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A rare case of Erdheim–Chester disease

Authors: R Chhabra and C Wall

Aims

Discussion of an extremely rare case of Erdheim–Chester disease (ECD), which is a rare, non-Langerhans cell histiocytosis with characteristic radiological and histological features. Only several hundred cases have been described in the medical literature to date.

Methods

We report a case of a 74-year-old gentleman who presented with a 2-week history of exertional chest pain and shortness of breath. His background history was significant for renal artery stenosis, hypertension and renovascular disease. There were no new changes on electrocardiograph or significant troponin rise.

Relevant initial investigations included a chest X-ray, which showed a moderate loculated right-basal pleural effusion. A subsequent CT thorax confirmed bilateral chest loculated effusions, and presence of soft tissue thickening around the thoracic aorta and subcapsular low attenuation soft tissue material around the left kidney.

In view of these findings, a right pleural drain was inserted and he proceeded to have a left perinephric soft tissue biopsy. The pleural fluid was exudative as per Light's criteria and no malignant cells were identified. Interpretation of radiology findings as well as perinephric biopsy results were consistent with a diagnosis of ECD.

Results

Haematology input was sought. He was treated with interferon α, to which he has responded well.

Conclusions

This is a rare multi-system disease, which can be challenging to diagnose and treat. The overall prognosis of the disease is poor. The 5-year survival of ECD is 68%. ■

Authors: AMNCH (The Adelaide and Meath Hospital), Dublin, Ireland