Recurrent malignant solitary fibrous tumour of the pleura – a case report of a delayed diagnosis

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Abstract

Malignant solitary tumours of the pleura are extremely rare neoplasms usually presenting with dramatic symptoms. A 54-year-old female presented with symptoms of confusion. Chest X-ray and CT showed a huge mass in the right pleural cavity. Diagnostic thoracoscopy yielded a false result of small cell cancer. The final pathology was positive for a malignant SFT. The operative procedure required removal of the mass. The follow-up exam showed a recurrent tumour and a pneumonectomy was required for a radical resection. The postoperative course was uneventful and follow-up exam showed no recurrence of the disease.

Key words: pleura, lung tumours, pneumonectomy, recurrence.

Streszczenie

Złośliwe guzy włókniste opłucnej (ang. *Solitary Fibrous Tumor*) są bardzo rzadkimi nowotworami zazwyczaj przebiegającymi bezobjawowo. 54-letnia chora zgłosiła się do lekarza z objawami okresowych zaburzeń świadomości. Zdjęcie przeglądowe klatki piersiowej i tomografia komputerowa uwidoczniły duży guz opłucnej prawej. Wideotorakoskopia diagnostyczna dała śródoperacyjny wynik raka drobnokomórkowego i w związku z tym odstąpiono od radykalnego zabiegu. Ostateczny wynik histologiczny uległ zmianie na złośliwy guz włóknisty opłucnej. Zabieg operacyjny polegał na radykalnym usunięciu guza. Badanie kontrolne pół roku później sugerowało nawrót guza i konieczna była pneumonektomia uzupełniająca. Przebieg pooperacyjny był bez powikłań i dalsze badania kontrolne wykazały radykalność operacji.

Słowa kluczowe: opłucna, rak płuc, resekcja płuca, nawrót.

Introduction

Malignant solitary fibrous tumours (SFTs) of the pleura are extremely rare. They account for approximately 12 to 40% of the entire SFT entity, which is itself an uncommon thoracic neoplasm [1, 2]. Previously classified as a benign form of mesothelioma, they have a variable potential for malignancy. Based on the mitotic activity, nuclear pleomorphism and the degree of cellularity, this potential can be estimated, thus requiring a close clinical follow-up.

We report a case of a 54-year-old previously healthy female presenting with short episodes of confusion. She had no other symptoms. During a work-up a chest X-ray was done, revealing a huge mass in the right lung (Fig. 1a). A CT scan was performed, confirming a giant 17 cm mass, shifting the mediastinal structures to the left side (Fig. 1b). The mass was partially infiltrating the descending aorta. The mediastinal lymph nodes were not enlarged. Fibreoptic bronchoscopy revealed a rotated, narrow bronchial lumen in the left bronchial tree. The episodes of confusion were linked to hypoglycaemia. Since the biopsies taken during bronchoscopy and transthoracic biopsies were negative, a decision to perform diagnostic VATS was made. The frozen section of the biopsy was positive for small cell cancer and the procedure was terminated. A week later however, the final pathology came back as a solitary fibrous tumour. The cells were found to be immunoreactive for CD34, vimentin and Ki67, supporting a mesenchymal origin of the mass. Subsequently, a thoracotomy was performed and a giant encapsulated tumour weighing 2450 g was encountered. The tumour was infiltrating the lung parenchyma (Fig. 2). The radical removal required a middle lobectomy. The postoperative period was uneventful and there were no more episodes of hypoglycaemia. During a routine 6-month follow-up CT of the chest was done, revealing a recurrent mass in the right hilum. The patient, however, still being asymptomatic, declined further intervention and disappeared from the clinic. She returned a year later with symptoms of persistent cough and a repeat CT scan showed progression of the tumour. Rethoracotomy was performed and a recurrent 10 × 8 cm mass was found. A radical resection required a right pneumonectomy. The postoperative period was uneventful and the patient was discharged on POD #8.

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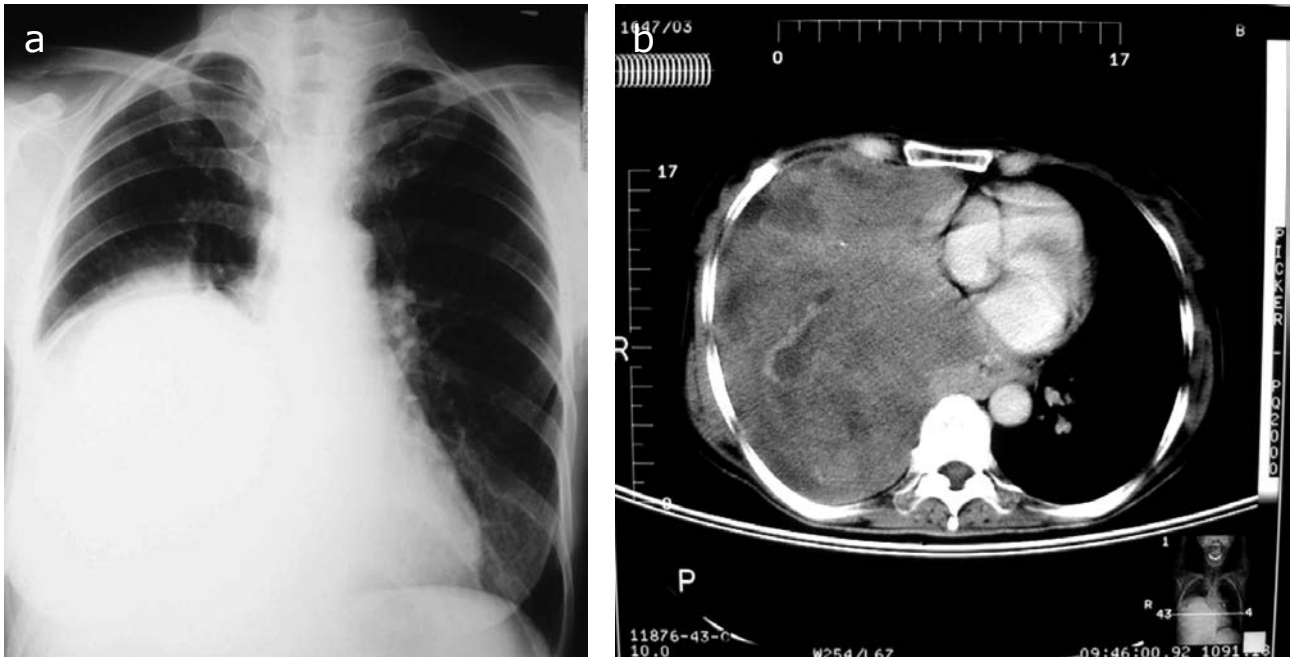

Fig. 1a-b. Chest X-ray and CT scan of the chest reveals a giant mass, shifting the mediastinal structures to the left

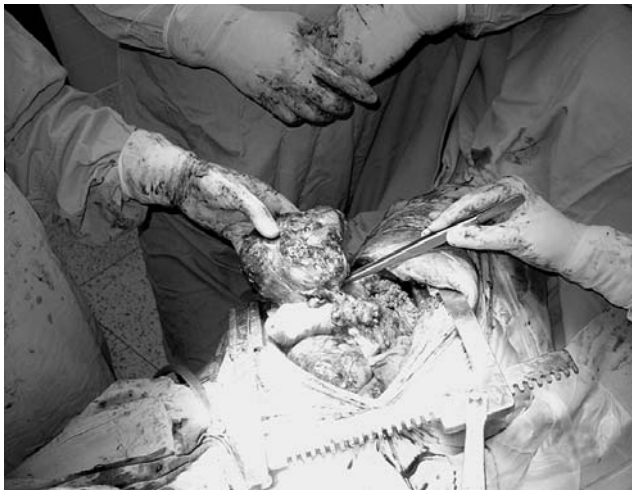

Fig. 2. Intraoperative view of the giant fibrous tumour of the pleura

A follow-up CT scan 12 months later showed no signs of recurrence and the patient remains asymptomatic.

Comment

Our case is unique because of the mass size, symptomatology and the pattern of recurrence. Most patients present with dramatic symptoms of dyspnoea, chest pain and cough. The presented patient in spite of the giant size of the tumour had only symptoms consistent with hypoglycaemia. The incidence of hypoglycaemia is estimated as 14% [1]. In spite of grossly radical resection a relatively fast recurrence was found. Unfortunately, the follow-up was interrupted by the patient, who did not want any intervention at that time. Although the repeat resection required a pneumonectomy, a good outcome was achieved. The tumour arose from the visceral pleura, requiring a resection of the involved lung parenchyma. This is consistent with the majority of reported cases [2].

SFT of the pleura can show a wide spectrum of histological features. The majority of previously reported tumours are benign in origin [1–3]. Local recurrence is typical only in the malignant variant [4]. Although a radical resection is still the mainstay of therapy, it is still unclear whether there is any role for neoadjuvant or adjuvant therapy. Patient follow-up is mandatory as the tumour's behaviour is unpredictable [5].

References

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