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Mesenteric paraganglioma's: an important differential diagnosis in intra-abdominal tumours

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DESCRIPTION

We present the case of a 63-year-old lady who complained of feeling constantly tired and lethargic, and was found to have iron deficiency anaemia. There was no history of per vaginal bleeding. Examination was normal apart from a large, mobile, firm, well-defined abdominal mass in the upper abdomen approximately 10cm in diameter. The abdominal CT scan revealed a large intra-abdominal tumour (figure 1). Histology of the specimen obtained by surgical resection revealed this to be a mesenteric paraganglioma (figure 2). To the best of our knowledge, there are eight other reported cases of mesenteric paraganglioma's. Ninety per cent of these sympathetic tissue tumours arise from the adrenals and are called phaeochromocytomas. Paraganglioma's are extra-adrenal phaeochromocytoma's, of which 85% arise in the abdomen, 12% in the thorax and the remaining 3% in the head and neck.[1] Paragangliomas that hypersecrete catecholamines may cause signs and symptoms identical to those in patients with hyperfunctioning adrenal pheochromocytoma. Recent studies implicate genetic factors in up to 25% of cases. The most frequent causes of phaeochromocytoma susceptibility are von Hippel–Lindau disease, multiple endocrine neoplasia type 2, and the newly delineated phaeochromocytoma–paraganglioma syndrome.

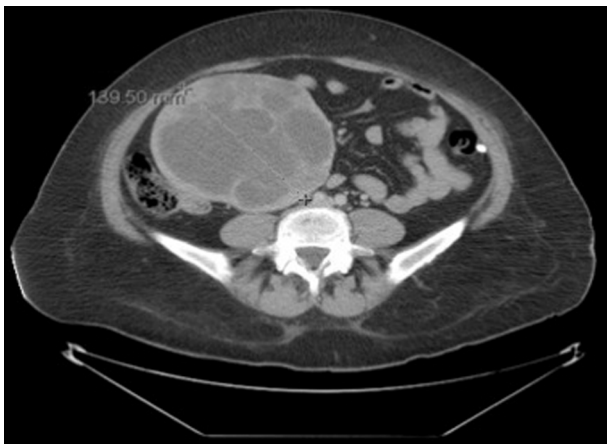

Figure 1 CT scan of the abdomen with intravenous contrast showed a 139×125 mm well-circumscribed mixed cystic/solid mass arising within the peritoneal cavity, to the right of the midline, displacing bowel loops. Posteriorly it was closely related to the inferior vena cave and anterior margin of the right psoas.

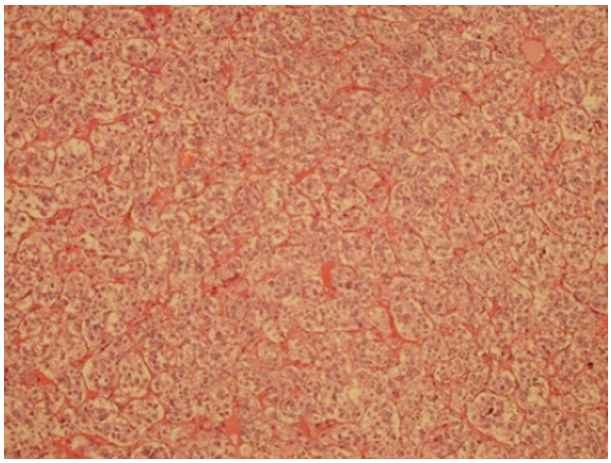

Figure 2 Classic nesting pattern (Zellballen) of paraganglioma – H&E staining, 100X magnification.

Germline mutations in three of the succinate dehydrogenase subunits (SDHD, SDHB and SDHC) increase susceptibility to head and neck paragangliomas, and familial and isolated phaeochromocytomas.[2] Differentiation between benign and malignant forms is difficult by imaging alone. There are no definite documented criteria for classification as a malignant form apart from the presence of distant metastases, which must be proven with biopsy, because paragangliomas may exhibit multicentricity.[3]

Learning points

- Keep in mind the possible differential diagnoses at all times, as even specialised imaging such as contrast CT scans may not always provide the answer.
- Excision of paraganglioma's is the treatment of choice after optimisation of the patient. Formal follow-up protocol, however, is yet to be established in view of the limited number of cases.
- Genetic testing should be considered in all patients presenting with paraganglioma's.

Competing interests None.

Patient consent Obtained.

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