Case report

Extrapericardial solitary fibrous tumour of the pericardium

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Abstract

Solitary fibrous tumour (SFT) occurs most commonly in the pleura and is extremely rare in the pericardium. The authors report a case of a 60-year-old man in whom a large mediastinal mass was accidentally discovered. Computed tomography showed involvement of the left anterosuperior mediastinum with displacement of the trachea, large vessels and oesophagus; histopathological findings after complete resection of the neoplasia demonstrated an SFT of the pericardium, the first reported case with extrapericardial pattern of growth. A review of the literature on SFTs of the pericardium is provided.

Keywords: Solitary fibrous tumour; Localized mesothelioma; Pericardial tumour

1. Introduction

Solitary fibrous tumour (SFT), so-called localized fibrous tumour or fibrous mesothelioma, was first described as a distinct entity in 1931 by Klemperer and Rabin [1]. SFT is a rare disease: from 1942 about 700 cases of SFT were described, in 85–90% of cases it arose from pleura [2], but it rarely occurred in other sites as mediastinum [3], peritoneum [2,4], retroperitoneum [1] and lung [2]. We present here the fifth reported case of SFT arising from pericardium [4–7].

2. Case report

A 60-year-old man with no history of occupational exposure to asbestos was referred to our department for a left mediastinal mass accidentally detected on a routine preoperative chest X-ray for herniated intervertebral disc.

On admission the patient had no complaints except for a slight exertional dyspnoea. Physical examination showed bilateral distension of the jugular veins with no evidence of a superior vena cava syndrome.

Chest X-ray showed a mass occupying the anterosuperior mediastinum and the left hemithorax, with compression and displacement of the trachea. On CT scan (Fig. 1) a large anterosuperior mediastinal mass displaced the oesophagus, the left common carotid and left suclavian artery and the trachea, whose lumen was reduced. Trans-thoracic fine needle aspiration biopsy detected a spindle cells tumour without evidence of malignancy.

The patient underwent a median sternotomy; resection of the left innominate vein, which proved to be incorporated and inseparable from the tumour at its origin, was required.

The superior vena cava, the left common carotid and left suclavian artery and the trachea were carefully dissected.

The left lung was atelelatic for the compression but not infiltrated; the left mediastinal pleura and residual thymic tissue were resected en bloc with pericardium, left vagus and phrenic nerve. No residual tumour was left at the end of the exeresis. A resternotomy was necessary for bleeding a few hours later; no further complications occurred in the post-operative period.

Four years later the patient is asymptomatic, with no evidence of local or distant recurrence.
Grossly, the neoplasia appeared as well circumscribed, oval, firm mass, of 20 × 18 cm in greater diameters and 1615 g in weight, with homogeneous, gray-white cut surface, with occasional cystic changes and no evidence of necrosis or haemorrhage. The mass arose from a portion of fibrous tissue referred by the surgeon as pericardium.

Histologically the neoplasia was characterized mainly by a spindle cell proliferation, which was arranged haphazardly or as short fascicles, in a richly collagenous background (Fig. 2). The neoplastic cells displayed vesicular nuclei with a sharply demarcated nuclear membrane, finely dispersed chromatin and inconspicuous nucleoli; the cytoplasm
was scant, poorly defined, and slightly eosinophilic. Vari-
ably sized cyst-like spaces lined by spindle cells and cellular
areas composed of closely packed spindled or oval cells
with increased nuclear size were also present. Mitoses
were rare, with an average of two per ten high-power fields.

Immunocytochemically the neoplastic cells showed dif-
cuse cytoplasmic immunoreactivity to vimentin and CD34.

Tumour cells were consistently negative for cytokeratin,
epithelial membrane antigen, carcinoembryonic antigen,
desmin, actin and S100 protein.

3. Discussion

SFT most often occurs in the pleura [2], 80% in the vis-
ceral and 20% in the parietal pleura [8]. Initially it was
thought to be of mesothelial origin [9]: in many papers it
is also called fibrous or localized mesothelioma. On the
other hand cases of SFT were reported at other sites and
originating from other serous membranes such as the peri-
toneum [4], the pericardium [4–7] or from the mediastinum
[3]: this suggested that SFT could arise from submesothelial
mesenchymal elements, from a primitive mesenchymal cell
closely allied to the multipotential subserosal cell, as
demonstrated by the immunoreactivity for vimentin but
not keratin.

SFT can cause problems in clinical differential diagnosis
and treatment: its clinical presentation is similar to that of
other mediastinal tumours (thymoma, haemangiopericy-
toma, lymphoma). Three out of the four cases of SFT arising
from the pericardium, described in the literature, showed
location within the pericardial sac [4,5,7] and of the last
one we failed to find information about its pattern of growth
[6]. A case of SFT arising from the epicardium and growing
in the pericardial cavity, was recently reported [10]. The
patient herein presented is the first case of SFT of the peri-
cardium with extrapericardial growth.

Surgical resection of SFT can be hazardous, due to the
usually large dimensions of these tumours and their connec-
tions with the great vessels, but it is usually the treatment of
choice: because the clinical behaviour of the disease is
unpredictable, complete excision of the tumour is always
advisable, even if it implies sacrificing nerves or vessels.

Local recurrence may also occur many years after the resec-
tion [4]: a long follow-up is required to detect recurrent
tumours.

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