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IMAGING IN INTENSIVE CARE MEDICINE

A patient with dermo-chondro-corneal dystrophy (François syndrome) and acute dyspnea

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A 38-year-old male with known François syndrome was admitted to the intensive care unit for acute respiratory failure. A contrast-enhanced CT scan revealed a spontaneous pneumothorax, an approximately $8 \times 7 \times 9$ cm partly thrombosed aneurysm of the left subclavian artery with signs of tracheal deviation and stenosis as well as multiple intrapulmonary calcifications and vascular aneurysms (Fig. 1). François syndrome is a rare genetic disorder with fewer than 20 reported cases. It is characterized by a triad of xanthomatosis, deformities of the extremities and corneal dystrophy. These patients are challenging to manage in an intensive care setting: Pulmonary and vascular malformations may complicate airway management and the establishment of vascular access. Pulmonary calcifications, likely caused by calcified aneurysms or chronic renal failure and subsequent hypercalcemia, may predispose patients to pneumothoraces or complicate mechanical ventilation. Whether there is an association between François syndrome and a predisposition to vascular malformations is not known. Therefore, appropriate radiologic investigations should be performed in advance, whenever possible.

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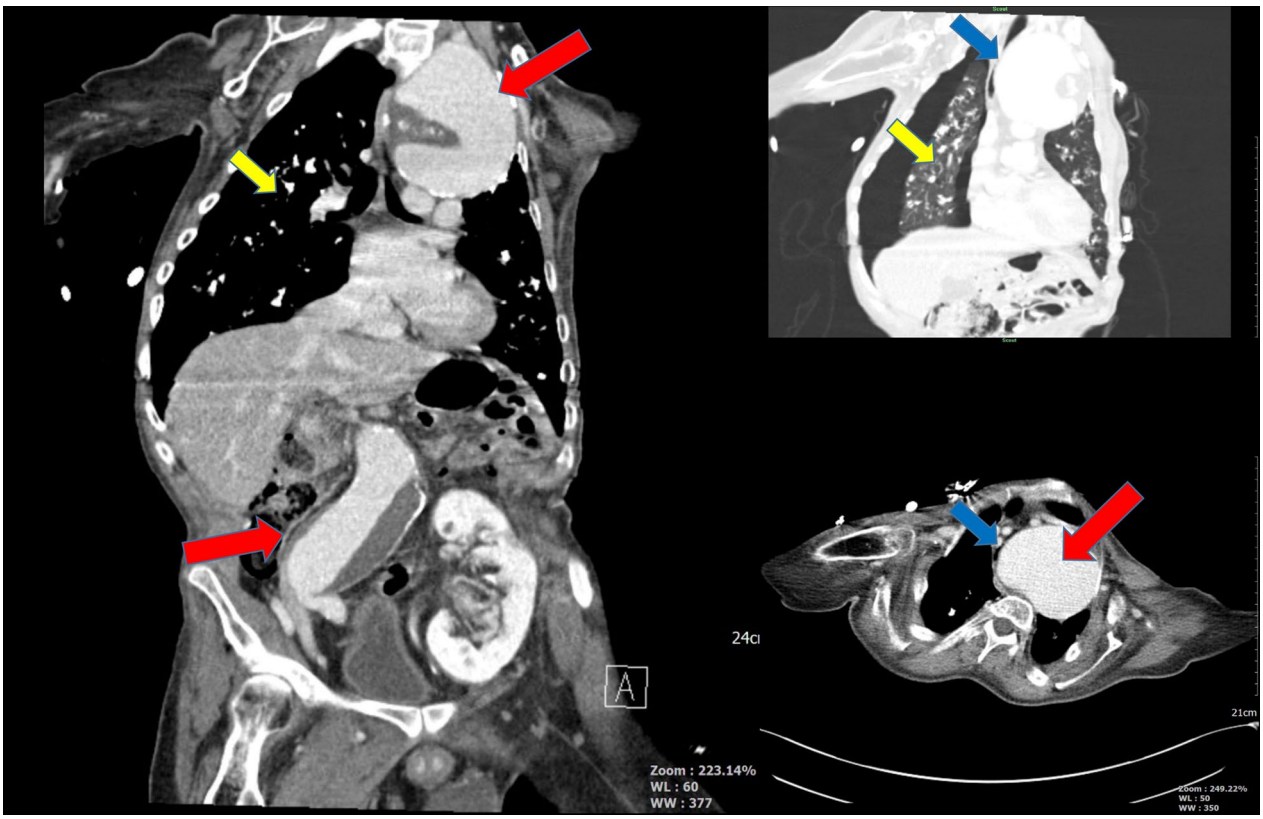

Fig. 1 Contrast-enhanced CT scan demonstrating tracheal deviation and stenosis (blue arrows), partly thrombosed aneurysms (red arrows) and pulmonary calcifications (yellow arrows)

Compliance with ethical standards

Conflicts of interest

On behalf of all authors, the corresponding author Dr. Gerrit Jansen states that there is no conflict of interest.

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