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INTRAPERICARDIAL PARAGANGLIOMA

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Key-word: Paraganglioma

Background: A 38-year-old woman presented for a follow-up CT of the neck and the chest. She had a history of familial disease and neck surgery 11 years earlier. Physical examination and laboratory results were normal. An intrapericardial lesion was suspected on a thoracic MR examination 5 years earlier.
Unenhanced CT scan, contrast-enhanced CT scan and scintigraphy (Octreotide scan) of the thorax were carried out.

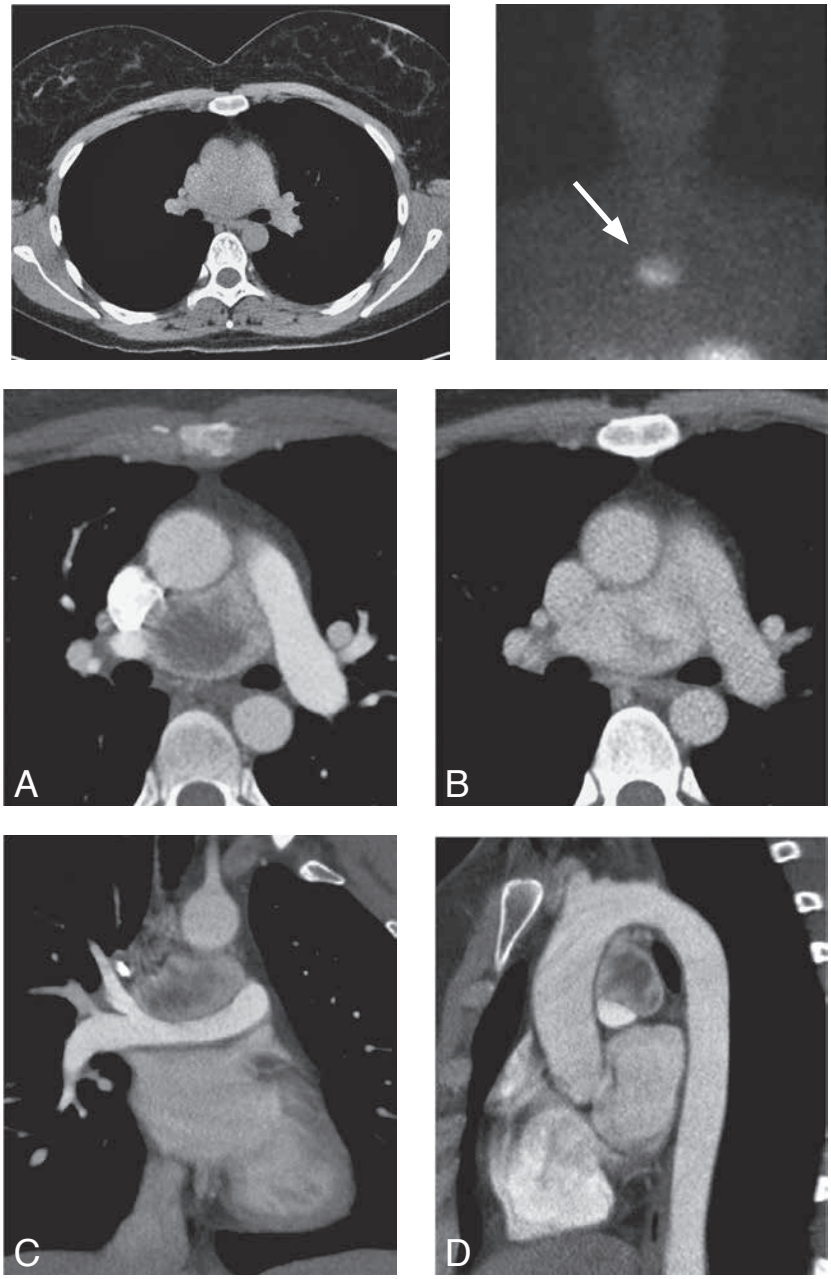

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Fig.	2A	2B
	2C	2D

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Work-up

Unenhanced CT scan of the mediastinum, section through the pulmonary arteries (mediastinal window setting) (Fig. 1) shows almost normal findings. In particular, no abnormal calcification is demonstrated.

Contrast-enhanced CT scan of the mediastinum (Fig. 2) consists of axial sections (A,B) which show a 44x25x34-mm heterogeneous tumor with intense peripheral and centripetal enhancement. The lesion is located just above the right pulmonary artery, posterior to the ascending aorta and medial to the superior vena cava. On reformatted images in the coronal (C) and sagitttal-oblique (D) plane, the tumor presents with a mass effect on the right pulmonary artery. The sagittal-oblique view demonstrates particularly well the close relationship between the tumor and the roof of the right pulmonary artery. This prompted the decision to operate the patient under extracorporeal circulation. At surgery, delineation between the tumor and the pulmonary artery was difficult. A breach of the artery eventually occurred without any complication thanks to the extracorporeal circulation.

Scintigraphic examination with 111In-diethylenetriaminepentaacetic acidoctreotide (Octreotide) (Fig. 3) demonstrates intense fixation in the lesion suggesting the presence of a large amount of receptors for somatostatin. Note also a limited fixation in the left cervical area.

The patient had a history of left tympano-jugular paraganglioma resected 11 years previously.

Radiological diagnosis

Based on the imaging findings and clinical history, the diagnosis of *intrapericardial paraganglioma* was made.

Discussion

Chromaffin (neuroendocrin cells found in the adrenal medulla and in sympathetic ganglia) cell tumors include pheochromocytomas (90%) that are located in the adrenal, and paragangliomas (10%) found in extra-adrenal locations. Paragangliomas are sporadic or observed in familial diseases (Von Hippel-Lindau, MEN2, NF1). Paragangliomas are locally invasive tumors, with local recurrence rate of 56%-71% and distant metastasis rate in 19%-27%.

Mediastinal paragangliomas represent 0,3% of the mediastinal tumors. They can be located in the middle mediastinum (two-thirds) arising from the perivascular adventitial tissue between aorta, pulmonary artery and ligamentum arteriosum, or exceptionally within the pericardium, and in the posterior mediastinum (one-third) arising from the sympathetic chain ganglia.

Most of mediastinal paragangliomas are non functional and will produce signs and symptoms related to compression or invasion of mediastinal structures. When functional, paragangliomas present with symptoms secondary to catecholamines secretion, including hypertension, headache, palpitation, sweats, and tremor. Diagnosis is made by biochemical tests, plasma metanephrine and/or urinary catecholamine metabolites. At CT scan, paragangliomas appear as soft tissue masses, usually larger than 3 cm, and intensely enhance after IV administration of iodinated CM. They are homogeneous when small, or heterogeneous due to necrosis when large.

On MRI, paragangliomas are iso-intense to muscle on T1-weighted images, hyperintense on T2-weighted images, with serpiginous vascular channels and cystic areas of necrosis or hemorrhage within larger lesions. Scintigraphy with MIBG or Octreotide (somatostatin receptor scintigraphy) is essential for the differential diagnosis of paragangliomas from other tumors but these techniques present limitations because of the variable tumor differentiation and the heterogeneous expression of somatostatin receptors. Nevertheless, the sensitivity of MIBG-scintigraphy is of approximately 90%. The imaging techniques of choice for paragangliomas are CT or MRI combined with MIBG-scintigraphy, except for primary tumors in the head and neck where MRI and somatostatin receptor scintigraphy are more sensitive. The paragangliomas can be metabolically active on 18-FDG-PET/CT.

Complete surgical resection is the treatment of choice. For functional paragangliomas, the hypertension must be treated before surgical intervention. When the diagnosis is confirmed preoperatively, tumor embolisation angiography can be performed in order to reduce intraoperative bleeding. Cardiopulmonary bypass is recommended if the tumor is close to vessels or heart to reduce the risk of bleeding and of hypertensive crisis due to intraoperative manipulation.

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