An unusual presentation of malignant mesothelioma as constrictive pericarditis

PUNEET KAKAR, VINAY S SREEGURU, SOMIT SARKAR, KATE A WILLMER, WILLIAM T BERRILL

Introduction

This case report discusses a 54-year-old woman who presented to hospital with recurrent bilateral pleural effusions. She was eventually found to have constrictive pericarditis secondary to malignant mesothelioma. This disease presents a challenge to the physician with considerable difficulties in diagnosis, classification and treatment. This particular presentation of malignant mesothelioma is highlighted in the article.

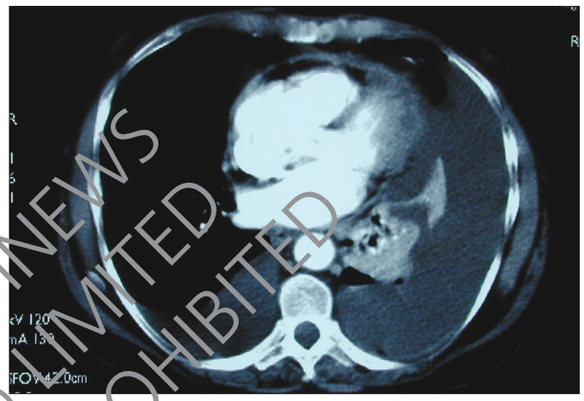
Br J Cardiol 2006;**13**:432–3

Case report

A 54-year-old woman, known to be a smoker, presented to the rapid access chest pain clinic as she had suffered with progressive shortness of breath on exertion for three weeks. She had been apparently healthy until three weeks previously and there was no preceding pre-morbid history or previous major illnesses. Preliminary investigations demonstrated bilateral pleural effusions, which were tapped, and the fluid was sent for laboratory analysis. She was then referred to the respiratory physician for further investigation.

On examination she appeared pale and lethargic. There was no clubbing, cyanosis, lymphadenopathy or pedal oedema. Her jugular venous pressure was not elevated and her heart sounds were normal. Respiratory system examination showed reduced air entry and dullness to percussion on both bases. Abdomen examination revealed a palpable, non-tender liver. Her haemoglobin was 14.2 g/dL and her white blood cell count (WCC) was 5.6 10^9/L. Urea, creatinine and serum electrolytes were all shown to be in normal ranges. Liver function tests, however, were deranged and showed a serum bilirubin of 49 nmol/L, a gamma glutamyl transferase of 233 IU/L and an alkaline phosphatase (ALP) of 206/L. Chest X-ray demonstrated bilateral pleural effusions (L > R). Abdominal ultrasound was normal.

Figure 1. CT scan of the thorax showing pericardial thickening



Pleural fluid analysis demonstrated a transudate with a fluid protein of 33 g/L and a fluid albumin of 21 g/L. Samples sent for malignant cytology, gram staining, culture and sensitivity and mycobacterium analysis, were all negative. Other investigations, including those for rheumatoid factor, autoantibody screen and thyroid function, were unremarkable. Plasma viscosity was marginally elevated at 1.84 mPa.

Meanwhile the patient's condition was rapidly deteriorating and she developed bilateral pedal oedema and raised jugular venous pressure. Echocardiography demonstrated under-filling of all four cardiac chambers with hyperdynamic overall wall contractility and a bright echo near the apex/posterior wall which appeared to restrict diastolic filling. The echocardiogram also showed a dilated, non-collapsible, inferior vena cava on sniffing, that implied raised right heart pressure.

A CT scan (figure 1) of her thorax demonstrated irregular pericardial thickening and effusion, bilateral pleural effusions and no demonstrable pulmonary mass or pleural plaques. Cardiac catheterisation was not possible as the patient was unable to lie flat and she was referred to the regional centre for thoracoscopy. Myocardial thickening and patchy pericardial infiltration was demonstrated. A pericardial biopsy was taken and histopathological analysis showed a sarcomatoid malignant mesothelioma.

In view of the biopsy findings and the patient's condition,

Department of Medicine, West Cumberland Hospital, North Cumbria Acute Hospitals NHS Trust, Hensingham, Whitehaven, Cumbria, CA28 8JG.
Puneet Kakar, Senior House Officer, Medicine
Vinay S Sreeguru, Senior House Officer, Medicine
Somit Sarkar, Senior House Officer, Medicine
Kate A Willmer, Consultant Cardiologist
William T Berrill, Consultant Respiratory Physician
Correspondence to: Dr P Kakar, Flat 1, 50 Mariner Avenue, Edgbaston, Birmingham, B16 9DT.
email: puneetkakar@hotmail.com

she was transferred back to the palliative unit of the West Cumberland Hospital. She died a few days later. The post-mortem report confirmed malignant mesothelioma involving the pleura, pericardium and myocardium. Tissue samples sent for asbestos analysis showed no asbestos fibres.

Discussion

Malignant mesothelioma can result from a comparatively short exposure to asbestos. It has been estimated that as a result of previous exposure to asbestos, annual mesothelioma deaths in men, in age-specific cohorts, up to 2020, may be up to 1% of all deaths, since the latent period in developing a mesothelioma may be as long as 40 years after exposure.[1]

Cases not due to mineral fibre exposure usually have no other cause and the prognosis is worse. These occur in a frequency of one to two per million per year.[2] Smoking does not increase the risk of mesothelioma with or without asbestos exposure, whereas smoking increases the risk 40-fold in bronchogenic carcinoma due to asbestos exposure.

There are still difficulties in establishing the criteria for diagnosis of mesothelioma as a primary tumour. The tumour arises from the mesothelium of the body's serous cavities. Cases not due to fibre exposure have a wider age range and a more varied cell pattern. Common clinical features are constrictive pericarditis, cardiac tamponade and cardiac failure. Further symptoms may arise due to compression of coronary arteries and local spread into the surrounding great vessels. Mesothelioma can resemble pericarditis or myxomas. Conduction block due to myocardial infiltration,[3] and tumour embolism causing neurological deficits[4] have also been reported. Breathlessness is the most common presenting symptom. It may be sudden or gradual in onset and usually heralds a pleural effusion.

The usual site of affliction is the pleura. When the peritoneum or pericardium are involved, much higher levels of asbestos exposure are required.[5] Aspiration may sometimes be difficult due to the toughness of the parietal tumour: repeated attempts at aspiration, needle biopsy and surgery, may encourage the tumour to track through the chest wall. CT scans are usually helpful in diagnosis and if a history of asbestos exposure is present, this can provide a relatively accurate diagnosis.

The outlook for the patient is relatively poor since death due to pulmonary embolism, myocardial infarction or mediastinal constriction is common. All mesothelioma cases should be referred to the coroner regardless of whether there has been any significant exposure to asbestos. Proof is provided by the lung fibre burden of the amphibole. Significant exposure is deemed to have occurred when the electron microscopy count is over a million amphibole fibres/dry gram using the digistran technique.[1] Half of those with pleural disease die within one year of diagnosis and few live for up to two years.[5] Aggressive therapy such as pneumonectomy, chemotherapy and radiation, sometimes appears to improve the prognosis.

Key messages

- Malignant mesothelioma is a difficult condition to diagnose, classify and treat
- It can result from exposure to asbestos; the prognosis is worse in cases with no known cause
- Common clinical features of the condition are constrictive pericarditis, cardiac tamponade and cardiac failure
- The CT scan can highlight that malignancy is a likely cause

Conclusion

This patient presented to cardiologists because of her recent onset of breathlessness combined with signs that were suggestive of cardiac failure. The key features suggestive of constrictive pericarditis were the raised right heart pressures with bilateral pleural effusions without evidence of pulmonary congestion. Malignancy as a likely cause was highlighted by the CT scan result. This is the key investigation in such cases.

Conflict of interest

None declared.

References

1. Baxter PJ, Adams PH, Aw TC, Cockroft A, Harrington JM (eds). *Hunter's diseases of occupations,* 9th edition. London: Arnold, 2000;692-4.
2. Warrell DA, Cox TM, Firth JD, Benx Jr EJ (eds). *Oxford textbook of medicine, Volume 2,* 3rd edition. Oxford: Oxford University Press, 2003;1544-50.
3. Fazekas T, Tiszlavicz L, Ungi I. Primary malignant pericardial mesothelioma. *Orv Hetil* 1991;**132**:2677-80.
4. Szczechowski L, Janiec K. Pericardial mesothelioma as a very rare cause of recurrent cerebral emboli. *Wiad Lek* 1992;**45**:857-61.
5. Kumar V. Abbas AK, Fausto N (eds). *Pathological basis of disease (Robbins and Cotran),* 7th edition. Philadelphia: Elsevier Saunders, 2005; 768-70.