A case of misdiagnosed arterial thoracic outlet syndrome as primary Raynaud's phenomenon

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ABSTRACT

Arterial thoracic outlet syndrome (aTOS) is a rare, but potentially, limb-threatening condition that is often misdiagnosed. We present the case of a 29-year-old man who was initially managed under the presumption of primary Raynaud's phenomenon for >1 year before the correct diagnosis of aTOS, and the delay in diagnosis was complicated by substantial distal thromboembolic occlusion. Successful staged treatment included thoracic outlet decompression, subclavian artery aneurysm repair with subclavian-to-axillary bypass, anticoagulation, and an unconventional axillary-to-ulnar artery bypass. This report highlights the diagnostic challenges of aTOS and the importance of considering it in patients with Raynaud's phenomenon and vaso-occlusive symptoms. (J Vasc Surg Cases Innov Tech 2024;10:101508.)

Keywords: Arterial thoracic outlet syndrome; Raynaud's phenomenon; Vascular surgical procedures

Arterial thoracic outlet syndrome (aTOS), the rarest form of TOS, is an objective abnormality of the subclavian artery caused by extrinsic compression and subsequent damage by an anomalous first rib or analogous abnormal structure (cervical rib or band) at the base of the scalene triangle.[1,2] aTOS is often seen in otherwise healthy young adults who participate in repetitive overhead activities, and patients can remain asymptomatic or present with acute thrombosis, chronic stenosis, nonthrombotic ischemia, distal embolization, or total occlusion.[1-4] We present a case of a 29-year-old man who was initially misdiagnosed with primary Raynaud's phenomenon (RP) for >1 year before the correct diagnosis of aTOS, which was complicated by significant distal thromboembolic occlusion. This report describes a successful multistage treatment plan that included thoracic outlet decompression, subclavian artery aneurysm repair with subclavian-to-axillary bypass, anticoagulation, and an unconventional axillary-to-ulnar artery bypass.

CASE REPORT

A previously healthy, 29-year-old, right-hand dominant, weightlifting man was referred to us for burnt-out aTOS. He was previously misdiagnosed and medically treated for RP.

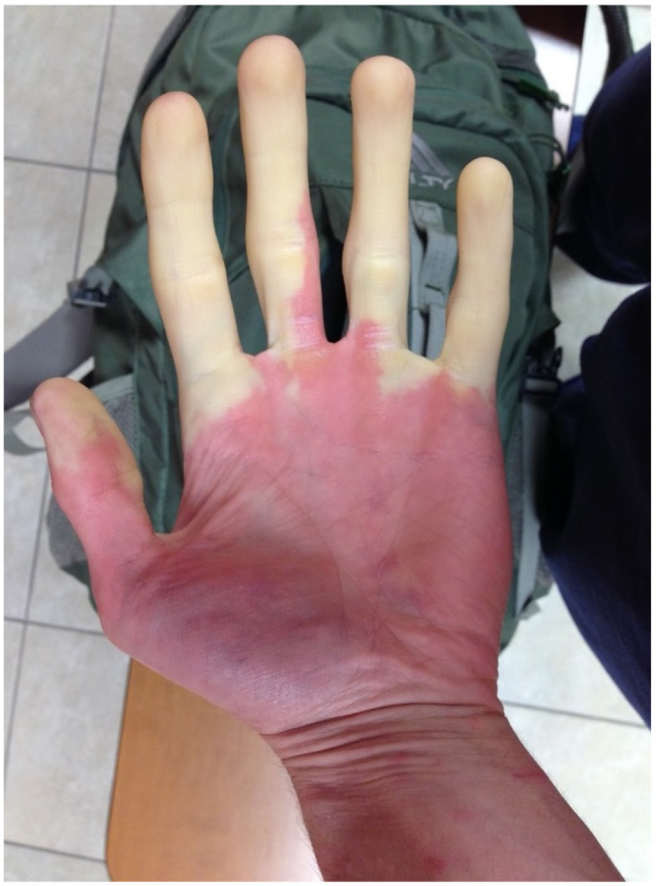

Fig 1. Photograph of the patient's left hand on initial presentation mimicking Raynaud's phenomenon (RP).

His symptoms began as blanching of the left fourth digit distal to the distal interphalangeal joint, which progressed to involve all five digits and part of the palm during a 6-month period (Fig 1). Cold, stress, and raising his arm triggered the blanching followed by hyperemia. A few months later, he developed worsening left forearm and hand claudication, increasing

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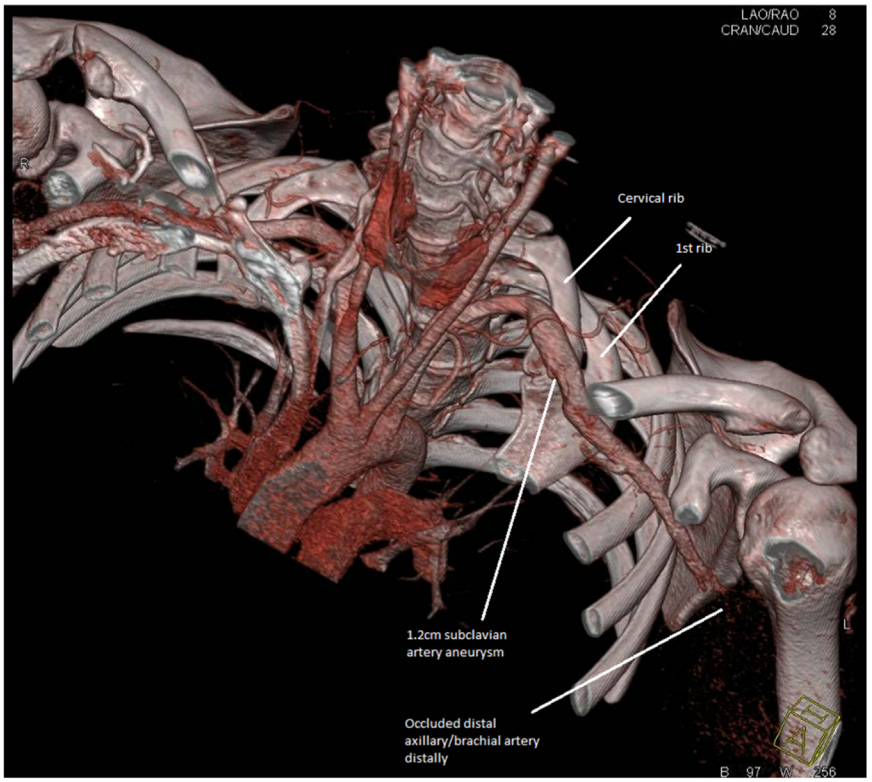

Fig 2. Three-dimensional reconstruction of computed tomography angiography (CTA) of the chest showing a complete cervical rib fused to the first rib, with an acute filling defect in the distal left axillary artery and extending into the brachial artery.

episodes of left hand paresthesia, tingling, burning sensations, and numbness. He also experienced significant muscular atrophy of his entire left upper extremity associated with weakness, limiting his ability to continue weightlifting and perform simple tasks such as tying his shoelace. Rheumatology initially managed the patient as having primary RP and started a course of prednisone for concern of vasculitis. However, due to the worsening arm claudication and diminished left radial pulses, his evaluation eventually led to computed tomography angiography (CTA) of the chest (Fig 2), which revealed a left-sided (complete) cervical rib that was fused to the first rib, with a 12-mm subclavian artery aneurysm. A large acute-appearing filling defect was noted in the distal left axillary artery, extending into the brachial artery and the thoracodorsal artery, that was concerning for acute thromboembolism. He was instructed to present to the emergency department and was admitted for inpatient management. The patient subsequently underwent catheter-directed thrombolysis with tissue plasminogen activator with minimal improvement.

We then performed left supraclavicular and infraclavicular exposure for resection of the left cervical and first ribs and repair and resection of the left subclavian artery aneurysm with an interposition subclavian-to-axillary artery bypass using an 8-mm PROPATEN ringed polytetrafluoroethylene graft (W.L. Gore & Associates). Embolectomy of the subclavian, axillary, brachial, and ulnar arteries was also attempted through an arm incision but was too chronic to be successful. After the surgery, intravenous heparin was started and anticoagulation therapy was continued to allow for recanalization. Approximately 6 weeks later, due to inadequate symptom relief, a repeat angiogram was performed to look for potential recanalization and targets for further revascularization. The angiogram demonstrated a patent left subclavian artery bypass but a persistently occluded distal axillary artery, brachial artery, and radial artery. There was, however, recanalization of the left ulnar artery through collateral vessels that extended down to the superficial palmar arch with limited outflow in the digital vessels (Fig 3). As such, we performed a left axillary-to-ulnar artery bypass using the great saphenous vein (Fig 4, *A*). Both anastomoses were

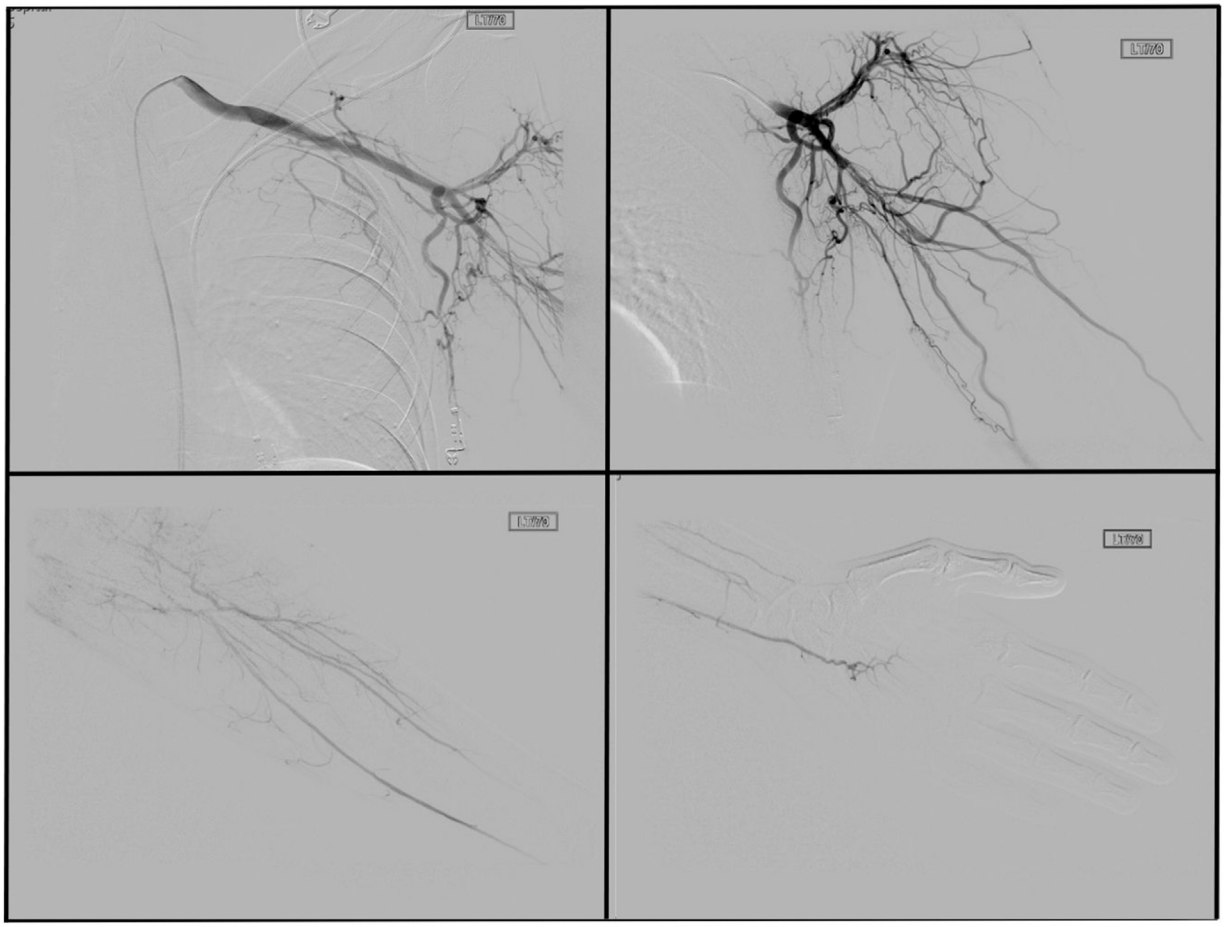

Fig 3. Angiogram after thoracic outlet syndrome (TOS) decompression showing a patent left subclavian artery bypass with recanalization of the left ulnar artery.

done in an end-side fashion. Prior thromboembolic events made axillary and ulnar artery dissection very challenging due to the presence of scar and inflammatory tissues. However, the remainder of the patient's postoperative course was uncomplicated, and he was discharged home with aspirin 81 mg and rivaroxaban (Xarelto; Janssen Pharmaceuticals) 20 mg. Subsequently, the patient returned to work and weightlifting with minimal difficulty and had no issues at his 5-year follow-up. The surgery site healed well (Fig 4, *B and C*), and his arm and hand ischemic symptoms resolved (Fig 4, *D*). Left upper extremity arterial duplex ultrasound at 3 years postoperatively showed widely patent subclavian-to-axillary bypass and axillary-to-ulnar artery bypass (Fig 5). The patient provided written informed consent for the report of his case details and imaging studies.

DISCUSSION

Unilateral hand or digital ischemia is a common presentation of aTOS and is likely due to proximal embolization from the subclavian artery, most commonly an aneurysm.[5] RP is a vasospastic disorder characterized by color changes in the affected fingers, typically in response to triggers such as cold or stress.[6-8] There are two forms of RP: primary and secondary. Primary RP is the most common form and is a benign idiopathic condition. Conversely, secondary RP is associated with a broad range of underlying diseases, most commonly rheumatic diseases.[6-8] We would like to emphasize that secondary RP should not be a standalone diagnosis, and patients should be treated for the primary disease.

aTOS is one of the causes of secondary RP and should not be overlooked because it can lead to limb-threatening ischemia.[3,9,10] A prior study revealed that individuals with primary RP who exhibit TOS-related symptoms cannot be distinguished from those without such symptoms when considering factors such as family

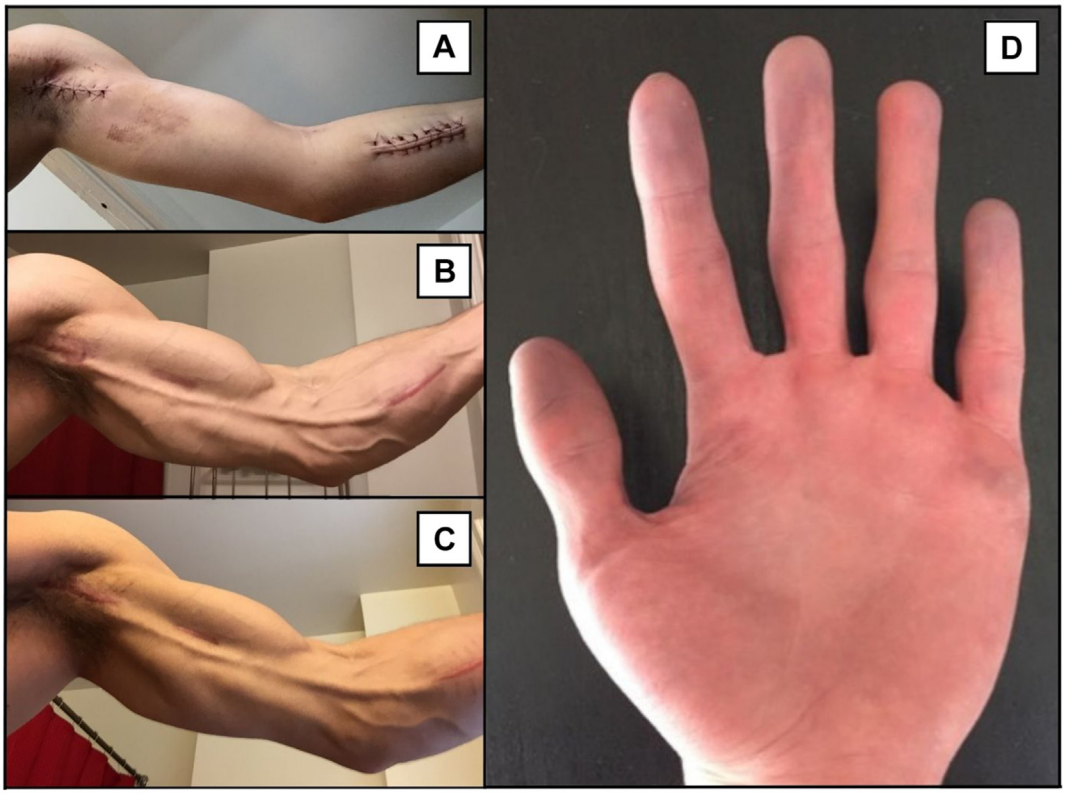

Fig 4. Photographs of the patient's arm after left axillary-to-ulnar artery bypass immediately postoperatively (**A**), at 24 months postoperatively (**B**), and at 60 months postoperatively (**C**) and resolution of Raynaud's phenomenon (RP) of the left hand (**D**).

history, sex, thumb involvement, and asymmetry.[11] In our case, the patient had been initially treated for primary RP for >1 year. One possible explanation for this underdiagnosis might be rooted in a positive family history of RP, a risk factor that is present for approximately one half of all primary RP patients.[8] Additionally, during the initial assessment, he only presented with cutaneous manifestations, and the autoimmune workup was negative, aligning with some of the proposed criteria for primary RP.[12] However, certain features were pointing toward a diagnosis of aTOS for our patient. His symptoms were unilateral, and he later developed vaso-occlusive symptoms such as claudication, muscular atrophy, and weakness, with a diminished left radial pulse. Notably, only about 7% of patients with primary RP present unilaterally,[5] highlighting the importance of conducting a comprehensive clinical assessment to rule out secondary causes.[8] It is essential to recognize that the absence of radial and ulnar pulses in aTOS can be inconsistent.[5] Therefore, the presence of distal pulses should not exclude arterial occlusion as the cause of secondary RP. In addition to aTOS, hypothenar hammer syndrome should also be considered in the differential diagnosis, especially in working-age individuals presenting with unilateral hand ischemia. A comprehensive pulse examination, complemented by imaging modalities such as CTA, can effectively guide the diagnosis.

Our patient underwent a staged treatment plan that included thoracic outlet decompression, involving the removal of a cervical rib and resection of the first rib, aneurysm resection, and replacement by a prosthetic graft to eliminate the source of thromboembolism. To address the aneurysm, a subclavian-to-axillary interposition bypass was performed, complemented by anticoagulation therapy to facilitate recanalization before the bypass. Our case serves as an illustrative example of the feasibility of an unconventional bypass from the axillary artery to the ulnar artery.

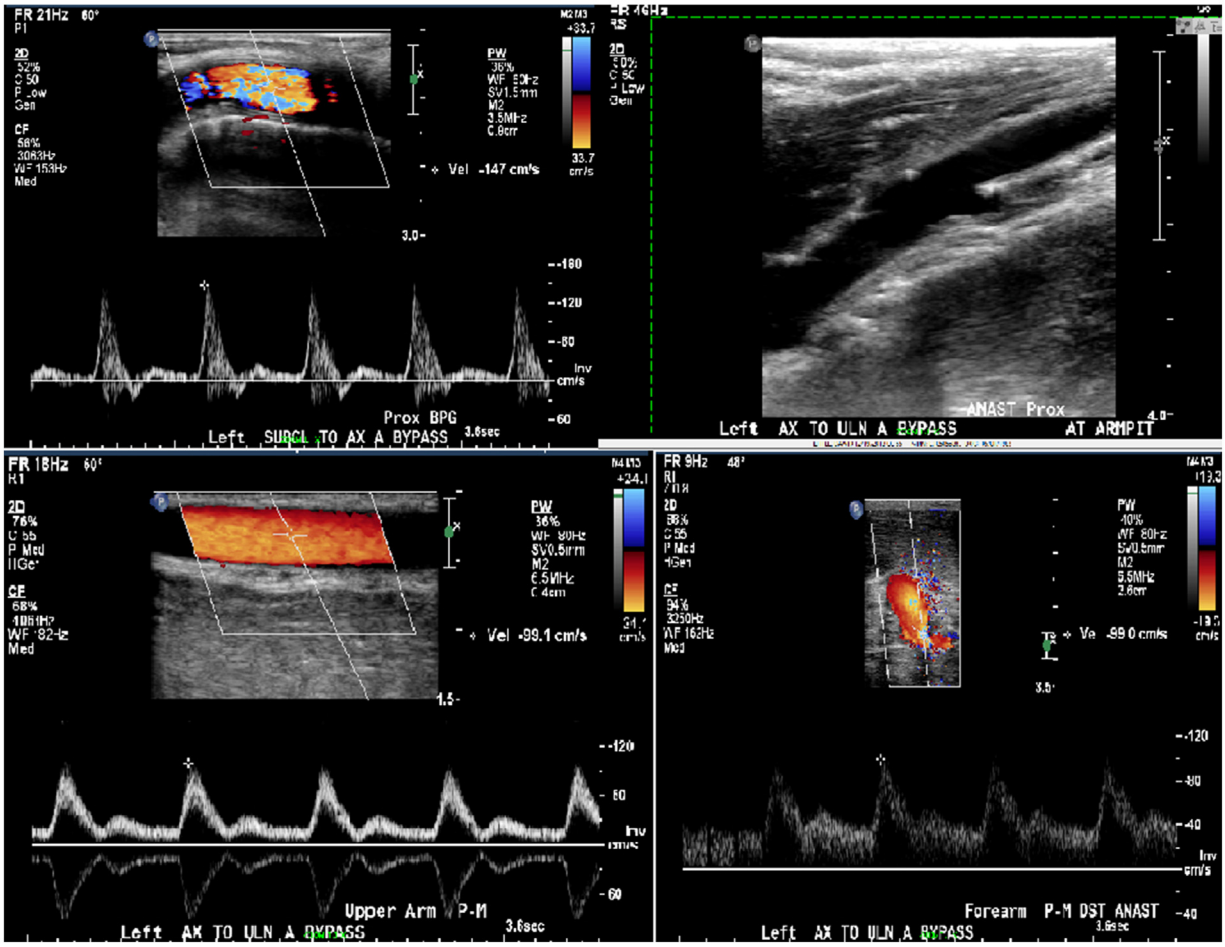

Fig 5. Left upper extremity arterial duplex ultrasound at 3 years postoperatively demonstrating patent left subclavian-axillary artery bypass and patent left axillary to ulnar artery bypass.

CONCLUSIONS

aTOS is a rare, but serious, condition that can cause significant morbidity and disability. It is often misdiagnosed because the symptoms can mimic other conditions, such as RP, as shown by this unique case of chronic aTOS that was missed for >1 year. CTA eventually revealed significant thromboembolic occlusion distally, necessitating staged treatment. aTOS should be considered when RP is associated with vaso-occlusive symptoms such as claudication, muscular atrophy, and weakness.

DISCLOSURES

None.

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