratumoral calcifications. It was intensely enhanced following injection of iodinated contrast agent with no necrotic/haemorrhagic changes (Fig. 1a). The adrenal glands were normal. There was also extensive thrombosis of the superior mesenteric vein. Some of the jejunal loops had signs of mesenteric

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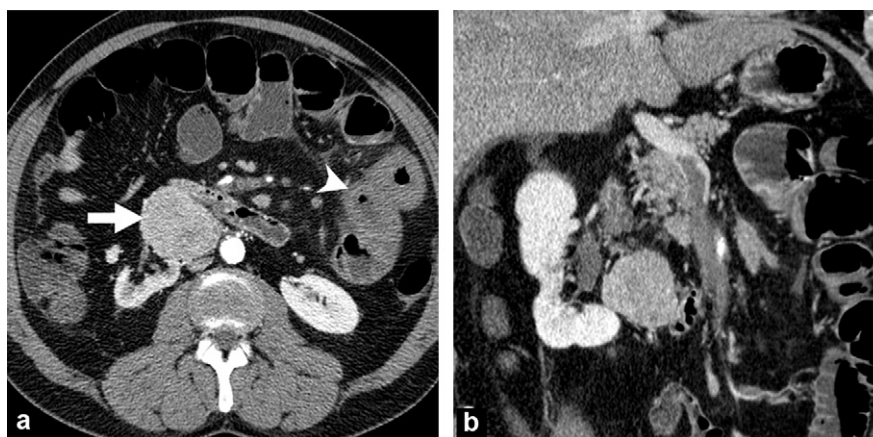


Figure 1. Abdominopelvic CT with injection of iodinated contrast agent: portal phase acquisition: a: axial slice: right lateral-aortic hyper-vascular tumour mass in the organ of Zuckerkandl (arrow). Small intestine loop with wall thickening (arrowhead); b: coronal reconstruction: hypodense defect in the superior mesenteric vein corresponding to a thrombosis.

venous ischaemia (parietal thickening with target-like enhancement of the wall) (Fig. 1b). Oesophagogastro-duodenal fibroscopy showed extrinsic compression of the duodenum at the inferior flexure without visible mucosal abnormality. Tests for hypercoagulability proved negative. The 24-hour urinary concentration of metanephrines was 2.5 times higher than the normal level and the chromogranin A level was six times higher. Scintigraphy using iodine 131 labelled metaiodobenzylguanidine (MIBG) showed intense radiotracer uptake by the right retroperitoneal mass with no other distant suspect focus of hyperfixation (Fig. 2). An ectopic pheochromocytoma of the organ of Zuckerkandl was therefore strongly suspected. Genetic research for associated hereditary conditions (multiple endocrine neoplasia type 2, von Hippel Lindau disease, neurofibromatosis type 1, familial paraganglioma) was negative. An effective dose of anticoagulation was initiated immediately. Later, complete surgical enucleation of the tumour took place after performing a full cardiorespiratory examination and suitable



Figure 2. Scintigraphy with iodine 131 labelled metaiodobenzylguanidine (MIBG): intense radiotracer uptake by the tumour.

anaesthetic preparation with drug blockade of adrenergic receptors.

Histological examination confirmed the diagnosis of pheochromocytoma. The ablated material measured $7 \times 6 \times 4$ cm and weighed 82 grams. There was no evidence of necrotic changes. Immunohistochemical examination showed that the cells expressed chromogranin and synaptophysin. Peripheral supratentacular cells expressed the S-100 protein (Fig. 3).

Discussion

Pheochromocytomas are rare primitive tumours secreting catecholamines which develop in 80–85% of cases in the adrenal glands. In 15 to 20% of cases, pheochromocytomas are ectopic and are also known as paragangliomas. In the abdomen, the most frequent extra-adrenal location is the organ of Zuckerkandl in the retroperitoneum. This organ corresponds to the sympathetic ganglia nerve chains around the aorta between the superior and inferior mesenteric arteries [1,2]. Pheochromocytoma is part of a hereditary condition in 24% of patients. Germline mutations have been identified for five genes (*VHL*, *RET*, *NF1*, *SDHB*, *SDHD*), associated respectively with von Hippel Lindau disease, neurofibromatosis type 1, multiple endocrine neoplasia type 2 and familial paragangliomas [1–3]. These mutations were not found in our patient.

The symptoms usually encountered are related to the secretion of catecholamines by the tumour. HT is present in 90% of cases and may develop paroxysmally depending on trigger factors (miction, defaecation). Pheochromocytoma is responsible for 0.1% of secondary HT. Ménard's classic symptomatic triad associates headaches, sweating and palpitations. The HT is sometimes malignant and associated with serious cardiovascular complications (acute pulmonary oedema, shock, cerebrovascular accident, myocardial infarction). Diabetes is found in a third of cases. In 25% of cases, pheochromocytoma has few symptoms or is discovered by chance [1]. Determination of urinary methoxylated derivatives and plasma catecholamines are the most sensitive biochemical tests for

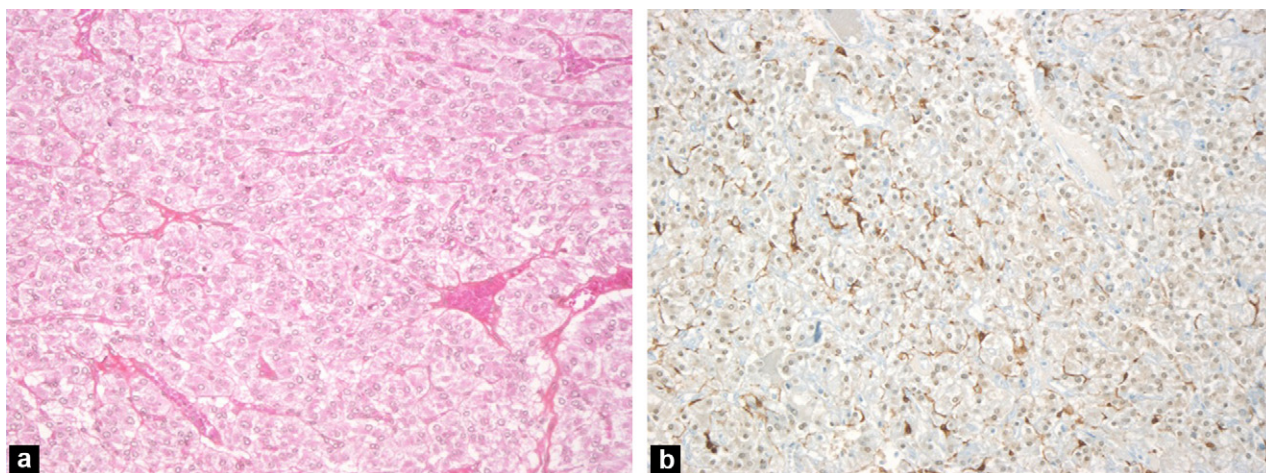


Figure 3. Histological examination of the ablated material under an optical microscope: a: lobulated endocrine architecture (HPS \times 25); b: labelling of supratentacular cells by anti-protein S-100 antibodies (\times 25).

diagnosing pheochromocytoma, with 97% and 99% sensitivity respectively [4,5]. CT and magnetic resonance imaging (MRI) perform similarly in terms of tumour detection [1,6]. CT allows the whole of the abdominopelvic cavity to be explored with better spatial resolution but has the disadvantage of being a technique involving irradiation. If pheochromocytoma is suspected, MRI can be performed as a first course of action because it detects lesions well – they appear as hypersignals in T2-weighting – and has the advantage of not involving irradiation. Tumour enhancement is very intense after injection of a contrast agent. Enhancement of large lesions (more than 3 cm in diameter) is heterogeneous and related to necrotic/haemorrhagic changes. The specificity of scintigraphy using iodine 123 or 131 labelled MIBG is close to 100% in terms of characterising a mass detected by CT or MRI. In the initial assessment, it is also performed to detect multifocal lesions or secondary locations of a malignant lesion. Later, it is useful for detecting tumour recurrence where this is suspected. Lumachi et al. suggest undertaking MRI exploration and MIBG scintigraphy where there is a suspicion of pheochromocytoma with sensitivity and positive predictive value close to 100% [6]. In our patient, the way the pheochromocytoma was revealed was unusual and is hardly described in the literature. In most cases reported, patients present a picture of acute mesenteric ischaemia originating from low arterial flow [7,8]. Brauchlin et al. reported a case of pheochromocytoma in a 51-year-old man in a state of shock with multi-organ failure and portal vein thrombosis [9]. The association of pheochromocytoma and extensive thrombosis of the superior mesenteric vein is unusual. With rapid administration of effective anticoagulation the condition evolved favourably with reduction in the CT signs of mesenteric venous ischaemia [10–12]. In our case, hypercoagulability was completely assessed and did not show any evidence of physiological predisposition [13]. The hypothesis of splanchnic vasoconstriction with reduction in the mesenteric flow may explain the formation of a mesenteric venous thrombosis. On the other hand, a paraneoplastic mechanism is also possible.

Conclusion

Pheochromocytoma is a rare endocrine tumour usually affecting the adrenal glands, and secreting catecholamines. In the abdomen, the most frequent ectopic location is the organ of Zuckerkandl. The association of pheochromocytoma and thrombosis of the superior mesenteric vein is an unusual means of the condition being revealed.

Disclosure of interest

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

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