Short Report

Upper Limb Ischaemia in a Young Female

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Abstract

We present the case of an acute or chronic ischaemia of the upper limb due to extrinsic compression and thrombotic occlusion of the left subclavian artery successfully bypassed with a subclavian to axillary artery venous auto-graft.

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Introduction

Acute limb ischaemia in teenage females without a past history of vascular disease is uncommon. This is a case report of acute subclavian artery occlusion in a young female displaying features of a congenital vascular–skeletal malformation syndrome.

Case Report

A 15-year-old left-handed Caucasian female presented with a 1-day history of cold left hand and severe pain affecting the left upper limb. Pain was exacerbated on minimal exertion in the left upper and lower arm and hand preventing her from combing her hair and writing. There was no history of trauma. She had almost a year of day history of cold left hand and severe pain affecting the left upper limb. Pain was exacerbated on minimal exertion in the left upper and lower arm and hand preventing her from combing her hair and writing. There was no history of trauma. She had almost a year of hearing impairment. She also had a phobia for needles.

On examination of the left hand of the patient, she had signs of ischaemia with pallor, coldness and slow capillary refill. Sensation and movement were intact and the forearm and arm muscles were non-tender. No muscle wasting was observed in the hand. On the left, there was a proximal left subclavian pulse but no pulses palpable beyond that point and moderate tenderness in the supraclavicular fossa with a palpable bony swelling. She had a full range of movement in her neck. She had a full set of pulses in the right arm. Radiographs of the spine showed a T1 hemivertebra and abnormal C5/6/7 spinal processes and a marked scoliosis to the left. A congenital abnormal left 1st rib shortened and fused to the 2nd rib was found (Fig. 1). The Duplex scan lab confirmed fresh thrombus in the subclavian artery (distal to the vertebral) to the distal axillary artery. A magnetic resonance (MR) angiogram showed left subclavian artery occlusion distally to its mid second part. The very distal axillary/brachial arteries were patent, fed by a collateral but no detailed run-off view of the arm was viewed (Fig. 2).

In view of her clinical presentation and radiological evidence a diagnosis of arterial thoracic outlet syndrome was made, possibly caused by the congenitally abnormal 1st rib damaging the distal portion of the subclavian artery. Her blood was sent for a thrombophilia screen due to the family history and Raynaud's type symptoms in the past. She was commenced on a heparin infusion while awaiting surgery. Due to the acute or chronic presentation, the immense phobia for needles and the risk of embolisation into vertebral artery, thrombolysis was not contemplated.

In the next 24 h during which time there was no further deterioration and there was marginal improvement with intravenous heparin, she proceeded to have an excision of the abnormal left 1st rib to widen the costo-clavicular canal. Subsequently,
thrombectomy and exploration of the subclavian and axillary arteries revealed increased wall thickness and intimal ulceration. The decision was taken to bypass this portion of artery to minimise the risk of further thrombosis and distal embolism. The long saphenous vein from the left thigh was harvested to provide a subclavian to axillary artery bypass graft. Postoperatively, the hand returned to its normal colour and capillary refill; however, there was no palpable brachial pulse. The brachial artery was explored and found to have changes of chronic embolic occlusion. An on-table angiogram confirmed the brachial occlusion from the level of the mid-arm down to its bifurcation and prominent collateral from the proximal brachial feeding the ulnar artery in the forearm and non-visualisation of the radial artery. This was confirmed also by an immediate postoperative duplex examination which confirmed that the ulnar was the dominant artery in the forearm which had good biphasic signals in the wrist. The appearance of the arterial vasculature was consistent with multiple long-standing embolic episodes. Further revascularisation would necessitate a brachial-ulnar bypass, which was deferred in view of favourable clinical result.

The thrombophilia screen sent earlier revealed the presence of lupus anticoagulant antibodies and the patient was started on warfarin in agreement with haematology advice. She recovered well after surgery with a symptom-free left arm and a warm well-perfused hand. Her graft was found to be patent on duplex assessment during the 6-week and 6-month follow-up and she had good biphasic signals recorded in the ulnar artery. She was discharged on warfarin. Further to this she has been referred to the geneticist due to the structural anomalies and positive antiphospholipid status. She was seen in a review clinic 2 months and 6 months post-surgery and was able to perform all her activities without any symptoms.

**Discussion**

Arterial thoracic outlet syndrome (aTOS) in paediatric age group is uncommon and most case series have venous TOS. There have been case reports with presentation of stroke, neurogenic and arterial TOS following non-union of fractured first rib after a sport injury and elongated transverse process of 7th cervical vertebra causing aTOS. One case series highlighted the presence of scalenus minimus, hypertrophy of scalenus anterior and medius muscle and cervical rib variants as the common causes of paediatric TOS.

While vTOS has a strong association with hypercoagulable states, the same is not established about aTOS. This case is unusual as the patient presented with a structural abnormality which predisposed her to arterial thrombosis as well as a haematological abnormality which increased her risk of thrombosis further. The presence of lupus anticoagulant antibody in this patient was possibly inherited. The presence of structural abnormality could be related to a number of syndromes.

The patient had microtia of the right ear as well as the absence of the right middle ear canal previously diagnosed when investigated for hearing impairment. The presence of scoliosis and the auditory canal anomaly raised the possibility of oculo-auriculo-vertebral (OAV) syndrome, also known as Goldenhar syndrome. However, no other facial or ocular anomalies were noted and an echocardiogram and abdominal ultrasound showed no evidence of abnormal internal organ development. A possible diagnosis of Klippel–Feil syndrome was also raised on account of the marked scoliosis, and rib and auricular anomalies. However, the patient did not present vertebral fusion of the vertebral column, which is a cardinal feature of the syndrome.

**Conclusion**

This case is reported to raise awareness on the unusual presentation of thoracic outlet syndrome in a young female. It also
highlights the importance of investigation of unilateral symptoms suggestive of Raynaud’s phenomenon. The presence of pro-coagulant disorder and a costal malformation increased the risk for arterial thoracic outlet syndrome. To this date, the literature search has not shown a combination of extrinsic and intrinsic cause for acute or chronic arterial occlusive disease affecting the upper limb in a teenaged female patient.

Conflict of Interest/Funding

None.

References