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Case report

A remarkably rare case of Adventitial Cystic Disease of the Popliteal Artery in a 51-year-old Middle Eastern female - A Case Report

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ABSTRACT

Introduction and importance: Adventitial Cystic Disease (ACD) is a vastly rare non-atherosclerotic vascular pathology that is principally manifested as intermittent claudication because of peripheral vascular ischemia. Precise etiological factors are not yet concretely identified, and it represents 0.1 % of all lower limb claudication causes. Middle-aged males are the most affected gender age group. Misdiagnosis of Popliteal Artery ACD could delay proper management and hence risk the loss of the affected limb due to critical limb ischemia.
Case presentation: We hereby explore the rare case of a 51-year-old female patient, who complained of vague left lower extremity pain accompanied by paresthesia for 1 month prior to admission without signs of local inflammation. The preoperative radiological assessment suggested the presence of thrombosis within the left Popliteal Artery which caused an occlusion in it and hence the proper blood flow was compromised.
Clinical discussion: Surgical intervention and the complete removal of the lesion along with establishing a patent synthetic anastomotic graft to maintain the preexisting vascular bundle was the key to treating our patient. Microscopic analysis of the excised specimen revealed an Adventitial Cystic Disease of the Popliteal Artery.
Conclusion: Adventitial Cystic Disease represents an extremely rare vascular pathology with a vast margin of non-specific symptoms that could lead to misdiagnoses. It is fundamental to establish suitable preoperative screening protocols for it and sustain adequate levels of clinical awareness so that we can timely diagnose and in turn, achieve proper therapeutic interventions to plummet the potential disastrous complications that could ensue.

1. Introduction

Adventitial Cystic Disease (ACD) is a tremendously rare non-atherosclerotic vascular anomaly that is predominantly manifested as intermittent claudication because of peripheral vascular ischemia. Ensuing compression of the concomitant arterial lumen by the resultant collection of cystic mucous substances is the reason behind the ischemic symptoms that patients complain from. Said mucous collection is comprised of several building blocks, such as mucoproteins and mucopolysaccharides. The site in which the buildup takes place is the arterial adventitial layer. This vascular condition chiefly favors occurring in the popliteal artery with a staggering (85 % incidence rate in contrast to other vascular structures), and it classically arises in males who are between youthful and middle-age groups with a general male-to-female predominance ratio of 15:1 [1]. Our case is remarkably rare because our patient was a female and was in the 6th decade of age.

The etiological factors behind the origin of ACD remain a subject of immense controversy as no specific reasons have been proven to be the progenitors for its occurrence [1]. Regarding preoperative diagnosis of ACD, the contemporary complex non-invasive imaging modalities, such as high-resolution Computed Tomography (CT) have paved the path towards establishing a clear clinical picture to help guide physicians to suspect and perhaps confirm the presence of ACD [2,3].

Treatment of ACD has taken the form of multiple different

Abbreviations: ACD, Adventitial Cystic Disease; CT, Computed Tomography; DUS, Doppler Ultrasound; ePTFE, Expanded Polytetrafluoroethylene; H&E, Hematoxylin and Eosin; P2 Popliteal Artery, Behind-the-knee Popliteal Artery; MRI, Magnetic Resonance Imaging.

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modalities. These methods' primary goal is to reestablish the normal blood flow within the affected Popliteal Artery, and they include excisional adventitial cystotomy [3], excision of the involved arterial segment, and establishing graft anastomosis [4], or cystic contents aspiration [5].

The work has been reported in line with the SCARE criteria and the revised 2020 SCARE guidelines [6].

2. Presentation of case

2.1. Patient information

Herein, we present the interesting case of a 51-year-old Middle Eastern female, who is a known case of controlled hypertension for 4 years. She was referred to our university hospital's Vascular Surgery clinic with the chief complaint of ambiguous left lower limb pain. Its onset was sudden 1 month prior to admission, it was vaguely felt in the lower left side of the left knee, was sharp in nature, intermittent even during periods of rest, scaled 05/10 according to her pain scale, had no accompanying pain radiation, was associated with the paresthesia and the feeling of “pins and needles” in her left lower limb, was not aggravated by any specific trigger, and was partially relieved by over-the-counter pain killers. Symptoms were not associated with local inflammatory signs, such as edema, hotness, redness, or swelling. Overlying skin changes, such as hypo-/hyperpigmentation, petechia, purpura, ecchymosis, pallor, and cyanosis were also denied by the patient. No co-occurring lower limb coldness or intermittent claudication was reported. The patient's review of systems yielded no relevant findings. Furthermore, the patient did not experience any night sweats, general fatigue, overlying skin changes, body weight variations, or fever. Additionally, our patient denied any history of trauma to the area or a similar previous incidence. She also denies any exposure to irradiation or chemotherapy. Additionally, she denied any history of recent infections.

Her drug history consisted of Enalapril, Amlodipine, and Bisoprolol for control of her hypertensive status. Whereas her family history merely included hypertension.

Moreover, her allergic history involved an allergy to sulfa-derived medications. Finally, her psychosocial history involved smoking as she was a 3-pack-year smoker. No relevant surgical history was reported.

Her body mass index was 28 kg/m^2.

3. Clinical findings

Vital signs readings were normal.

Via inspection, no overlying skin alterations, such as hyper-/hypopigmentation, ecchymoses, cyanosis, pallor, or pin-point spots were seen.

Via palpation, no palpable arterial pulse was demonstrated neither above the course of the Popliteal Artery nor along the ipsilateral distal arterial axis.

Via auscultation, we could not hear any relevant thrills over the left popliteal region or along the ipsilateral arterial axis.

It's worth noting that the examination findings of the right lower limb were normal.

3.1. Laboratory investigations

An appropriate laboratory panel was analyzed, and its results were normal.

4. Diagnostic assessment

Non-invasive preoperative assessment was initiated by performing Duplex Ultrasound (DUS). It revealed a marked thrombosis within the left behind-the-knee Popliteal Artery (P2 popliteal artery) (Fig. 1). The Popliteal Artery diameter measured approximately (1.1 cm). No pulse was emitted via ultrasound and the ipsilateral arterial tree elicited a mono-dimensional signal along the entirety of its axis. No concomitant aneurysms were demonstrated, the remaining ipsilateral arterial vessels were compressible with no signs of thrombosis, and the remaining examination findings of the vascular tree of the right lower limb yielded normal results.

To obtain a better visualization of the clinical findings, a Computed Tomography Angiography scan was done. The report stated the presence of a left Popliteal Artery aneurysm where the Popliteal Artery measured (1.1 cm). Said aneurysm was seen to cause an occlusion within the left Popliteal Artery for (2.2 cm) (Fig. 2A–B). The remaining arterial tree was free of any lesions. Based on the previous radiological assessment, a

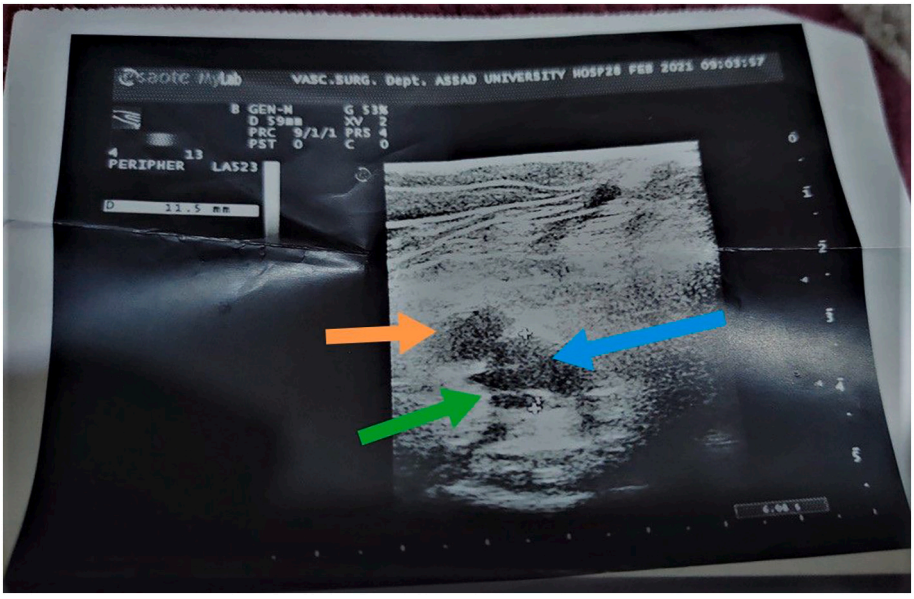

Fig. 1. Preoperative DUS depicting the popliteal fossa where the Popliteal Artery measured (1.1 cm) with notable stenosis and thrombosis within the lumen in addition to dilation of the adventitial layer. Orange arrow identifies the Popliteal Vein. Blue arrow identifies the lesion. Green arrow identifies the compressed segment of the Popliteal Artery with subsequent thrombosis within it.

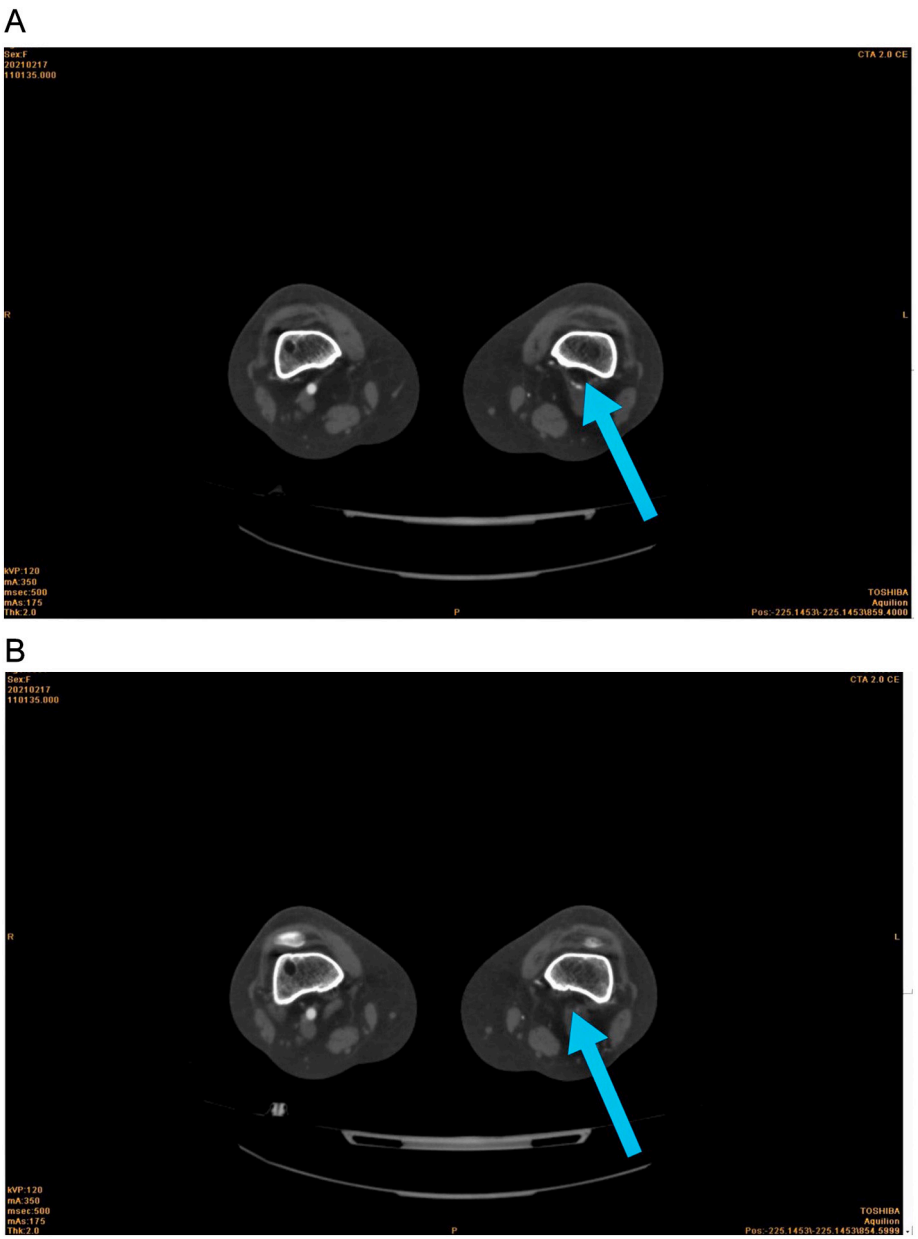

Fig. 2. (A–B): Computed Tomography Angiography scan of the lower limb arterial tree demonstrates the presence of a left lower limb's P2-Popliteal Artery thrombosis where the Popliteal Artery measured (1.1 cm).

popliteal fossa disease was largely suspected (i.e., Adventitial Cystic Disease, Baker's cyst, Popliteal Artery stenosis, peripheral arterial disease, or a soft tissue tumor).

Primary patient preoperative preparation included keeping her in a nil-per-mouth status, establishing patent intravenous access, administrating the necessary prophylactic antibiotics, and blood sampling for crossmatching.

No notable obstacles were faced during the preoperative phase.

5. Therapeutic intervention

Based on the previous clinical picture, surgical intervention was deemed necessary. The operation was successfully carried out at our specialized university hospital by a Vascular Surgery consultant with 35 years of Vascular Surgery experience. Additionally, it took place under combined spinal and epidural anesthesia without the occurrence of any perioperative complications. A classical S-shaped incision in the left popliteal fossa was performed. During intraoperative exploration, a loculated and round cystic lesion overlapping the left Popliteal Artery was revealed. It was soft, rubbery, and intact with well-defined borders. The corresponding segment of the Popliteal Artery was seen to have an abnormal dilation in its central segment. To avoid recurrence due to incomplete resection, it was excised along with the affected segment of the concomitant Popliteal Artery. Then, direct Popliteal-Popliteal anastomosis was successfully achieved via a reinforced synthetic Expanded Polytetrafluoroethylene (ePTFE) graft. The excised specimen was directly sent for careful histopathological analysis.

Histopathological analysis through Hematoxylin and Eosin (H&E) staining demonstrated how the cystic wall is comprised of compact fibrous materials which conform with the arterial adventitial layer. Furthermore, foci of hemorrhage in addition to scattered segments of inflammatory contents are vivid within the composition of the wall of the lesion. Moreover, protein-derived contents are distributed across the field with marked irregularly dilated cystic space within the adventitial layer (Fig. 3A–B–C–D–E–F–G). Our patient underwent an uneventful and healthy postoperative recovery with no notable complications. We

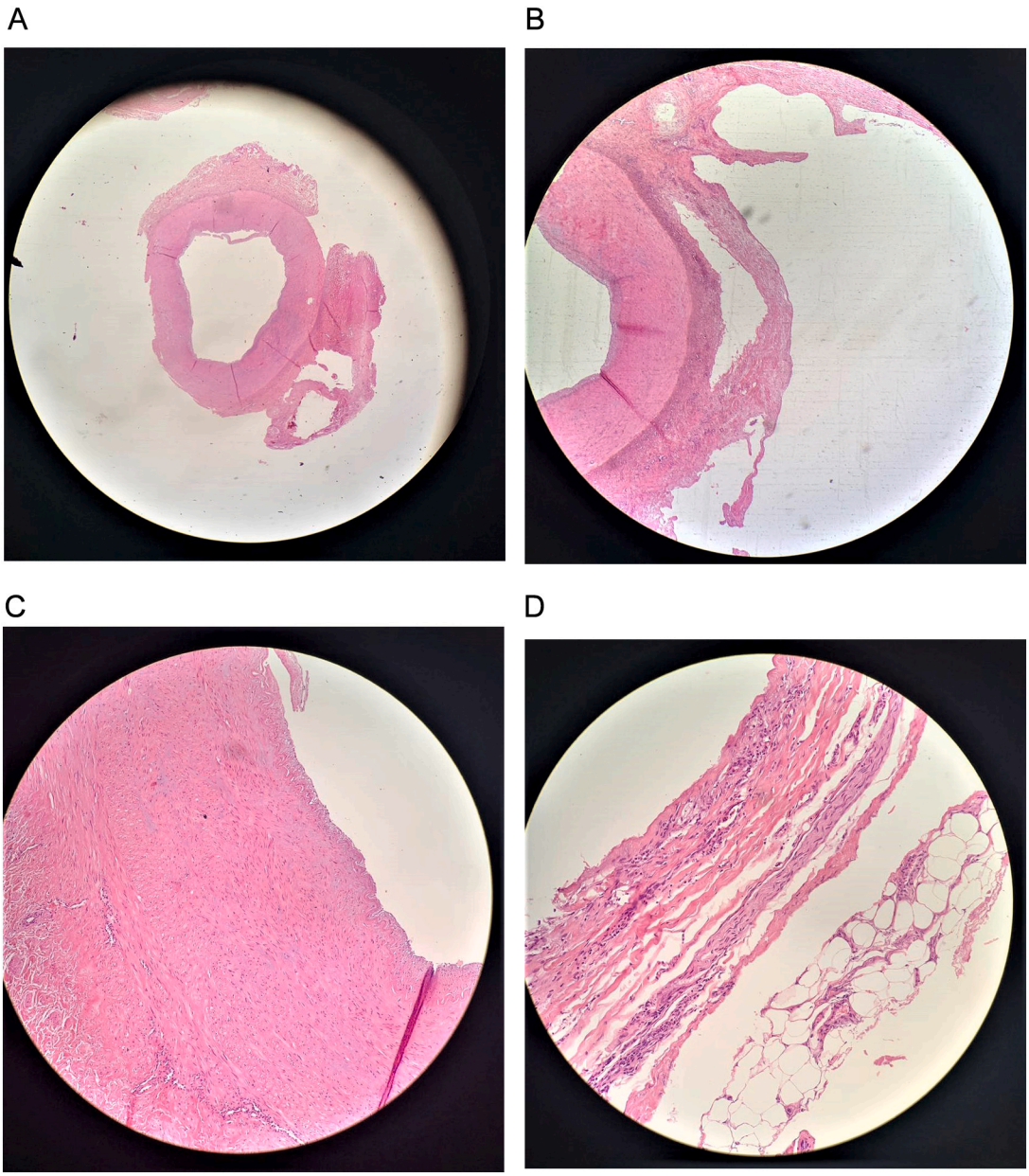

Fig. 3. (A–B–C–D–E–F–G): Histopathological analysis through H&E staining demonstrated how the cystic wall is comprised of compact fibrous materials which conforms with the arterial adventitial layer. Furthermore, foci of hemorrhage in addition to scattered segments of inflammatory contents are vivid within the composition of the wall of the lesion. Moreover, protein-derived contents with marked irregularly dilated cystic space within the adventitial layer are distributed across the field.

facilitated the performance of daily sterile wound dressings of her wound by a medical provider, prescribed her appropriate analgesics to manage any residual pain, and administered suitable postoperative antibiotics to avoid wound infections. Furthermore, she was administered medications for prophylactic anticoagulation, such as Apixaban, Aspirin, and Clopidogrel.

We followed up with our patient after her surgery for 18 months now. She was scheduled for regular appointments at our university hospital's Vascular Surgery clinic to meticulously carry out thorough bedside examinations and perform DUS imaging series. The patency of the installed graft was vividly marked, and the examination of the bilateral lower limbs' arterial tree yielded normal results (i.e., three-dimensional signals were demonstrated across the entirety of the left lower limb, no stenosis at the site of the installed graft was seen, and no aneurysms or pseudoaneurysms were seen). Now, she's assigned to annual clinical visits to undergo vascular reevaluations.

6. Discussion

Atkins and Key were the first to document an Adventitial Cystic Disease in 1947 in London [7]. In 1954, Ejrup and Hiertonn were the first to depict the first reported case of ACD affecting the Popliteal Artery [8]. From that point to this day slightly over 700 cases have been diagnosed. Of those cases, 80.5 % were involving the Popliteal Artery [9]. Adventitial Cystic Disease comprises merely 0.1 % of the cases of lower limb intermittent claudication [10]. Furthermore, most cases where the Popliteal Artery is involved are unilateral in occurrence [9]. Whereas the next most frequently involved arteries are the Femoral and

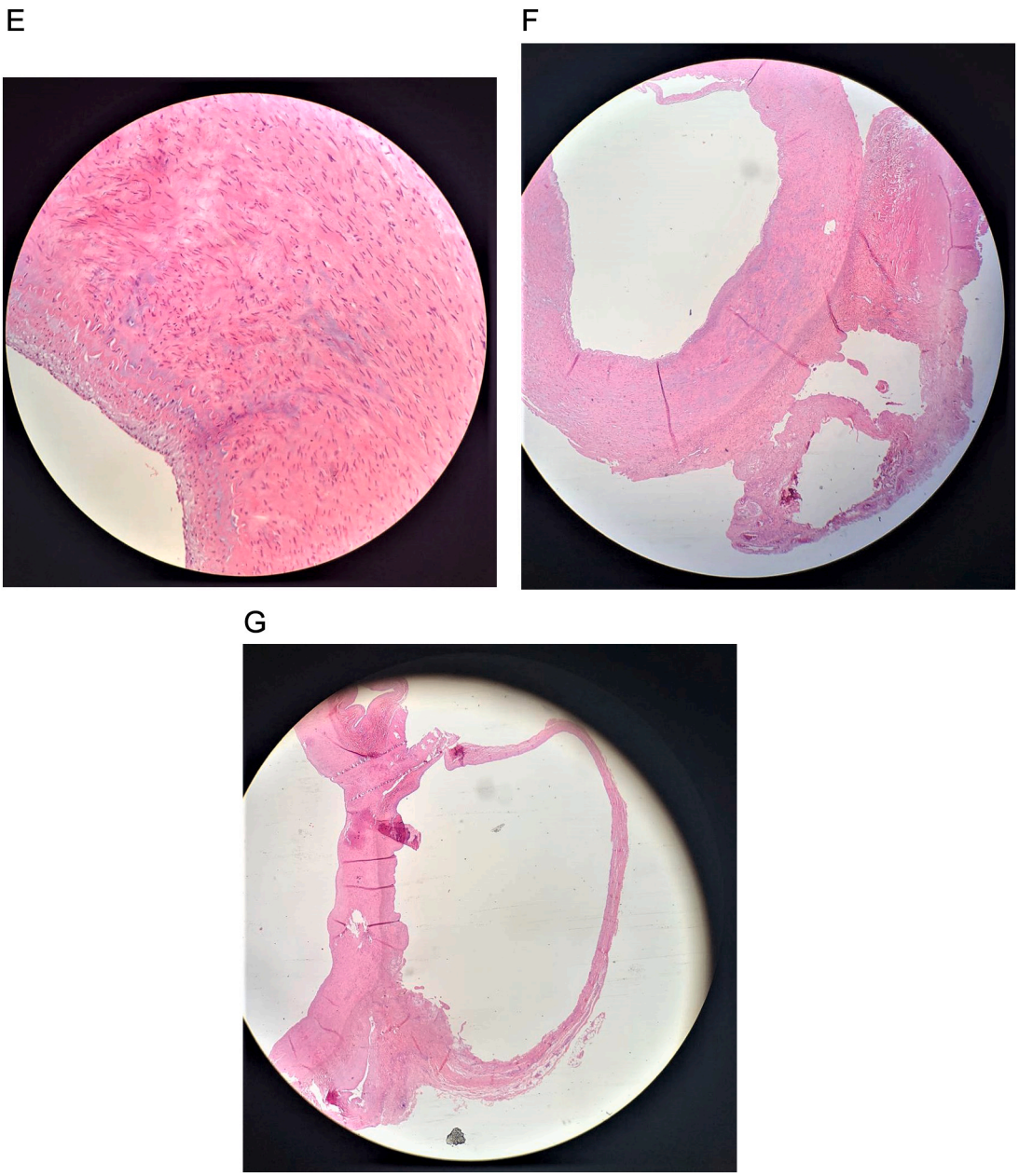

Fig. 3. (*continued*).

External Iliac Arteries [11,12]. Regarding gender-specific prevalence, ACD tends to occur more prominently in males. Moreover, the male-to-female ratio is estimated to be 15:1 where the affected individuals are most likely middle-aged, particularly in their 5th decade of life [9].

From an epidemiological standpoint, the precise prevalence of ACD among patients with vascular claudication has been estimated to be 1 in 1200 cases, regardless of their population age group [13]. The etiological factors remain vague and ill-defined, and the pathogenesis remains poorly understood. Several theories have been suggested. These include ganglion formation, repeated trauma, articular diseases, developmental anomalies, and systemic disorders [13,14]. When the illness starts to manifest itself, patients with ACD affecting the popliteal artery classically are middle-aged males who report a sudden occurrence of intermittent claudication on short distances [13]. The duration of their chief complaint is somewhat limited ranging from weeks to months and is usually happening in one limb rather than bilaterally. On the other hand, symptoms of vascular claudication might spontaneously resolve for a short timeframe, but nonetheless, they swiftly recur and progress to cause a change in patients' daily lives. Vascular claudication in patients with ACD is usually up to 20 min when compared to that of other causes for vascular claudication [15].

When ACD strikes the Popliteal Artery, it is estimated that two-thirds of those cases involve arterial stenosis rather than occlusive episodes. Moreover, upon physical examination of said patients, physicians could elicit a response to guide towards the diagnosis and this clue is called "Ishikawa Sign". It can be illustrated by a normal or rather dim pedal arterial pulse and by a vivid arterial bruit in the affected popliteal fossa. The ipsilateral pedal arterial pulses, which are normally evident during rest, might disappear upon ipsilateral hip and knee flexion [16].

When we want to establish a preoperative diagnosis, contemporary radiological modalities serve an immense role in achieving this endeavor. Current guidelines support the utilization of DUS followed by CT or Magnetic Resonance Imaging (MRI) [13]. DUS remains the gold standard to initiate the radiological assessment for ACD [17,18]. However, in cases of ACD affecting the Popliteal Artery, CT is utilized more frequently because it possesses the capability to visualize the cystic

lesions along with their anatomical relationship to their surrounding structures [19].

When deciding on the available therapeutic options. It's worth noting that there are excisional and non-excisional surgical interventions. The latter include percutaneous transluminal angioplasty - with or without the utilization of stents, ultrasound or CT-guided percutaneous aspiration of cystic contents, and evacuation of cystic contents - with or without resection of the said cyst [9,13]. These methods' primary goal is to reestablish the normal blood flow within the affected Popliteal Artery, and they include excisional adventitial cystotomy [3], excision of the involved arterial segment and establishing graft anastomosis [4], or cystic contents aspiration [5].

Interventional methods which don't involve resection of the affected segment are chosen in cases where stenoses without vessel occlusion are encountered. However, complete excision with respective vessel reconstruction is the gold standard where utter occlusion of the Popliteal Artery is present due to thrombosis or in cases where there is thrombosis in the setting of prominent arterial wall degeneration [9,13]. Furthermore, excision of the Popliteal Artery segment that is affected by an ACD renders the possibility of disease recurrence highly unlikely when compared to other available modalities of therapy. However, recurrence could possibly occur regardless of which method has been applied [20]. When it comes to patient symptoms, they tend to rise again in 10 %–30 % of individuals who underwent aspiration of cystic contents during an average post-aspiration period of 15 months [15,21]. On the other hand, cystic recurrence has been seen in 15 % of individuals who underwent cystic evacuation, whereas it only occurred of 6 %–10 % in patients who underwent complete surgical cystic removal [13,22].

7. Conclusion

Adventitial Cystic Disease is an extremely rare vascular pathology that requires immediate attention and tends to occur in the vascular tree of the popliteal fossa. Sufficient levels of clinical awareness must be present when faced with such a clinical presentation especially when the site of the complaint is the popliteal fossa. Attending to it in a timely fashion allows the medical provider to perform the necessary surgical interventions that aid in avoiding the wide variety of ensuing complications. Documentation of such rare clinical diseases is vital because this disease could potentially lead to devastating complications like critical limb ischemia and loss of the affected extremity. As a result, we could successfully conduct pre-/postoperative studies to create better screening methods, intraoperative surgical techniques, and postoperative surveillance modalities.

Abbreviations

ACD	Adventitial Cystic Disease
CT	Computed Tomography
DUS	Doppler Ultrasound
ePTFE	Expanded Polytetrafluoroethylene
H&E	Hematoxylin and Eosin
P2 popliteal artery	Behind-the-knee Popliteal Artery
MRI	Magnetic Resonance Imaging

Consent of patient

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Availability of data and materials

The datasets generated during and/or analyzed during the current study are not publicly available because the data were obtained from the hospital computer-based in-house system. Data are available from the corresponding author upon reasonable request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

Institutional review board approval is not required for deidentified single case reports or histories based on institutional policies.

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OA, OH: Conceptualization, resources, who wrote, original drafted, edited, visualized, validated, and literature reviewed the manuscript.
YA: Resources, project administration, visualization, and review of the manuscript.
MN: Vascular Surgery consultant, who performed and supervised the operation. Supervision, project administration, and review of the manuscript.
OA: The corresponding author who submitted the paper for publication.

All authors read and approved the final manuscript.

Conflict of interest

The authors declare that they have no competing interests.

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- Pathology Department at Al Assad University Hospital, Damascus, Syria.

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