SHORT REPORT

Pseudoaneurysm of the Peroneal Artery: Presentation of Ehlers-Danlos Syndrome Type IV

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Abstract
Introduction: Pseudoaneurysms in deep or unusual sites raise the possibility of an underlying vessel wall disorder.
Report: A 28-year-old woman presented with pain and swelling of her calf, with no history of trauma. Angiography diagnosed a peroneal artery pseudoaneurysm, which we embolised successfully. Subsequent genetic analysis revealed the COL3A1 mutation, confirming Ehlers-Danlos syndrome type IV.
Conclusion: To our knowledge, this is the first report of a peroneal artery pseudoaneurysm associated with underlying collagen vascular disease.

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Introduction
We present here a rare case of a peroneal artery pseudoaneurysm. The unusual site of the lesion, in the absence of trauma, prompted us to investigate this patient further.

Report
A 28-year-old female smoker presented to the emergency department with a two day history of pain and swelling of her right calf, after reaching up to some high shelves. There was no history of preceding trauma.
On further questioning, she reported a tendency to “bruise easily”. Her past medical history included an episode of acute abdominal pain the previous year, for which she underwent a CT scan. This showed a 9.5 cm haemorrhagic cyst arising from the left liver lobe. This was thought to be a subcapsular haematoma from an adenoma.
However, on a later CT scan, the “adenoma” had spontaneously resolved.

On examination, the patient was apyrexial and had a swollen right calf, with pain in the posterior compartment on palpation and dorsiflexion. All pulses were present and there was no motor deficit, however there was numbness in the distribution of the tibial nerve over the sole of her foot. Her inflammatory markers were minimally raised.

An ultrasound showed a well defined, pulsatile mass in the posterior compartment, consistent with a peroneal artery pseudoaneurysm. The patient was transferred to our vascular unit and underwent urgent angiography (Figs. 1 and 2).

This confirmed a wide-necked false aneurysm arising from the peroneal artery 1 cm distal to the bifurcation of the tibio-peroneal trunk, with a 1 cm sac. Both the anterior and posterior tibial arteries were patent despite some mild extrinsic compression from the hematoma.

The sac of the aneurysm was embolised with four 3 mm (diameter) × 3 cm (length) coils (Cook). It was not possible to pack the sac fully with coils without affecting the patency of the peroneal artery. Therefore, we embolised proximally and distally to the aneurysm with a total of six coils. There was successful occlusion of the aneurysm. The posterior and anterior tibial arteries remained intact at the end of the procedure.

The patient made a full clinical recovery and was discharged a few days later. She was subsequently referred to a clinical geneticist. DNA analysis revealed a mutation in the COL3A1 gene, leading to abnormal type III procollagen synthesis. This is diagnostic of Ehlers-Danlos syndrome type IV.

Discussion

Pseudoaneurysms of the peroneal artery are extremely rare. A review of the literature revealed only 28 case reports, with the vast majority traumatic in aetiology.1 Four cases were iatrogenic in nature,1 two infective2 and one a complication in Behcet’s disease.3 Therefore, to our knowledge, this is the first report of a peroneal artery pseudoaneurysm associated with underlying collagen vascular disease.

Importantly, this patient had no history of significant trauma. The development of a “spontaneous” pseudoaneurysm in such an unusual site led us to investigate her further.

Ehlers-Danlos syndrome Type IV, or “vascular” Ehlers-Danlos, is a rare disorder which is associated with multiple medical and surgical complications.4 Inheritance can be autosomal dominant, but many cases are sporadic mutations. This patient’s family members had no history of similar vascular events, but have been offered genetic testing.

As the clinical presentation is variable, there is often a delay in diagnosis.4 Features include easy bruising, a striking facial appearance, translucent skin with visible veins, and rupture of blood vessels, gravid uterus or intestines.4 There is no specific treatment for the disease, but severity varies widely between individuals. Patients benefit from genetic counselling and regular outpatient follow-up,
however often present acutely with ruptured vessels in unexpected locations. There is a poor prognosis and a reduced life expectancy, with many patients dying from artery dissection or pregnancy-related complications. Most surgeons are unfamiliar with genetic testing, and involvement of a clinical geneticist in suspected cases of collagen vascular disease is mandatory.

In conclusion, pseudoaneurysms of any vessel are most commonly associated with trauma. In the absence of a significant history, or if it is an unusual or deep site, the clinician must be alert to the possibility of underlying collagen vascular disease.

Conflict of Interest

The authors have no potential conflicts of interest.

References