Case report

An unusual presentation of solitary fibrous tumour of the pleura: right atrium and inferior vena cava compression

Walid Shaker, Tchao Meatchi, Daniel Dusser, Marc Riquet*

Department of Thoracic Surgery, Georges Pompidou European Hospital, 20-40 rue Leblanc, 75908 Paris Cedex 15, France

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Abstract

Solitary fibrous tumours of the pleura are rare and usually present with unpredictable clinical signs. We report a 58-year-old patient presenting with signs of cardiac decompensation and external compression of the right atrium and inferior vena cava by a voluminous solitary fibrous tumour arising from the diaphragmatic pleura. Marked clinical improvement was obtained after excision of the mass with complete disappearance of the cardiac signs. © 2002 Elsevier Science B.V. All rights reserved.

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1. Introduction

Primary tumours of the pleura can be divided into two major groups: diffuse and solitary. Diffuse pleural tumours or mesotheliomas are the commonest. The solitary fibrous pleural tumours (SFPT) are rare and originate from the mesenchymal tissue underlying the mesothelial layer of the pleura. They are usually asymptomatic. Some cases may have uncommon presentations [1]. We observed a case of rare clinical presentation that, to our knowledge, has not yet been reported.

2. Case report

A 58-year-old female, smoker, was admitted to the hospital for a rapid onset effort dyspnoea grade III associated with bilateral oedema of the lower limbs. The patient had lost 6 kg of her body weight in 3 weeks. Her main past medical and surgical history consisted of: a depressive syndrome controlled medically, chronic bronchial asthma, allergy to iodine and a hysterectomy 11 years ago. On initial clinical examination, we noticed bilateral lower limbs oedema reaching the mid calves with peripheral cyanosis, hypoxemia (83% saturation), no palpable lymph nodes, neither hepatic nor splenic enlargement. Cardiac examination revealed sinus tachycardia 100 beats/min with no audible murmur. On chest examination no air entry was found on the right infrascapular area. The plain chest X-ray showed a right side basal opacity masked by moderate amount of pleural effusion. The cardiac echocardiography revealed (Fig. 1) compressed and displaced right atrium and inferior vena cava, by a huge intrathoracic mass explaining the cardiac signs, and dilation of the inferior vena cava with high filling pressure. The pulmonary artery pressure was estimated at 42/20 mmHg. Very mild tricuspid insufficiency grade 1/4 and mitral insufficiency grade 1/4 were observed. Further investigation by computed tomographic (CT) scanning (Fig. 2a) showed a huge heterogeneous opacity of 15 cm in length with multiple necrotic foci occupying the basal part of the right hemithorax; the mass extended superiorly to the right border of the heart, compressing and displacing the right atrium without intra-vascular invasion. The right lower pulmonary lobe was completely compressed and collapsed, the rest of pulmonary parenchyma was normal. The right diaphragmatic dome was everted and the liver pushed inferiorly (Fig. 2b). Bronchoscopy confirmed lower lung lobe external compression. Cytological examination and biopsies were non-significant with negative smears for mycobacteria. Extra-thoracic investigations included a normal bone and brain scan and abdominal echography. Pulmonary function and blood gases showed the following results: vital capacity, 1.8 l (61%); forced expiratory volume in 1 s, 0.61 (24%); arterial oxygen tension, 62 mmHg and; arterial carbon dioxide tension, 43 mmHg. Other laboratory results included normal blood sugar level, serum albumin and liver enzymes. Surgery was scheduled and performed after careful left lateral position-
ing for fear of cardiac decompensation. Following a large thoracotomy with sixth rib excision, a huge mass was found that was firmly adherent to the diaphragm. The mass was resected inferiorly together with 8 cm of the right dome reaching the inferior vena cava which was free. The peritoneum remained intact during the entire procedure. Ante-

iorly, the mass was strongly attached to a short segment of the phrenic nerve pedicle and the pericardium. Wedge resection of the lower lobe was performed with an almost complete preservation. The diaphragmatic defect was sutured by separate stitches. The excised tumour mass was 18 × 17 × 12 cm in diameter and 1.800 g in weight. Post operative histological examination revealed a tumour having the classic histological pattern of SFPT with a network of fibroblast-like cells. An immunohistochemical study of the tumour mass confirmed this diagnosis. Others characteristics were a very low mitotic rate (number of mitosis (4/10 High Power Field)) and presence of a few necrotic areas, a description of potentially malignant solitary fibrous tumour. However, pericardium and lung parenchyma resected together with the tumour were not infiltrated. The early post operative period showed marked clinical improvement, and disappearance of clinical signs of cardiac tamponade. The forced expiratory volume passed from 24 to 58%. Oxygen demand decreased to 2 l/min over a few days. The patient was faring well with no recurrence over 2 years follow-up.

3. Discussion

The solitary fibrous tumours of the pleura are relatively rare. They are usually asymptomatic and may present with unpredictable clinical behaviour. In most cases they affect males more than females. The most common presentation are small tumours less than 10 cm in diameter with incidental discovery by radiological studies, whether chest X-ray or CT scan, in asymptomatic patients [1–6]. Larger tumours, more than 10 cm in diameter occupying a larger space in the thoracic cavity, present more commonly with dyspnoea, chest pain, fatigue and dry cough. The extra-thoracic presentations include weight loss, night sweat, hypertrophic pulmonary osteoarthropathy and hypoglycemia. Hypertrophic osteoarthropathy (Pierre Marie–Bamberg syndrome) [1–6] is related to the abnormal production of hyaluronic acid by the tumour cells and affects up to 20% of the patients [6]. Hypoglycaemia is a rare presentation due to pathological secretion of insulin-like growth factor IGF2 by the tumour. Rena et al. [1] stated that IGF2 could suppress insulin and IGF1 secretion and that an abnormal IGF1:IGF2 ratio might indicate an IGF2 secreting tumour. They also found that both these hormones level returned to normal once the tumour was removed and that the recurrence of this paraneoplastic syndrome was a sign of recurrence of the solitary fibrous tumour in one patient. In our reported case, the giant solitary fibrous tumour of the pleura presented with bilateral lower limbs oedema due to cardiac chambers compression, mainly the right atrium, which was displaced anteriorly by the mass, and inferior vena cava compression. The lower limbs oedema reaching the mid calves was associated with dyspnöea on effort grade III and hypoxemia. The other causes of bilateral lower limbs oedema were excluded by clinical examination,

Fig. 1. Echocardiography showing compressed and displaced right atrium (RA).

Fig. 2. (a) TMD, huge heterogeneous opacity extending to the right border of the heart. (b) TMD lower view, the liver (*) is pushed inferiorly by the mass.
laboratory studies and radiography. The lower limbs oedema and the chest pain disappeared with marked improvement of dyspnoea in the early post operative period. The occurrence of cardiac chambers compression by solitary fibrous tumour of the pleura must be very rare even with large size tumours and to our knowledge has not yet been reported. In our case, this presentation confirmed by echocardiography must be attributed to the diaphragmatic pleural origin of the tumour which was in near relation to the cardiac chambers. The SFT originates usually from the visceral pleura. Cardillo et al. [6] found that 87.28% of their case group were based on visceral pleura and only 12.72% from the parietal. The origin of the SFPT in our case from the parietal diaphragmatic pleura may be responsible for the occurrence of cardiac and inferior vena cava compression.

4. Conclusion

Solitary fibrous tumours are localized tumours of the pleura which may have unpredictable clinical presentations. In small sized tumours, they are usually discovered incidentally by chest X-ray in asymptomatic patients. In tumours larger then 10 cm, they may be presented by dyspnoea, chest pain, cough, weight loss and fatigue. Rare presentations include hypoglycaemia, hypertrophic pulmonary osteoarthropathy or signs of cardiac chambers compression as seen in our reported case.

References