Mesenteric paraganglioma

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Paragangliomas are uncommon tumours that have been described in virtually every site in which normal paraganglia are known to occur. Paragangliomas of the carotid body represent the most common group of extra-adrenal paragangliomas. The other less common sites include jugulotympanic, vagal, mediastinal and retroperitoneal. Paragangliomas have been described in the gastrointestinal tract especially in the duodenum. Paraganglioma as a mesenteric mass is extremely rare and only occasional reports have been published.[1–3] Our case of mesenteric paraganglioma in a 76-year-old woman is probably only the fourth published case.

Case report

A 76-year-old woman, known to have diabetes mellitus and osteoarthritis, presented with a painless mass of 2 years' duration, which was gradually increasing in size. Examination of abdomen revealed a well-circumscribed mass, measuring 20 cm × 15 cm, that was firm, nontender and mobile. General physical examination and the rest of the systemic examination revealed no other abnormality. Ultrasonography and computed tomography showed a solid, cystic mass with increased vascularity peripherally that appeared to arise from the small-bowel mesentery. Exploratory laparotomy confirmed the presence of a large, solid, cystic mass in the mesentery of the small bowel. The mass was excised. The patient was well without evidence of recurrence or metastasis at 15-month follow-up.

Grossly, the specimen consisted of a round, multilobulated cystic mass filled with blood clots. At the periphery, the lesion was solid and fleshy. Microscopic sections revealed a cellular neoplasm composed of nests and groups of tumour cells separated by fibrovascular connective tissue, giving a characteristic "zellballen" nested pattern. The tumour cells showed monomorphic vesicular nuclei without significant cytologic atypia. Cytoplasm was moderate and granular. Occasional mitotic figures were identified (Fig. 1). Immunohistochemical studies revealed the presence of chromogranin A, synaptophysin and S-100 protein and the absence of cytokeratins, MNF 116 and anticytokeratin Cam 5.2. On the basis of histologic and immunohistochemical features, a diagnosis of mesenteric paraganglioma was made.

Discussion

Extra-adrenal paragangliomas are rare neoplasms. They may develop anywhere from the neck to the pelvis; however, mostly they arise intra-abdominally.[4] Generally, the tumours develop in the third to fifth decade of life.[5] Increased catecholamine secretion is responsible for presenting symptoms in at

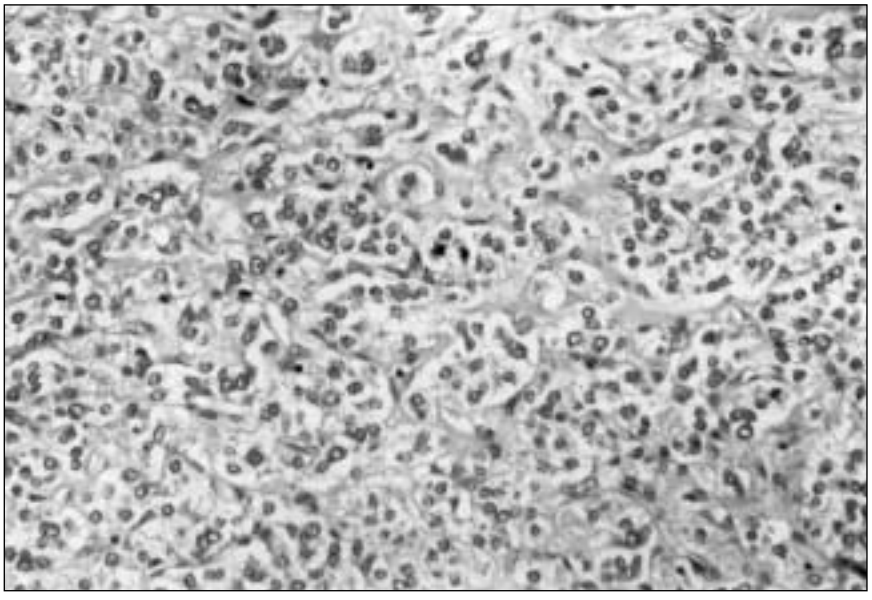

FIG. 1. Photomicrograph of the mesenteric mass. Note the typical "zellballen" arrangement of tumour cells (hematoxylin–eosin stain, original magnification × 200).

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least 25% of the cases.[6] These extra-adrenal paragangliomas tend to behave more aggressively than others, and the incidence of malignant change has been reportedly ranged from 14%[7] to 50%.[6] In the present case, the paraganglioma presented as a gradually increasing abdominal mass of 2 years' duration that did not produce any systemic or local obstructive symptoms. The case probably represents the fourth report of this condition in literature.

The recognition of this tumour as a cause of an abdominal mass is extremely important both for surgeons and pathologists. Preoperative image-guided fine-needle aspiration or intraoperative consultation may help an operating surgeon to plan the surgical approach. This lesion could be a true diagnostic dilemma for a pathologist; however, cytologic appreciation of neuroendocrine features of the tumour cells may help to avoid an erroneous diagnosis of carcinoma or sarcoma.

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