Short reports

Mediastinal paraganglioma presenting as an intracardiac mass with superior vena caval obstruction

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Abstract

A case of mediastinal paraganglioma presenting with superior vena caval obstruction is reported. The tumour extended into the right atrium and ventricle. Tru-Cut biopsy under ultrasonographic guidance was performed safely to provide a diagnosis before death.

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Mediastinal paraganglioma is a rare tumour of the aortic bodies. Superior vena caval obstruction arising from mediastinal paraganglioma is extremely rare. Very few cases of intracardiac paragangliomas have been described, and intracardiac paraganglioma presenting with superior vena caval obstruction has never been described before.

Case report

A 52 year old housewife presented with "heaviness of the head," progressive swelling of the face and neck, dyspnoea on exertion, dysphagia, and hoarseness of voice. She had been well until 18 months before presentation when she noticed "heaviness in the head" on bending forwards. She also gave a history of anorexia, epigastric discomfort after meals, and had lost 20 kg in weight over 18 months. She was a non-smoker and had no significant occupational exposure. The past and family history were non-contributory.

Examination revealed a middle aged lady with obvious swelling of face and neck. The neck veins were distended but non-pulsatile. Prominent veins were noted over the anterior chest and abdomen. There were no palpable lymph nodes. The blood pressure was 120/70 mm Hg. Examination of the respiratory system revealed dullness of percussion and tubular breath sounds in the parasternal region. Soft, smooth, non-tender 5 cm hepatomegaly with a 2 cm palpable spleen was present. There was no evidence of free fluid in the abdomen.

A clinical diagnosis of an anterior mediastinal tumour causing superior vena caval obstruction was made. Chest radiography and a computed tomographic scan (fig 1) showed a large lobulated enhancing anterior mediastinal mass with areas of dense calcification. The mass extended into the superior vena cava and right atrium. The inferior vena cava was dilated and a small pericardial effusion was noted. The extension of the tumour into the right atrium and ventricle was confirmed by two dimensional echocardiography. A Tru-Cut biopsy from the mediastinal part of the mass was performed under ultrasonographic guidance without complications.

Tumour tissue was fixed in 10% buffered formalin, routinely processed and paraffin embedded, and 5 μm sections were cut and stained with haematoxylin and eosin.

The tumour was composed mainly of compact alveolar cell clusters separated by delicate connective tissue septae (fig 2) containing thin walled capillary blood vessels. In a few areas the cells were arranged more diffusely. They had round to oval nuclei with ill defined pale eosinophilic cytoplasm. Pleomorphism was evident but mitoses or necrosis were not observed.

Immunohistochemical stain by the avidin biotin conjugate (ABC) immunoperoxidase method was performed with anti-neurone specific enolase (NSE) antibody (Dako Corporation, USA). The tumour cells were positive for NSE.

On the basis of the above histological and immunohistochemical findings a diagnosis of paraganglioma was made.

The patient refused further treatment. She was discharged on her request and lost to follow up.

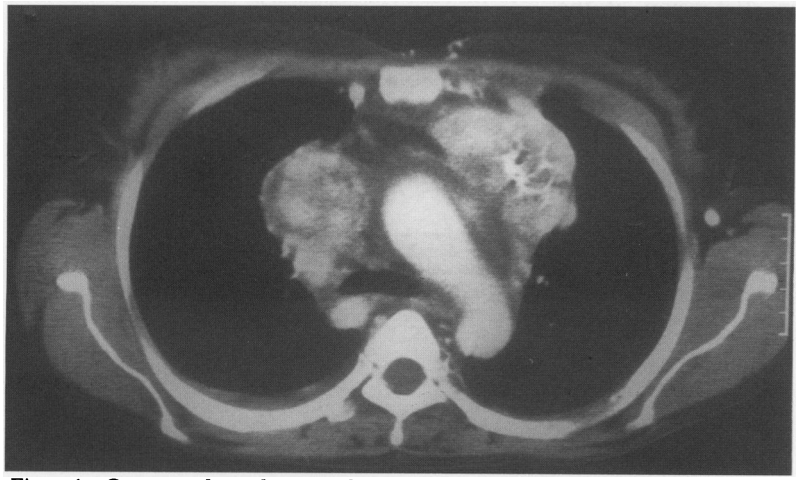

Figure 1 Contrast enhanced computed tomographic scan of chest showing a large lobulated mass, with dense calcification, anterior to the arch of the aorta. The superior vena cava is grossly dilated and filled with the mass. The dilated azygos vein can be seen.

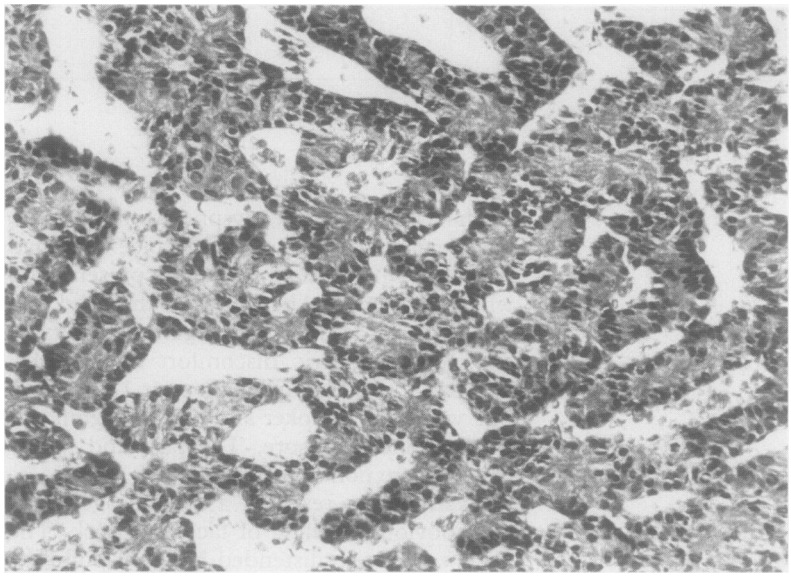

Figure 2 Photomicrograph showing clusters of tumour cells separated by delicate vascular connective tissue septa. Stain: haematoxylin and eosin. Original magnification × 450, reduced to 63% in origination.

Discussion

Paraganglioma (also known as non-chromaffin paraganglioma, chemodectoma) of the mediastinum is an extremely rare tumour, fewer than 100 cases having been reported in the English literature. Extra-adrenal paragangliomas arise from neural crest cells. They have been classified into four groups: (a) branchiomeric (associated with the great vessels of the thorax and neck); (b) intravagal; (c) associated with the thoracolumbar sympathetic chain; (d) in association with other visceral organs.

The tumour is asymptomatic in about one half of the patients and is revealed on routine chest radiography. Symptomatic patients present with hoarseness, dysphagia, chest discomfort, and cough. The superior vena caval obstruction as a result of invasion by the tumour is rare, despite the proximity of this vessel to the origin of these tumours. Only four cases of superior vena caval obstruction have been described among the 90 cases of mediastinal paragangliomas reported.[1-4] In our case the tumour had invaded the superior vena cava from the mediastinum leading to obliteration of its lumen. Extension of the tumour into the right atrium and ventricle had produced a free floating mass inside these chambers.

The heart is an unusual site for this tumour, only six cases of paraganglioma involving the heart having been described,[5-10] four of which were found to lie within the pericardium.[6-9] Only two intracardiac tumours have been reported[6 9] and both involved the interatrial septum. One of these produced a mass within the right atrium.[5]

The localisation of mediastinal paragangliomas is possible with non-invasive imaging techniques such as echocardiography, computed tomography, or magnetic resonance imaging. The presence of calcification, as in our case, may not necessarily discourage the diagnosis. The diagnosis, accomplished at surgery or post mortem examination, usually comes as a pathological surprise because of the rarity of these tumours.

Since these tumours are highly vascular and bleed profusely at operation it is hazardous to obtain Tru-Cut biopsy samples. In our case, however, we unsuspectingly performed this procedure without complications.

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