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Sudden juvenile death: Simultaneous spontaneous dissection of the left extracranial internal carotid artery and the left common iliac artery

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Abstract Arterial spontaneous dissection is a pathological condition that recognises various causes, from hereditary diseases such as connective tissue diseases to traumatic events. Although the pathogenesis of arterial disease remains unknown in most cases, nevertheless, it is important to define possible causes.
We report on the case of a 19-year-old young man, smoker, who presented with a spontaneous dissection of the extracranial left internal carotid artery, and subsequently a spontaneous dissection and early rupture of the left common iliac artery which was the cause of death.
In our case, the histopathological findings at autopsy revealed features consistent with fibromuscular dysplasia, a rare non-atherosclerotic vascular disease that mainly involves medium-sized muscular arteries.

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PALABRAS CLAVE
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Arteria carótida interna;
Arteria ilíaca común;
Displasia fibromuscular;
Muerte súbita juvenil

Muerte súbita juvenil: Disección espontánea simultánea de la arteria carótida interna izquierda extracraneal y de la arteria ilíaca común izquierda

Resumen La disección arterial espontánea es una condición patológica que reconoce diversas causas, desde enfermedades hereditarias como las enfermedades del tejido conectivo hasta eventos traumáticos. Aunque la patogénesis de la enfermedad arterial sigue siendo desconocida en la mayoría de los casos, es importante definir las posibles causas.
Presentamos el caso de un joven de 19 años, fumador, que presentó una disección espontánea de la arteria carótida interna izquierda extracraneal y, posteriormente, una disección espontánea con ruptura temprana de la arteria ilíaca común izquierda, lo que fue la causa de su fallecimiento.
En nuestro caso, los hallazgos histopatológicos en la autopsia revelaron características compatibles con displasia fibromuscular, una rara enfermedad vascular no aterosclerótica que afecta principalmente a las arterias musculares de mediano calibre.

Introduction

Spontaneous arterial dissection can affect all arteries, with various causes such as trauma, connective tissue diseases like Marfan and Ehlers-Danlos syndromes, polycystic kidney disease, and fibromuscular dysplasia (FMD).[1–4] FMD, a non-atherosclerotic, non-inflammatory vascular disorder, typically impacts medium-sized muscular arteries, especially the renal arteries, and less frequently the internal carotid, vertebral, subclavian, and iliac arteries.[5–8]

First described in 1938, FMD was later named by McCormack in 1958,[5] with its aetiology likely multifactorial, with smoking possibly worsening the condition in those with multifocal lesions.[7] FMD is more common in women but tends to progress more rapidly in men, especially in smokers.[8] It can occur at any age, typically affecting young adults.[7]

FMD is often asymptomatic, though symptoms vary by artery location. Cervical artery involvement may cause migraine, pulsatile tinnitus, and dizziness, with neurological symptoms in cases of cerebral ischemia or embolism.[5,8] Iliac artery involvement may lead to lower limb ischemia, but dissection or rupture is rare.[9] Diagnosis is typically made using duplex ultrasound, CT, or MR angiography, with catheter-based arteriography as the gold standard.[8] Treatment depends on lesion location and clinical presentation.

Medical-forensic description

A 19-year-old male smoker (ten cigarettes per day), with a history of Achilles tendon surgery and recurrent unilateral pulsating headaches, was admitted to the emergency room. He experienced scotomas, dysarthria, dizziness, and weakness on the right side, following a left-sided headache two hours earlier. A similar, milder episode occurred three months prior. The neurological exam, CT scans, and lab tests were all negative, leading to a diagnosis of migraine with and without aura. The patient was prescribed home therapy and advised to undergo a brain MRI with angio-MRI.

Eleven days later, MRI and angio-MRI revealed a flow defect in the left internal carotid artery due to dissection. He was discharged with antiplatelet therapy and a follow-up appointment. However, five days later, after several syncopal episodes, he was re-admitted with a diagnosis of carotid dissection. His condition worsened rapidly; after a sudden collapse, he was intubated but suffered cardiac arrest. A CT scan revealed significant abdominal bleeding. Despite resuscitation efforts, a second cardiac arrest occurred, leading to death. The autopsy was performed with the toxicological tests being negative.

The autopsy confirmed a 4-cm dissection of the left internal carotid artery (Fig. 1A–B) with haemorrhagic infiltration. The right carotid artery was unremarkable. Dissection and rupture of the left common iliac artery (Fig. 1C) with surrounding tissue damage were observed, along with diffuse haemorrhagic infiltration in the abdominal cavity (Fig. 1D).

The heart weighed 330 g (reference value: 352.7 g), with dimensions of 11 cm transversely, 10.5 cm longitudinally, and 3.5 cm anteroposteriorly. Wall thicknesses: left ventricle anterior 1.4 cm, lateral 1 cm, posterior 1.3 cm; interventricular septum 1.4 cm; right ventricle 0.3 cm. The mitral valve showed myxoid degeneration. The aorta had lipid striae and a thinned wall. Microscopically, the left internal carotid (Fig. 2A) and common iliac artery (Fig. 2B) showed musculo-elastic atrophy and fibrosis (Fig. 2C-F), with some segments replaced by fibrous tissue.

The pathological diagnosis indicated diffuse medial FMD, affecting the left internal carotid and common iliac arteries, with dissection and rupture of the left iliac artery causing sudden acute death.

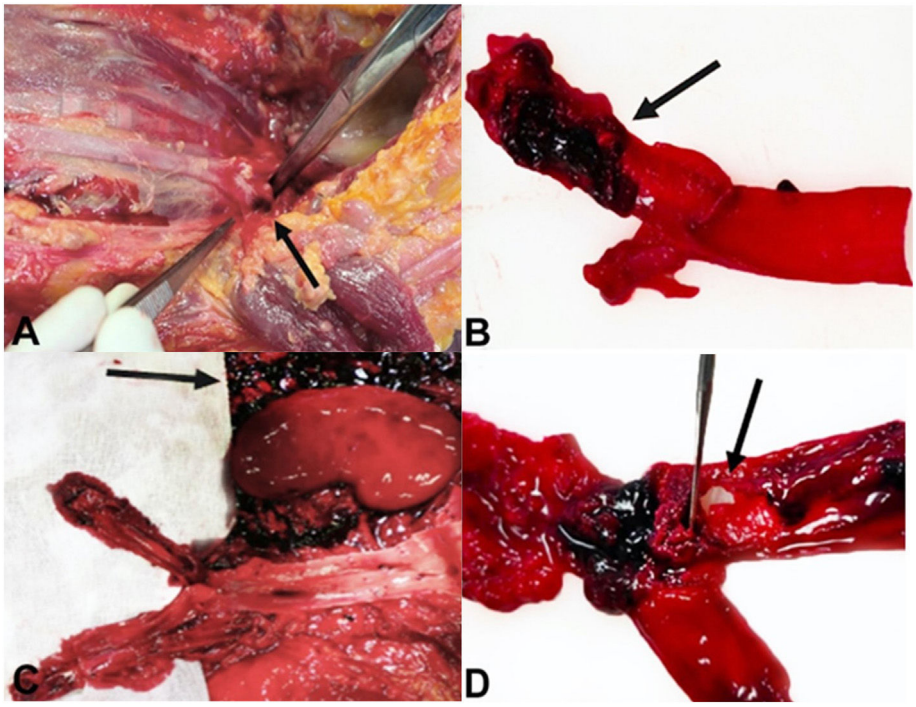

Figure 1 (A and B) Dissection (black arrow) of extracranial left internal carotid artery. (C) Haemorrhagic infiltration of retroperitoneal perivascular soft tissues (black arrow). (D) Dissection and rupture (black arrow) of left common iliac artery.

Discussion

We present the case of a 19-year-old male smoker with a history of unilateral pulsatile headaches 2–3 times per month, who died suddenly. His family history was negative for cardiovascular diseases or sudden deaths. The autopsy revealed dissection of the left internal carotid artery and the left common iliac artery, along with rupture and haemorrhagic infiltration of the arterial walls and abdominal soft tissues, primarily on the left side. Histologically, the affected arteries showed significant musculo-elastic atrophy and fibrosis, with similar features observed in all examined vessels. These findings point towards a systemic arterial disease, likely FMD, which typically affects medium and large arteries.

FMD often involves the carotid artery, and rarely, the iliac artery.[9] In this case, both the left internal carotid and common iliac arteries, along with other vessels, were affected. FMD predominantly affects women, but when it occurs in smoking men, especially young adults, it is more severe and typically presents with asymptomatic or non-specific symptoms.[7,8] This patient's clinical history of unilateral pulsatile migraine further supports the FMD diagnosis.

Pathologically, FMD is classified into five subtypes based on the arterial wall layer involved: intimal fibroplasia, medial hyperplasia, medial fibroplasia (most common), perimedial fibroplasia, and adventitial fibroplasia.[5,10,11] Some authors argue that medial hyperplasia may be a precursor to medial fibroplasia.[11–13] In this case, the media, characterised by disorganisation due to elastic fibre atrophy and fibrosis, was predominantly affected, with mild involvement of the adventitia. Additionally, haemorrhagic infiltration of the soft tissues in the left iliac fossa supports the fatal dissection and rupture of the left common iliac artery.

FMD may have a genetic component, with rare familial cases and a suspected autosomal dominant inheritance pattern. Recent studies have identified the rs9349379-A SNP in the PHACTR1 gene, potentially influencing endothelin-1 regulation and vascular remodelling, though genetic testing is not yet clinically recommended.[14]

Differential diagnosis with other causes of spontaneous dissection, such as Marfan Syndrome, Ehlers-Danlos Syndrome, and others, is crucial for clinical purposes and may have legal implications. These conditions can present with overlapping vascular manifestations like aneurysms, dissections, and tortuosity. However, unlike FMD, these syndromes are caused by specific genetic mutations and are often associated with systemic connective tissue signs, such as joint hypermobility, fragile skin, or distinct physical traits. For instance, Marfan syndrome commonly involves ascending aortic dilatation and tall stature. Distinguishing between these conditions requires careful clinical evaluation, imaging, and genetic testing.[15]

Conclusions

In the presented case, the autopsy demonstrated the occurrence of dissection of the extracranial left internal carotid artery, associated with dissection and rupture of the left common iliac artery, and haemorrhagic infiltration of

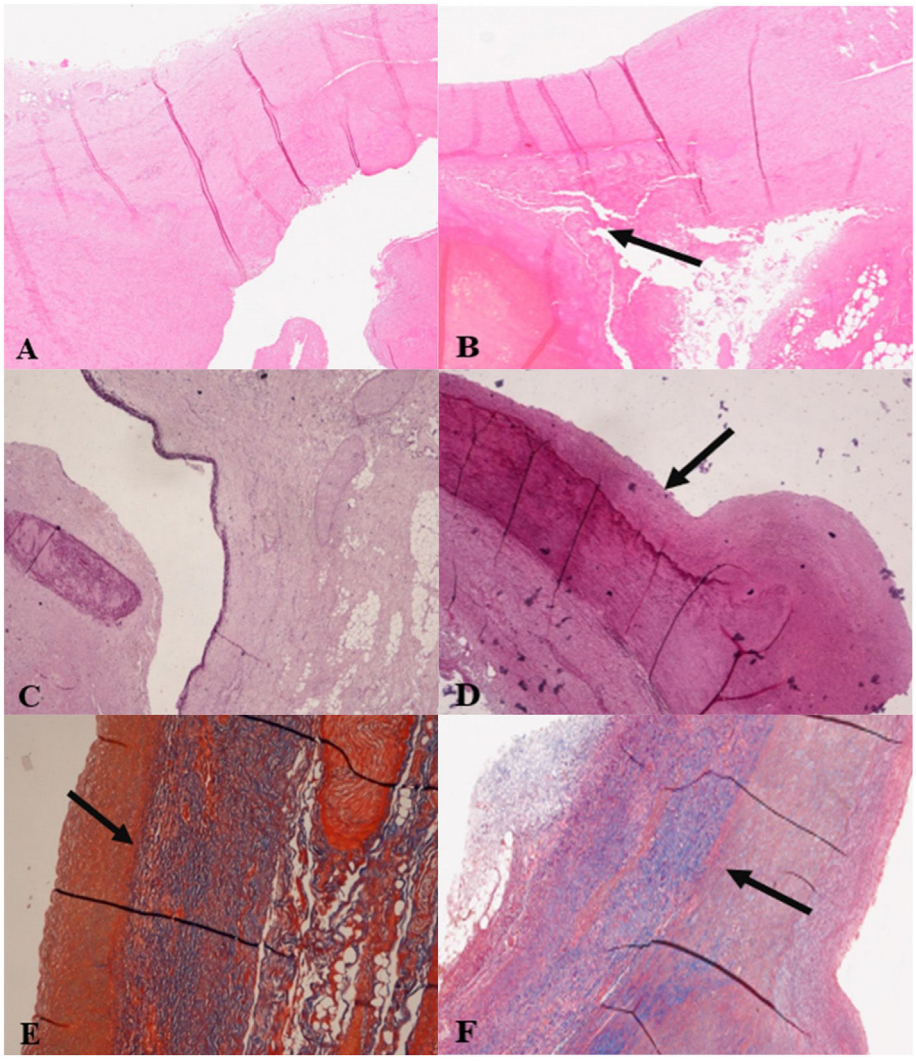

Figure 2 (A) Dissection and haemorrhagic infiltration of extracranial left internal carotid artery (HE; 2.5×). B) Dissection and haemorrhagic infiltration (black arrow) of left common iliac artery (HE 2.5×). (C) Extracranial left internal carotid artery characterised by marked muscular-elastic atrophy of tunica media (Elastic stain; 4×). (D) Left common iliac artery characterised by marked muscular-elastic atrophy (black arrow) (Elastic stain; 4×). (E) Extracranial left internal carotid artery characterised by medial and adventitial fibrosis (black arrow) with altered parietal structure (Trichrome stain; 4×). (F) Left common iliac artery with medial and adventitial fibrosis (black arrow) with damaged parietal structure (Trichrome stain; 4×).

the left iliac fossa, which suggests that death occurred due to acute and unpredictable rupture of the left common iliac artery.

Furthermore, histology showed diffuse and severe abnormalities of the medial layer of the arterial wall of all the arteries studied, which suggests a systemic arterial disease responsible for the dissection and rupture, consistent with the diagnosis of Fibromuscular Dysplasia. Therefore, FMD should be considered as a possible cause of acute dissection especially in young individuals with no other predisposing factors for cardiovascular diseases.

Informed consent statement

Informed consent for publication was obtained from the parents of the patient.

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CRediT authorship contribution statement

Cecilia Salzillo: Conceptualization, Investigation, Writing – original draft. **Andrea Marzullo:** Writing – original draft, Writing – review & editing. **Stefania Galizia:** Conceptualization. **Domenico Angiletta:** Investigation, Writing – review & editing. **Antonio De Donno:** Supervision.

Conflicts of interest

The authors declare no conflict of interest.

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