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ENDGAMES

SPOT DIAGNOSIS

Recurrent haemoptysis in a child with advanced cystic fibrosis lung disease

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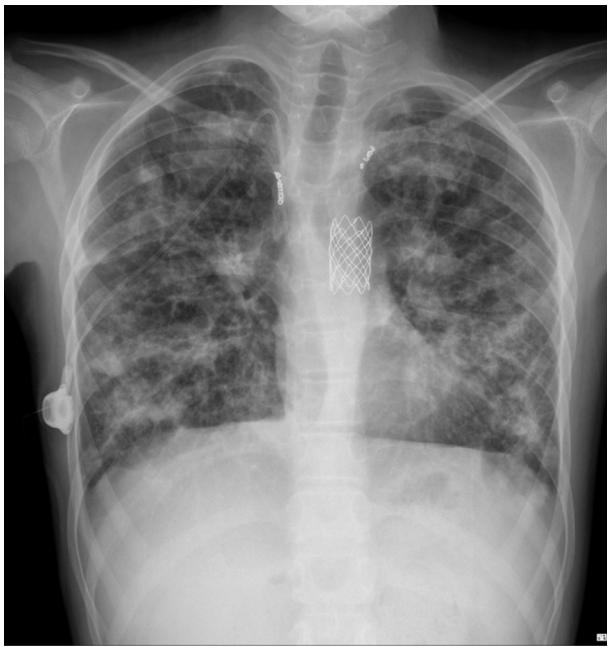
A 12 year old boy with cystic fibrosis has advanced lung disease (FEV_1 30%-40% predicted) and needs monthly intravenous antibiotics given through an implanted venous access device (visible in the radiograph in fig 1). He has experienced recurrent life threatening haemoptysis, for which he was treated with tranexamic acid and propranolol. His condition has not improved. What further intervention, seen on the radiograph (arrows), has he undergone?



Chest radiography of a child with advanced cystic fibrosis lung disease and an implanted venous access device in situ on the right side

Answer

Radio-opaque coils have been inserted to embolise the internal mammary arteries and a covered stent to occlude the origins of the bronchial arteries in the thoracic aorta.

Discussion

Major haemoptysis is a recognised complication of cystic fibrosis. It can be challenging to treat and is potentially life threatening.[1] The cause is typically aberrant bronchial arteries, which normally originate from the aorta to supply the bronchi and lung parenchyma.[2] Bronchial artery embolisation is the intervention of choice for life threatening bleeds, but can be technically challenging, has a high failure rate, and has substantial potential complications, including postembolisation syndrome, contrast media hypersensitivity, groin puncture haematomas, femoral artery pseudoaneurysms, vasospasm, dissection, perforation with wire or catheter, and some neurological complications.[2 3] Embolisation of non-bronchial systemic arteries, such as the internal mammary arteries, is an option if haemoptysis continues.[3] When embolisation fails, surgical resection of a lobe or lung can be curative; however, in patients with cystic fibrosis, a disease characterised by non-homogeneous bilateral lung pathology, this is rarely an option.[1] The placing of a covered stent within the thoracic aorta to occlude the origins of the bronchial arteries is a novel approach, which can be considered when other options have failed.[4]

Patient outcome

The bronchial stent and bronchial artery embolisation failed to control the haemoptysis. The patient had bilateral cystic fibrosis bronchiectasis, bilateral aspergillomas, and an FEV_1 persistently less than 30% predicted, which meant the only treatment option

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remaining was bilateral lung transplant, which was performed successfully.

Learning points

Major haemoptysis is a recognised complication of cystic fibrosis, and can be life threatening.

Bronchial and non-bronchial systemic artery embolisation is the intervention of choice for recurrent or acute massive haemoptysis.

Bronchial artery embolisation is technically challenging and has a high failure rate.

We have read and understood BMJ policy on declaration of interests and declare no competing interests.

Patient consent: Patient consent obtained

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