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CASE BASED REVIEWS

Largest known malignant solitary fibrous tumour of the pleura-extended resection warranting cardiopulmonary bypass support

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Abstract
This case reports the largest known malignant solitary fibrous tumour of the pleura treated with en bloc surgical resection warranting the use of cardiopulmonary bypass support. A 60-year-old male presented with dyspnoea and a dry cough. Following extensive investigations, a radiological and histologic diagnosis of malignant solitary fibrous tumour of the pleura was made. This 4.3 kg tumour occupied the entire left hemithorax, involved the left lung and infiltrated into the pericardial cavity. Although the postoperative course was uneventful with a 12-day length of stay, the patient opted not to undergo adjuvant radiotherapy to a single positive margin site and died 6 months later due to local recurrence.

Keywords Malignant · Pleura · Tumour

Introduction

Solitary fibrous tumours of the pleura (SFTP) are rare mesenchymal tumours, with a reported incidence of 2.8/100,000. They represent less than 5% of all tumours of the pleura. They occur in all age groups, with peaks in the fourth and sixth decades of life. Malignant SFTPs represent 10–20% of all SFTPs, with a reported 5-year survival of 81% [1].

Following a review of the literature, less than 1000 cases of mSFTP have been reported, mostly as single case studies, as well as a few case series. As a result, there is no established consensus on treatment modality or adjuvant treatment protocol. The average tumour size in one case series was 2.13 kg [1]. This reports the largest mSFTP ever resected at 4.3 kg.

This case has previously been presented as a poster at the Society for Cardiothoracic Surgery for Great Britain and Ireland annual meeting, March 2017, Belfast, Northern Ireland, and also as a poster at the Irish Thoracic Society annual scientific meeting, November 2017, Limerick, Ireland

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Case report/description

A 60-year-old male farmer presented to his local hospital in June 2016 with a 3-month history of dyspnoea, orthopnoea and paroxysmal nocturnal dyspnoea, as well as a dry cough. He denied any chest infections, constitutional symptoms such as weight loss or night sweats, exposure to asbestos or radon gas or other occupational exposures. He was a nonsmoker and nondrinker.

His only past medical history was that he had presented to hospital in 1989 with headaches. An incidental finding of a pulmonary nodule was made on chest x-ray at that time, however the patient was lost to follow-up. He had no past surgical history. His family history was significant for colorectal cancer. His regular medications included a combined albuterol/ipratropium inhaler, esomeprazole, paracetamol and a macrogol laxative.

Positive findings on initial physical examination included clubbing, tracheal deviation, a raised jugular venous pulse, a murmur consistent with aortic stenosis and peripheral oedema.

Several investigations were carried out in the local hospital. Chest x-ray revealed a left-sided pleural effusion. CT thorax confirmed the effusion and diagnosed a large mass occupying the left hemithorax measuring 21 × 21 × 17 cm, with almost total atelectasis of the left lung. It was intimately related to the

descending aorta, left ventricle and left side of the mediastinum at multiple levels. There was pronounced mediastinal shift and compression of the mediastinal vascular structures.

A chest drain was inserted and 4 l of haemorrhagic fluid was drained; subsequent cytology analysis was negative for malignancy. CT-guided biopsy was performed and returned a histological diagnosis of a benign spindle cell neoplasm with features of a solitary fibrous tumour. Repeat biopsy yielded the same result.

The patient was then transferred to the care of the thoracic surgery service at a major tertiary hospital, where further investigations were carried out. MRI confirmed the earlier CT findings including the significant mass effect. PET CT was performed and revealed a multilobular mass which was extensively necrotic and showed small areas of dystrophic calcification. It avidly accumulated fluorodeoxyglucose.

On echocardiography, only the right parasternal and right apical views were available, consistent with the heart being pushed toward the right hemithorax. The chambers and valves were all structurally normal. The large pleural effusion was noted but there was no pericardial effusion.

Spirometry showed a restrictive picture, with marked reduction in the FEV1 and FVC, but maintenance of the FEVI/FVC ratio. Of note, the patient still had a reasonably good exercise tolerance given both the radiological picture and the spirometry: he was able to walk 200 m on the flat, and seemed to be quite physiologically well, despite almost total left-sided autopneumonectomy.

In consultation with the respiratory multidisciplinary team, a plan for surgical resection was made, and the patient was brought to theatre in July 2016. Approach was initially via a left anterior thoracotomy with transection of the sternum (hemiclamshell) later extended to a full bilateral clamshell incision to facilitate better access (Fig. 1). Decortication was performed and intrapericardial pneumonectomy was necessary due to the extent of the disease. As access to the left pulmonary artery and pulmonary veins was particularly difficult due to mediastinal shift, the patient was put on cardiopulmonary bypass (CPB) to empty the heart and allow for stapling of the vessels and en bloc explantation.

Pericardiectomy was performed with vicryl mesh reconstruction. At the time of closure, there was no macroscopic evidence of any remaining disease (Fig. 2).

There was significant blood loss, and haemostasis was achieved with considerable difficulty using clips, diathermy, argon, floseal and surgicel. As a result, the left hemithorax was electively packed and the patient was returned to theatre 24 hours later for delayed closure. The patient went to the intensive care unit intubated and ventilated with three chest drains in situ.

He was successfully extubated at day 2 postoperatively, made an uneventful recovery, and was transferred back to his local hospital on day 13.

The histopathology was reported as a 30 × 25 × 12 cm malignant solitary fibrous tumour of the pleura, weighing 4.3 kg, inseparable from the left lung and pericardium. There was one positive margin at the anterior chest wall. There was no nodal involvement. Cytology of pleural and pericardial fluid was negative for malignancy. Immunohistochemistry stained positive for CD34 and vimentin, and this excluded the major differential of malignant mesothelioma.

In consultation with the sarcoma multidisciplinary team, a repeat CT was performed, as adjuvant treatment was being considered. Disease was noted on the anterior chest wall at the previous positive margin site, and this was queried as recurrence/progression. A repeat CT-guided biopsy was recommended but the patient declined.

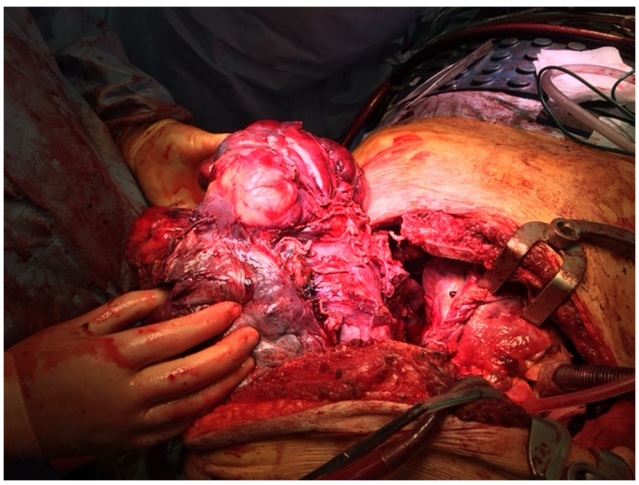

Fig. 1 Superior view of clamshell thoracotomy with CPB support

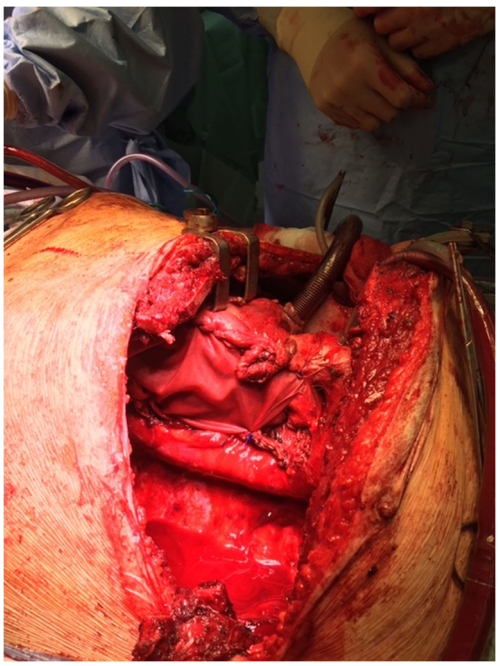

Fig. 2 Vicryl mesh reconstruction following pericardiectomy

The patient was offered but refused adjuvant radiotherapy to the single involved chest wall site. At 4 months postoperatively, he began to deteriorate clinically, due to recurrence of his tumour. A decision was made not to treat with palliative chemotherapy, especially because the tumour was not particularly chemosensitive. The disease progressed locally and unfortunately the patient died at 6 months postoperatively.

Comment

The paucity of reported cases of mSFTP in the literature makes any consensus on primary and adjuvant treatment approach difficult to reach. As such, a highly individualised treatment plan is required. Extended en bloc resection, if technically feasible, would appear to offer increased 5-year survival when compared to isolated mass excision (87.5 vs 0% in a review by Lococo and colleagues) [1]. There is no evidence to support a role for chemotherapy or radiotherapy in the management of these tumours, and as such surgery is the primary, and only feasible treatment modality [2].

Interestingly, the preoperative histology in this case was reported as benign on two separate biopsies, while the final operative specimen was reported as malignant. Image-guided biopsy for intrathoracic tumours has a well-established evidence base in terms of its diagnostic accuracy [3, 4]. However, when dealing with such large and heterogeneous tumours, one must maintain a healthy degree of scepticism with regard to preoperative histological diagnoses. Tumour biology and clinical behaviour should be given as much, if not more weight than histology in planning the multimodality treatment of these malignancies.

The use of CPB support in this case facilitated the en bloc resection of a tumour which would otherwise have only been a candidate for isolated mass excision. The use of CPB to facilitate a more ambitious surgical approach to thoracic tumours was described as early as 1971 [5]. CPB is used both emergently in the management of massive haemorrhage and electively in carefully selected patients to facilitate the excision of tumours infiltrating or abutting the heart or great vessels [6]. Following its successful use in this case, we suggest that CPB support should continue to be used to facilitate the resection of large and invasive thoracic tumours.

We suggest that early adjuvant radiotherapy may have been of benefit to this patient, given that his local recurrence occurred at a single chest wall site. However, we recognise that there is a lack of strong evidence in the literature around adjuvant treatment and as such it is difficult to make any substantial predictions as to the potential benefit of early adjuvant radiotherapy. This patient's refusal of early adjuvant radiotherapy reinforces the need for frank discussion of multimodality treatment and the potential for adjuvant therapy up front with patients, before surgery is performed.

In conclusion, we report a case of the largest known mSFTP treated with en bloc surgical resection. Cardiopulmonary bypass facilitated a more ambitious surgical approach, and while literature would suggest that this should confer an improved prognosis it was unfortunately not the case with our patient.

Compliance with ethical standards

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was taken for images used in this article.

Conflict of interest The authors declare that they have no conflict of interest.

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