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**Case Report**

# Spontaneous innominate artery rupture in a patient with systemic sclerosis

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**Abstract**

We present the case of a 57-year-old female with systemic sclerosis who presented in extremis to our hospital with an acute onset of right upper chest and neck pain with swelling. She deteriorated rapidly due to haemodynamic compromise from suspected bleeding and suffered a cardiac arrest with prolonged resuscitation. Emergency thoracotomy demonstrated an acute longitudinal tear of the innominate artery/brachiocephalic trunk at the junction of the subclavian and common carotid arteries. This is the first reported case of spontaneous arterial rupture in a patient with systemic sclerosis, and while direct causation is difficult to prove, her history of previous vascular complications with potential ongoing microvascular damage makes a contributory role likely.

## CASE REPORT

We present the case of a 57-year-old female who presented in extremis to our emergency department with an acute onset of right upper chest and neck pain with swelling, immediately after bending down in her garden. She had a background of systemic sclerosis complicated by a renal crisis 15 years previously, for which she was on mycophenolate mofetil maintenance therapy. She also had rectal carcinoma in 2016 with subsequent abdominoperineal resection and end-colostomy formation.

After developing dyspnoea within minutes, she was transported to hospital via ambulance where her neck swelling worsened, causing hypoxia and cardiorespiratory arrest. Endotracheal intubation was not possible due to severe tracheal deviation, and an emergency surgical tracheostomy was performed. Sustained return of spontaneous circulation was achieved after 60 minutes, with multiple instances of loss of cardiac output in the interim. She continued to bleed profusely from her tracheostomy site and was transferred to theatre immediately for emergency surgery due to suspicion of ongoing active bleeding within the upper chest. Clamshell thoracotomy was performed which revealed an acute longitudinal tear of the innominate artery/brachiocephalic trunk at the junction of the subclavian and common carotid arteries. This was successfully repaired; however, the patient died on the intensive care unit 48 hours later of multi-organ failure and hypoxic brain injury.

## DISCUSSION

Systemic sclerosis (SSc) is an immune-mediated disorder resulting in fibrosis of the skin and internal organs as well as a fibroproliferative vasculopathy [1]. Changes in the microvasculature and endothelium are a prominent feature of SSc, but macrovascular changes related to atherosclerosis can be accelerated as in other autoimmune conditions such as rheumatoid arthritis and systemic lupus erythematosus, leading to a higher mortality risk from cardiovascular disease [2].

Arterial aneurysms and dissections have been occasionally reported in SSc patients, with focal arteritis affecting small and medium sized vessels thought to be the underlying pathogenesis for aneurysmal changes in connective tissue disease [3]. Long-term steroid usage may have a contributory role in vascular fragility [4], and there is a suggestion that prolonged steroid use may lead to disintegration of the intima media [5]. Intracranial aneurysms have been reported in the context of CREST syndrome; however, these vessels may be particularly susceptible to focal changes due to the lack of perivascular support [3].

Non-traumatic innominate artery rupture is an extremely rare event. Certain conditions may predispose to this outcome including aneurysms from degenerative arterial disease, Takayasu's disease, Marfan's syndrome and syphilis [6]. Invasive infection, radiotherapy, malignant invasion and heritable vascular conditions such as Ehlers-Danlos syndrome may also predispose to rupture [6].

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To our knowledge, this is the first reported case of spontaneous arterial rupture in a patient with systemic sclerosis. Our patient did not have any of the above risk factors and aneurysmal disease was not found at the time of surgical repair. Tracheostomy can lead to innominate artery injury [7] but is unlikely to be the cause here given the nature of presentation and pronounced haemodynamic compromise prior to insertion. Whilst direct causation from her systemic sclerosis is difficult to prove, her history of previous vascular complications in the form of a renal crisis along with potential ongoing microvascular damage, in the absence of known risk factors for spontaneous rupture, makes a contributory role to her fatal outcome likely.

## CONFLICT OF INTEREST STATEMENT

The authors declare we have no conflicts of interest.

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## ETHICAL APPROVAL

Not required.

## CONSENT

*Written* consent for publication has been obtained from the next of kin.

## GUARANTOR

Dr Sabreen Ali.

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