CASE REPORT

Solitary fibrous tumour: a rare tumour of the pleural cavity

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SUMMARY

Solitary fibrous tumours are a rare group of tumours that originate from connective tissues such as the pleura and mediastinum. When growing within the thoracic cavity these tumours exert pressure on vital organs and large vessels. We have successfully operated on two patients with large solitary fibrous tumours and managed the pathophysiological changes manifested as superior vena cava syndrome resulting from these tumours.

BACKGROUND

Massive solitary fibrous tumours may create respiratory and circulatory disorders through pressure on vital organs within the thoracic cavity, causing lung collapse and deviation of the mediastinum, affecting the heart and the large vessels, which can lead to syndromes such as superior vena cava syndrome (SVCS). These tumours are rare and may reach up to 30 cm in diameter, and are often deemed unresectable.

We describe our experience treating two patients with large solitary fibrous tumours within the thoracic cavity, one originating from the pleura and the other from the pericardium. After 2 years of follow-up, the patients were still well with no recurrence.

CASE PRESENTATION

The first case is of a patient with dyspnoea and palpitations with limited physical activity during the 3 months before surgery, during which the patient could not lie down due to worsening dyspnoea and cyanosis of the face. The patient underwent chest computed tomography (CT) scan which showed a mass measuring 7×8×19 cm in the anterior mediastinum (figure 1). The tumour exerted pressure on the superior vena cava (SVC), aortic arch, the right side of the heart and the anterior aspect of the right lung, with resulting deviation of the mediastinum with symptoms corresponding to SVCS. On needle biopsy the mass was identified as a solitary fibrous tumour. An operation was performed under general anaesthesia in the anti-Trendelenburg position to avoid the central cyanosis and oedema that result from SVCS while lying supine. Surgery was performed using an anterolateral approach; an additional incision was made parallel to the lateral border of the sternum (figure 2). A solitary fibrous tumour was excised intact, measuring 9×13×15 cm and weighing 1040 g. During excision haemodynamic instability was observed, necessitating further

treatment in the intensive care unit (ICU) for haemodynamic stabilisation. The patient was extubated successfully and was transferred to the thoracic surgical ward. After an uneventful recovery, the patient was discharged a week after surgery. Post-operative follow-up included echocardiography and a CT scan of the chest that demonstrated a return to the normal anatomical position of the viscera and normal function of the heart. Pathology results showed the tumour was benign, with positive immunohistochemistry tests for CD34, bcl-2 and CD79, and negative for keratin.

A second patient, with hypertension and Parkinson's disease, presented with a lobulated intrathoracic mass that he had had for 3 years, and which was initially seen on a chest X-ray for pneumonia. On a previous CT scan (3 years previously) it was found to be a well-defined mass with a regular border, hypodense and heterogeneous in character. The mass was in close proximity to the heart and hilum of the left lung, and measured 6×6.5×8 cm. An MRI suggested a thymoma in the left mediastinum. The patient refused a needle biopsy. Three months before hospitalisation his health deteriorated and he was admitted due to dyspnoea and chest pain during minimal effort and turning in bed. A CT scan and needle biopsy identified a huge solitary fibrous tumour originating from the left pleura, with deviation of the mediastinum to the right and pressure on the left side of the heart (figure 3). Surgery was performed via an anterolateral thoracotomy approach; the solitary fibrous tumour was located on a pedicle growing from the left pleura, measuring 12×15.8×22 cm and weighing 2400 g. Excision entailed meticulous haemostasis and closure of the lung with staples and biological glue. The size of the tumour required a second incision below the original thoracotomy incision to allow access to push the tumour up through the original incision. In this case the tumour was classified pathologically as of uncertain malignant potential, and immunohistochemistry tests were positive for CD34, CD-99 and bcl-2. Immunohistochemistry for Ki-67 was positive in 5%–8% of cells in low cellular areas, and in 8%–10% of cells in hypercellular areas.

INVESTIGATIONS

The diagnostic investigation of a solitary fibrous tumour of the pleura is the same as for any other mass in the chest cavity, starting with imaging by X-ray and CT scan, which visualise the characteristic well-circumscribed mass.

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Figure 1 A CT scan of a solitary fibrous tumour exerting pressure on the heart and large vessels.

Most authors recommend avoidance of percutaneous transthoracic needle biopsy since it does not influence the need for surgical treatment.¹ However, Sung *et al* report a moderate success rate of 43% with fine-needle aspiration in obtaining a definite preoperative diagnosis of solitary fibrous tumours of the pleura, although it did not appear to affect their choice of surgery as treatment for all of their patients.²

OUTCOME AND FOLLOW-UP

Both patients were followed up in the outpatient clinic for 2 years. They both made full recoveries, returning to their previous physical status.

DISCUSSION

Solitary fibrous tumours are rare, usually arising in the elderly, and between 10% and 30% may be malignant.³ These tumours have similar histological characteristics, being composed of spindle cells. Solitary fibrous tumours were first described in 1931 by Klempeper and Rabin.⁴ With the development of

Figure 2 The anterior part of the tumour.

Figure 3 A scan of pleural solitary fibrous tumour from mediastinum.

immunohistochemical tests, fibroblasts have been found to be the source of solitary fibrous tumours. This may explain their presence outside the thoracic cavity, originating from tissue without mesothelial cells,⁵ such as the pleura, peritoneum and pericardium, and organs without serosal cells, such as the lung, mediastinum, nose, paranasal sinuses and liver.⁶

Most thoracic solitary fibrous tumours originate from the pleura (in 80% of cases), with an incidence of 2.8 per 100,000,³ with only a few tumours reported as originating from the pericardium. Most solitary fibrous tumours are found incidentally on chest X-ray, with the main symptoms including cough, chest pain, dyspnoea and fever, which worsen as the tumour grows. These tumours are characteristically septate masses with a well-defined envelope, usually growing on a pedicle.⁷

The only successful treatment is surgery, with a risk of recurrence of about 50% in cases with malignant pathology.¹

Large intrathoracic solitary fibrous tumours may cause a shift of the mediastinum, thus exerting pressure on the heart and great vessels. Due to raised intrathoracic pressure on the thin wall of the SVC, the low pressure inside the vein and the close proximity to firm organs, the pressure that is exerted on the SVC leads to reduced venous return from the head, neck and upper extremities; this is termed 'superior vena cava syndrome'. This syndrome is indicated by cyanosis and oedema of the upper body (face, neck, chest and upper extremities) and oedema of the larynx leading to stridor and dyspnoea. Low blood pressure and dilatation of upper body veins are also possible. Increased pressure on the SVC will cause symptoms such as headache, nausea, vomiting and change in consciousness. SVCS is a challenge for anaesthesia (intubation) and surgery because these patients cannot tolerate being supine for a prolonged period of time, due to insufficient venous return. Our two patients were anaesthetised and intubated in a semi-recumbent position that prevented central cyanosis and hypoxaemia. A double-lumen tube and single-lung ventilation assisted in maintaining ventilation to the contralateral side from the tumour.

It seems optimal to avoid a thoracotomy in the lateral position for large tumours because the tumour may exert additional pressure on the heart. This may reduce venous return and lead

to further haemodynamic deterioration. The anterior approach was chosen with a modified anterolateral thoracotomy incision and parasternotomy incision. Our modification included incising the ribs just lateral to the sternum instead of the traditional midline sternotomy, a manoeuvre that allowed access to the tumour and intrathoracic organs in order to separate them while maintaining haemostasis.⁸ With this approach the incision may be extended as required, as in the first case. In the second case, the original incision was sufficient for dissection of the tumour, but the tumour was too large to deliver through the incision, so an additional incision was performed two intercostal spaces below, in order to push out the tumour. Another advantage of the anterolateral approach is the maximal exposure of the hilum of the lung and the vessels of the mediastinum and, in case of injury to them, this incision affords a better approach for repair of the injury.

A drop in blood pressure and tachycardia were noted after tumour removal, indicating haemodynamic instability, requiring inotropic support in the ICU. The haemodynamic instability was caused by the acute decrease in cardiac pressure while excising the tumour from the thoracic cavity. Prior to tumour extraction the heart had adapted to work against the progressive rise in pressure which led to chronic pathophysiological changes to maintain circulation. The sudden decrease in pressure from the heart caused an acute increase in venous return, without the

heart being able to dilate. The haemodynamic instability was not due to bleeding, as that was minimal (mostly from small vessels to the tumour envelope and while performing adhesiolysis). Both patients improved haemodynamically in the ICU, and inotropic support was discontinued on the second postoperative day.

During the recovery period the patients had pain at the incision and atelectasis; these are the most common complications after thoracotomy, and require effective pain management and physiotherapy.

Contributors RB is the main author. EA was responsible for planning. AG and AS were responsible for conception and design.

Competing interests None declared.

Patient consent Detail has been removed from this case description/these case descriptions to ensure anonymity. The editors and reviewers have seen the detailed information available and are satisfied that the information backs up the case the authors are making.

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Learning points

- ▶ A solitary fibrous tumour is a rare condition.
- ▶ Superior vena cava syndrome significantly challenges the anaesthesiology.
- ▶ A large intrathoracic tumour can be operated on with good long-term outcome.

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