Near-fatal haemoptysis as presentation of a giant intralobar pulmonary sequestration

A 50-year-old female with no prior respiratory disease or symptoms presented with massive haemoptysis and respiratory failure. Multidetector computed tomographic angiography demonstrated an aberrant artery supplying a lobulated mass occupying two-thirds of the right chest (fig. 1a and b). Aortography confirmed a large aberrant systemic artery originating from the supra-diaphragmatic aorta (fig. 1c), with drainage into the pulmonary veins (fig. 1d). Emergent transcatheter arterial embolisation of the feeding artery was performed (fig. 1e). 2 weeks later, after recovering from respiratory failure, lobectomy was performed because of persistent bleeding and mild fever despite antibiotics. A massive haemorrhage within the intralobar sequestra with a thrombosed feeding artery as a result of the intravascular coil was confirmed (fig. 1f).

Pulmonary sequestration is a rare sporadic developmental abnormality in which a region of the lung parenchyma has abnormal connection with the airways and is supplied by an aberrant artery arising from the aorta or one of its branches. Most sequestration are intralobar (75–85%), with incomplete communication with the adjacent lung and venous drainage *via* the pulmonary veins. They are generally observed in the medial or posterobasal segments of the left (60%) and right (40%) lower lobes [1]. It presents most commonly in childhood with recurrent infections, although massive haemoptysis in young adulthood has been reported [2, 3]. Conversely, extralobar sequestrations are characterised by the absence of connection with the airway, venous drainage *via* systemic veins, left lower lobe predominance, male predilection and frequent concomitant congenital abnormalities. They generally present in the neonatal period as left-to-right cardiac shunting, respiratory distress and cyanosis (large sequestration), or are found as an incidental finding later in life.

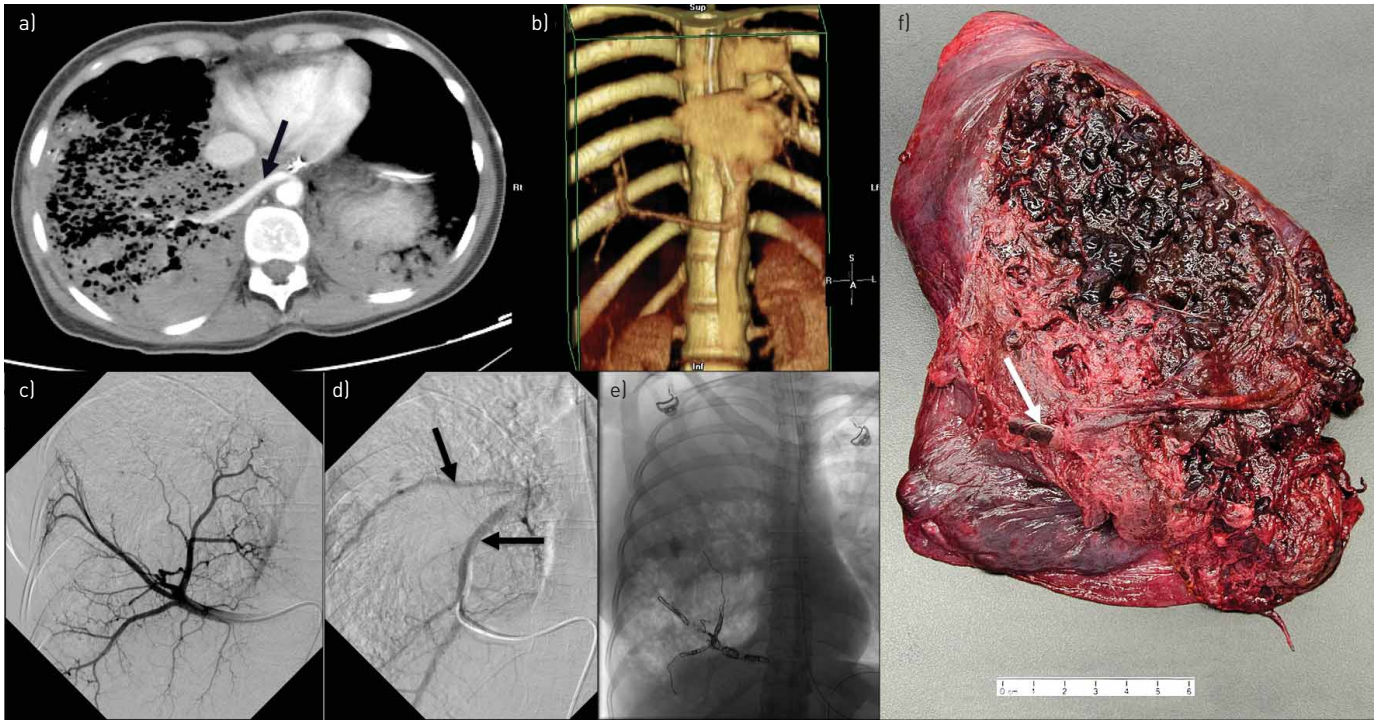

FIGURE 1 a) Multidetector computed tomographic angiography with b) three-dimensional reconstruction showing the aberrant systemic artery (arrow in a) arising from the supra-diaphramatic aorta. The aberrant systemic artery originating from the aorta was c) visualised on aortography with d) drainage into the pulmonary veins (arrows). e) The anomalous vessel was selectively catheterised and embolised. f) Pathological examination revealed large sequestration with massive haemorrhage supplied by a thrombosed 6-mm wide feeding artery (arrow) with no evidence for malignancy or cystic adenomatoid malformation. Scale bar = 6 cm.

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Intralobular pulmonary sequestration may present in adulthood as massive haemoptysis
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