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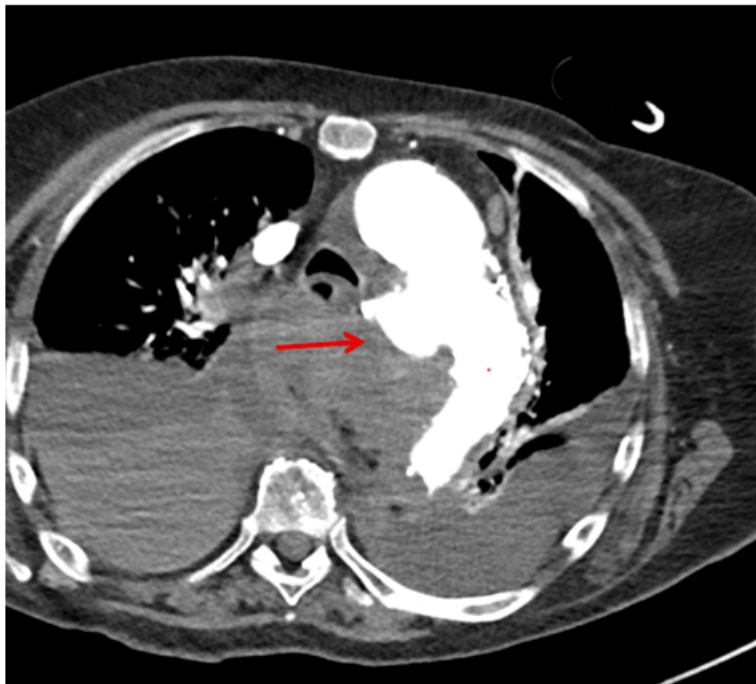
A “*lusus naturae*” dyspnea

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Descriptive legend

An 85-year-old female patient with a medical history significant for hypertension, chronic kidney disease, renal artery stenosis, Sjogren syndrome, and hypothyroidism presented to the emergency department with sudden severe dyspnea. Physical examination revealed a blood pressure of 130/80 mmHg and a heart rate of 120 bpm. The electrocardiogram showed sinus tachycardia. Point-of-care Ultrasound (US) documented a moderate bilateral pleural effusion, normal cardiac function, and a collapsible inferior vena cava. Compression US excluded lower extremity deep vein thrombosis. An arterial blood gas showed lactic acidosis (pH 7.31, lactates 36 mg/dL – normal value < 15) and a severe respiratory failure (PaO₂/FiO₂ ratio of 167). Enhanced Computed Tomography (CT) of the thorax was performed.

Question

Based on the CT imaging, what is the most likely diagnosis?

1. aortic dissection
2. oesophageal diverticulum
3. saccular aneurysm of the thoracic aorta
4. ruptured Kommerell's diverticulum

Answer

The correct answer is ruptured Kommerell's Diverticulum (KD) (Figure 1). First described in 1936, KD is a rare aortic anomaly characterized by a diverticulum that arises from either the left or the right aortic arch, as in our patient (Figure 2).¹ This condition results from a developmental error with a remnant of the 4th dorsal aortic arch. The prevalence of aberrant subclavian arteries is estimated to be 0.7 – 2%, and 20 – 60% of aberrant subclavian arteries have an associated KD.² Aneurysms of KD occur in 3 – 8% of cases as a fusiform dilation of the diverticulum at the origin of the aberrant subclavian artery. Rupture can occur in approximately 19% of cases.³ Recent

histological studies have shown the presence of cystic medial necrosis in the diverticulum wall, which explain the high rates of dissection and rupture.⁴

In most of the patients KD is asymptomatic but can cause symptoms of oesophageal or tracheal obstruction.⁵ Pediatric patients often present with airway symptoms⁶ whereas dysphagia⁷ (known as “dysphagia lusoria”) and chest discomfort⁸ are more common in the adult patients. The major complications are rupture or dissection.⁹ CT or magnetic resonance imaging can provide details of KD, associated arch anomalies, and its relationship with surrounding organs.¹⁰ Uncomplicated KD, saccular aneurysm of the thoracic aorta and ductus diverticulum are mimics of Kommerell aneurysm. The differential diagnosis is based on the origin of the aneurysm, outpouching contents, and morphology.¹¹

Due to its rarity, there are no established management guidelines. Surgical intervention should be considered when the diameter of the diverticulum orifice exceeds 30 mm and/or when the diameter of the descending aorta adjacent to the diverticulum exceeds 50 mm.⁵ Treatment options include open surgical repair, hybrid endovascular repair, and total endovascular repair, based on the anatomy, comorbidities of the patient, and surgical expertise available.¹²

Given the patient’s advance directives, that excluded surgical intervention, and in the absence of cardiovascular indications, we started palliative sedation. The patient died one hour later.

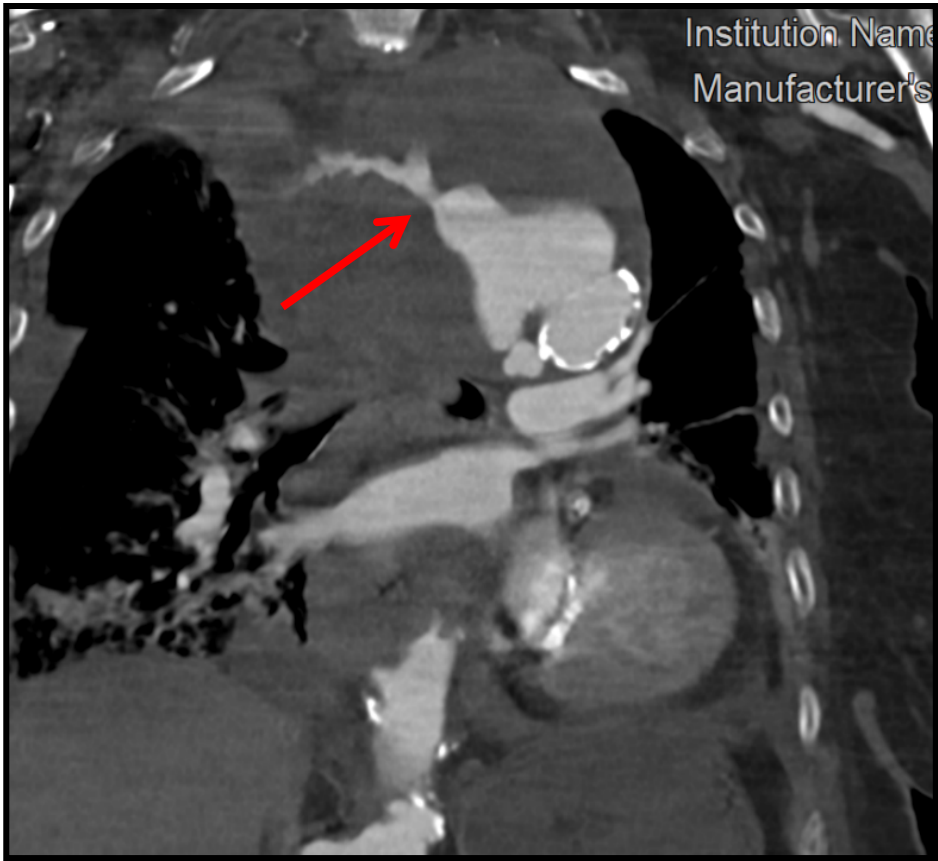


Figure 1. Enhanced CT (sagittal arterial phase) showing the ruptured Kommerell's diverticulum (red arrow).

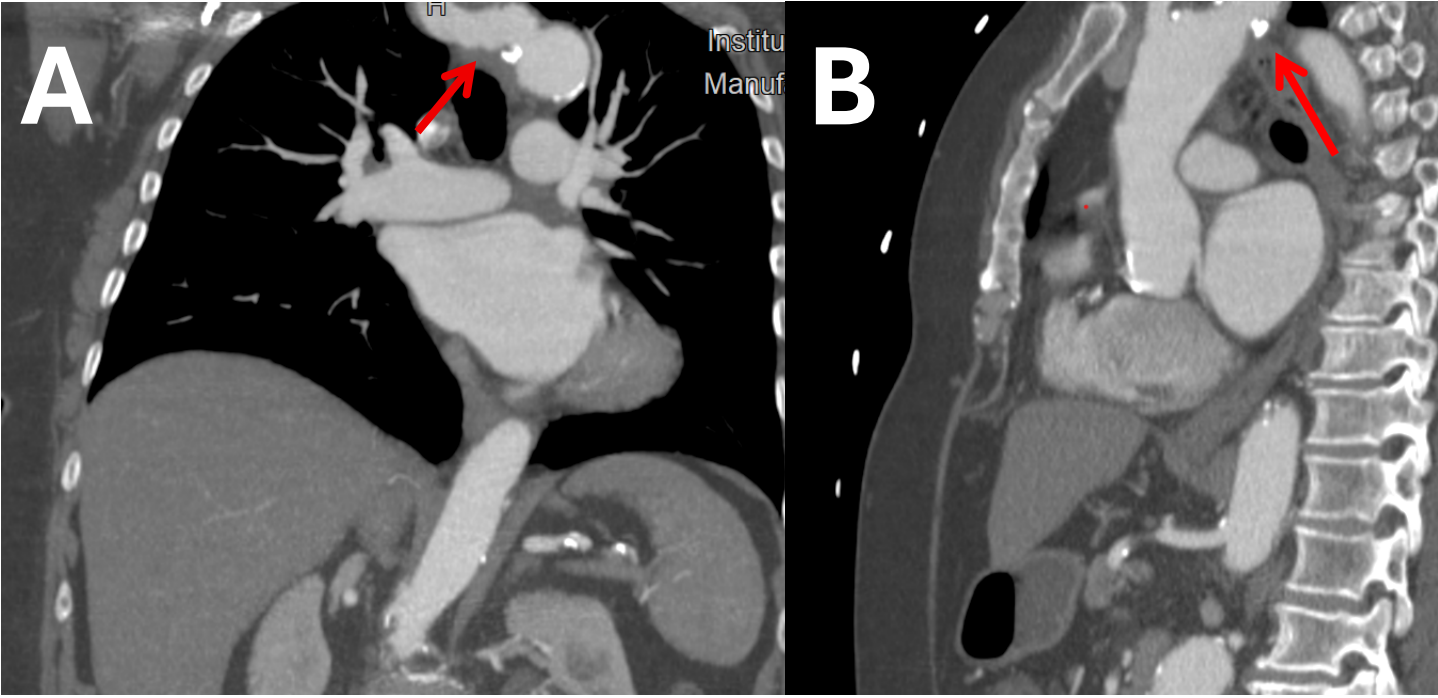


Figure 2. Enhanced-CT of the thorax (dated 2013) showing an aberrant right subclavian artery (arteria lusoria) with Kommerell's diverticulum (red arrow) (Panel A, coronal arterial phase; Panel B, sagittal arterial phase).

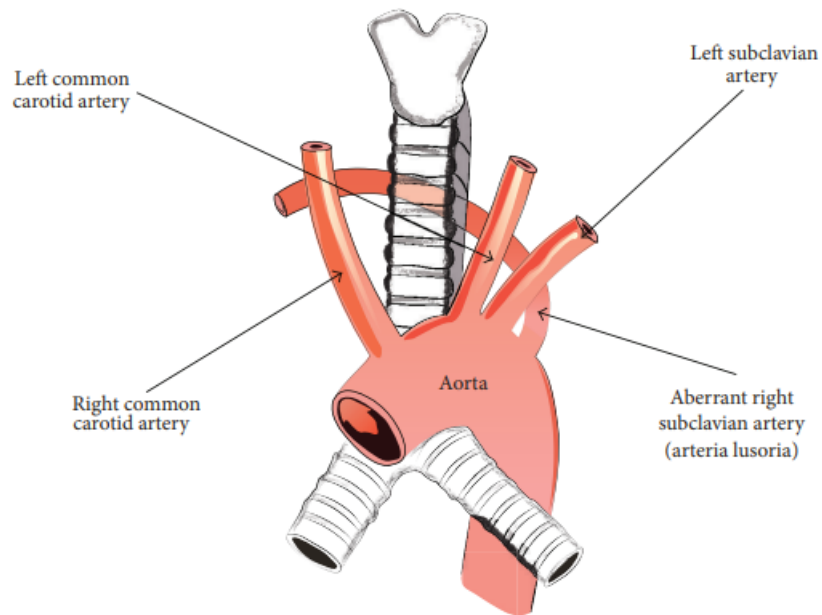


Figure 3. Aberrant right subclavian artery (arteria lusoria).¹³

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Conflicts of interest: Erika Poggiali is member of the editorial board of Emergency Care Journal. This work was not supported by any grant.

Availability of data and materials: the datasets used during the current case report are available from the corresponding author on reasonable request.

Ethics approval and consent to participate: as this was a descriptive case report and data was collected without patient identifiers, ethics approval was not required under our hospital's Institutional Review Board guidelines.

Informed consent: the patient provided consent for the access to medical records at the time of admission.