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CASE REPORT

Popliteal artery aneurysmal dilatation by intimomedial mucoid degeneration causing emergency limb ischemia: a case report and literature review

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Abstract

Intimomedial mucoid degeneration (IMMD) is an extremely rare vascular condition characterized by mucin accumulation in the intima and media, resulting in arterial wall elastic tissue degradation and aneurysm formation. We report the case of a 34-year-old woman who presented to the emergency department with a 1-month history of left lower limb pain. On local physical examination, both lower limbs were warm with palpable regular pulses in the entire lower limbs and normally capillary refill. No palpable masses were felt. Computed tomography angiography (CTA) of the lower limbs revealed an aneurysm with a thrombosed sac in the left popliteal artery behind the knee joint. Thoracic and abdominal CTA revealed normal findings. The patient underwent posterior exploration of the left popliteal artery, which revealed a large aneurysm requiring excision, and an interposition vein graft end-to-end anastomosis was performed. Histopathological examination of the aneurysm disclosed the presence of fibrosis, scattered chronic inflammatory cells, and a hyalinized fibrin thrombus associated with marked fragmentation and distortion of the elastic tissue. Extensive mucoid deposition in the intima and media of the artery was also detected. The postoperative recovery was uneventful, and the patient was discharged home with regular follow-up during the last year in our outpatient clinic. IMMD can affect smaller vessels such as the brachial, coronary, temporal arteries, and dorsalis pedis. The main histological hallmarks of IMMD include intimal and medial thickening caused by the formation of mucin pools, which leads to elastin fibers fragmentation and aggregation. The prognosis of IMMD-related aneurysms is determined by the extent of the disease and the time it is discovered. IMMD-related aneurysms should be included in the differential diagnosis of isolated popliteal artery aneurysms. Open surgical repair is the gold standard of treatment for IMMD-related aneurysms.

Keywords

Intimomedial mucoid degeneration; Emergency limb ischemia; Arterial disease; Popliteal artery; Aneurysm; Open repair

1. Introduction

Intimomedial mucoid degeneration (IMMD) is an uncommon vascular condition characterized by mucin accumulation in the intima and media, resulting in arterial wall elastic tissue degradation and aneurysm formation [1–4]. IMMD was documented to impact the large branches of the aorta, as well as medium and small vessels such as the coronary and brachial arteries, in prior studies [1, 5–8]. The cause of IMMD remains unknown. The illness was initially reported by Pepler's study in South Africa [9]. Decker *et al.* [1] coined the term "intimomedial mucoid degeneration" in 1977 in a case series of nine patients with aortic aneurysms. The disease was defined in pathological terms by characterizing its histological properties because its underlying pathology was unclear [1, 10, 11]. In IMMD, aneurysms usually have a saccular or fusiform morphology, and they induce location-specific symptoms [6, 8]. A bleeding diathesis differing from disseminated intravascular coagulation frequently complicates surgery, but it resolves with surgical treatment of the affected vessel [2]. A meticulous technique during surgery is extremely important [2, 6]. The coagulation profile and platelet function should be closely monitored and adjusted perioperatively. Unfortunately, because of the rarity of this pathology, it remains poorly studied, and no guideline regarding its management exists. In this study, we report a patient with a painful popliteal artery aneurysm caused by IMMD that was treated surgically. According to the English literature, this is the first case report

of IMMD affecting the popliteal artery. In addition, we present a literature review of this rare vasculopathology.

2. Case presentation

A 34-year-old woman receiving medical treatment for asthma patient presented to the emergency department complaining of a 1-month history of left lower limb pain. The pain was intermittent, sudden in onset, and associated with coldness and numbness in the same limb. The pain was aggravated by walking and prolonged standing and relieved by relaxing. The patient denied any history of abdominal pain or other lower limb pain. The past surgical history included adenoidectomy 2 years prior to presentation, laparoscopic cholecystectomy 18 months prior to presentation, dilation and curettage for abortion 6 months prior to presentation, and laparoscopic sleeve gastrectomy 3 months prior to presentation. Her family history was unremarkable. Upon general physical examination, the patient expressed that the pain was mild. She was vitally stable and afebrile. Both lower limbs were warm with palpable regular pulses in the entire lower limbs and normal capillary refill. No palpable masses were felt. Laboratory examination revealed the following findings: hemoglobin, 14.1 g/dL; leucocyte count, 7.56 $\times$ 10^9/L; platelet count, 256 $\times$ 10^9/L, and D-Dimer, 1.87 μ/mL; partial thromboplastin time, 27 seconds; and international normalized ratio, 1.1. Negative results were obtained for antineutrophil cytoplasmic antibody, antinuclear antibody, rheumatoid factor, cryoglobulin, a hepatitis panel, a cytomegalovirus test, a thrombophilia screen, a human immunodeficiency virus test, and syphilis serology. Homocysteine and C-reactive protein levels, liver and renal function indices, and the erythrocyte sedimentation rate were all within normal ranges. Computed tomography angiography (CTA) of the lower limbs revealed an aneurysm with a thrombosed sac in the left popliteal artery behind the knee joint, and distally, all runoff vessels exhibited good opacification (Fig. 1A,B). Thoracic and abdomen showed normal findings. The decision was made to perform surgery. As the aneurysm was large and confined to the popliteal space, the decision was made to use the posterior approach with interposition vein graft. Then, the aneurysm sac was opened, and a thrombus was found and removed. Back-bleeding geniculate collaterals were oversewn, and interposition vein graft end-to-end anastomosis into the transected popliteal artery was performed (Fig. 2A,B). Histopathological examination of the aneurysm revealed the presence of fibrosis, scattered chronic inflammatory cells, and a hyalinized fibrin thrombus associated with marked fragmentation and distortion of the elastic fibers (Fig. 3A–C). The examination also disclosed extensive mucoid deposition within the intima and media of the artery (Fig. 4A–C). The postoperative recovery was uneventful. The patient was discharged home in a good condition with regular follow-up during the last year after operation in our outpatient clinic.

3. Discussion

In 1993, Cooper *et al.* [11] presented a series of six patients with IMMD detected in the subclavian, common carotid, mesenteric, and iliac arteries, revealing that IMMD can also have an extra-aortic presentation. IMMD was documented to impact the large branches of the aorta, as well as medium and small vessels such the brachial, coronary, and temporal arteries and dorsalis pedis [5, 7]. In our case, only the left popliteal was involved.

Several studies demonstrated that IMMD-associated aneurysms affect a younger population than affected by conventional nonspecific degenerative aneurysms [1, 4, 6], in line with our patient age. Patients with IMMD have symptoms that are specific to the location of the aneurysm [7, 9]. The clinical signs include abdominal and back pain, the presence of a pulsatile mass, limb claudication, and aneurysm rupture symptoms [6]. These clinical signs could be explained by the most common locations of involvement such as the thoracic aorta; infra-renal aorta; and common carotid, subclavian, and common iliac arteries [6]. However, our patient had left lower limb claudication, which could be explained by the local effects of the aneurysmal left popliteal artery. Aneurysms in patients with IMMD frequently have fusiform or saccular morphological characteristics [8]. To establish the extent of the disease and dismiss the presence of a dissection in the affected artery, imaging modalities such as duplex ultrasonography, CTA, and/or magnetic resonance angiography are frequently used [6, 8]. In our patient, chest and abdominal CTA showed normal pathology.

The main histological hallmarks of IMMD include intimal and medial thickening caused by the formation of mucin pools, which leads to elastin fiber fragmentation and aggregation, as observed in our case [8, 11]. Aneurysms arise as a result of weakness of the artery wall structure [2, 11]. The lack of inflammatory reaction on histological inspection is a noteworthy feature [1]. The characteristics of IMMD differ from those of cystic medial necrosis in which the mucin accumulation affects only the media [1, 4]. IMMD has been reported to affect extra-aortic arteries, whereas cystic medial necrosis is usually limited to the aorta [1, 11]. In IMMD, extra-aortic disease can occur without aortic involvement [2, 11]. The lack of a luminal thrombus in the aneurysm sac is a distinguishing hallmark of IMMD [7], which contradicted the findings in our patient, in whom a large thrombus was found on imaging and during surgery. During surgery, patients frequently experience bleeding [8]. Surgical manipulation aggravates this bleeding diathesis, which is reversed once the aneurysm is repaired. It is thus hypothesized that the primary fibrinolytic process starts from the diseased aneurysm, which could explain why thrombus formation and occlusive illness are uncommon in IMMD [4–6, 8], which also contradicts the findings in our patient. The gold standard for treating IMMD-related aneurysms is open surgical repair [6, 8]. In our patient, left popliteal artery aneurysm open repair with interposition vein grafting was performed. Endovascular repair has gained popularity in the treatment of popliteal artery aneurysms as an alternative to the open surgical approach. It is associated with acceptable long-term patency rates, but stent fractures occurred in almost one-third of cases [12]. The prognosis of IMMD-related aneurysms is determined by the extent of the disease and the timing of discovery [8]. The morbidity and mortality rates are higher in patients with acute presentations and dissection [8]. Major blood loss, which necessitates large blood transfusions,

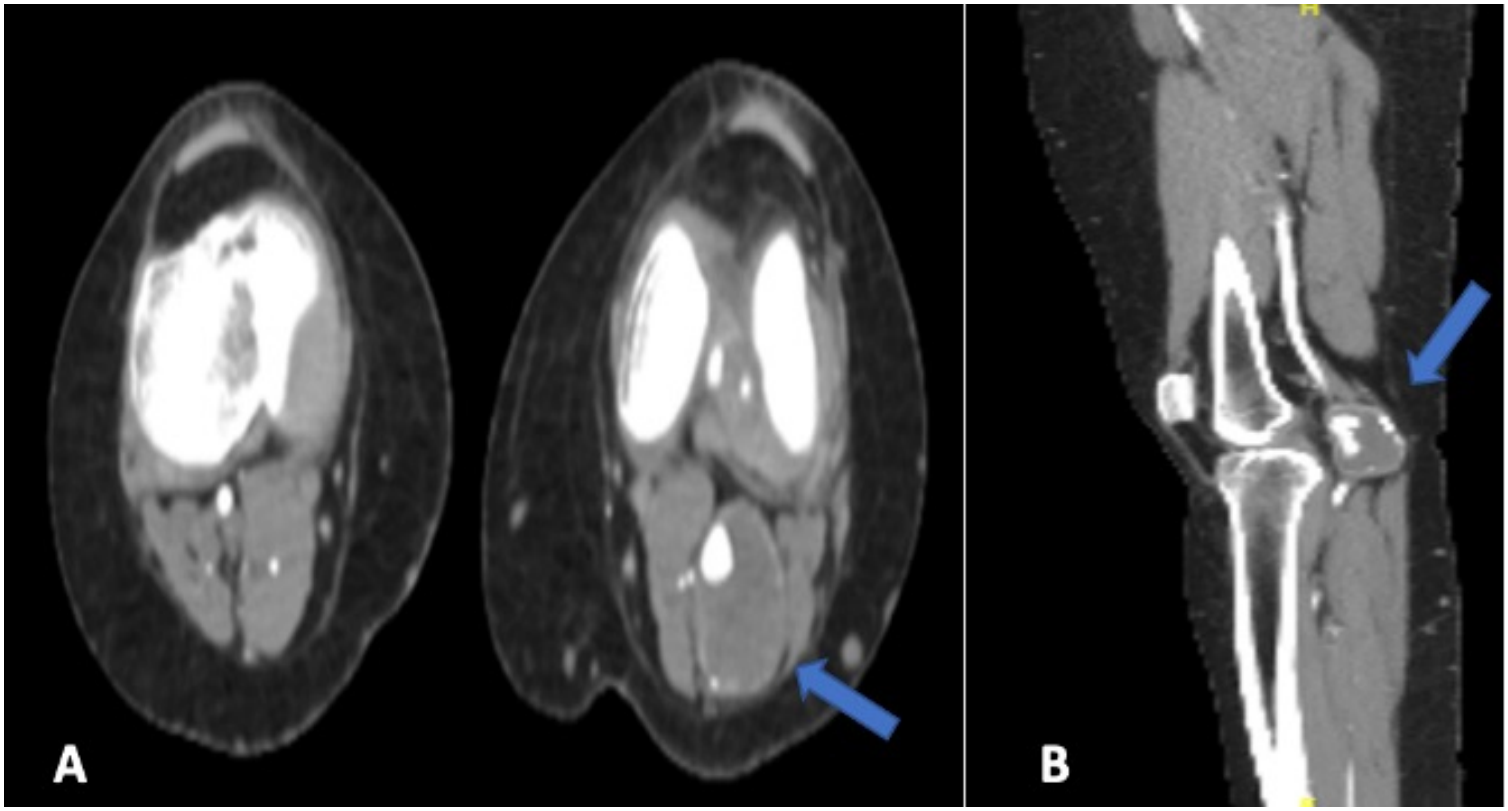

FIGURE 1. Computed tomography angiography of the lower limbs. (A) CTA axial section revealed a left popliteal artery aneurysm with a thrombosed sac behind the knee joint (arrowhead), and distally, all runoff exhibited showed good opacification. (B) CTA sagittal section of the same image.

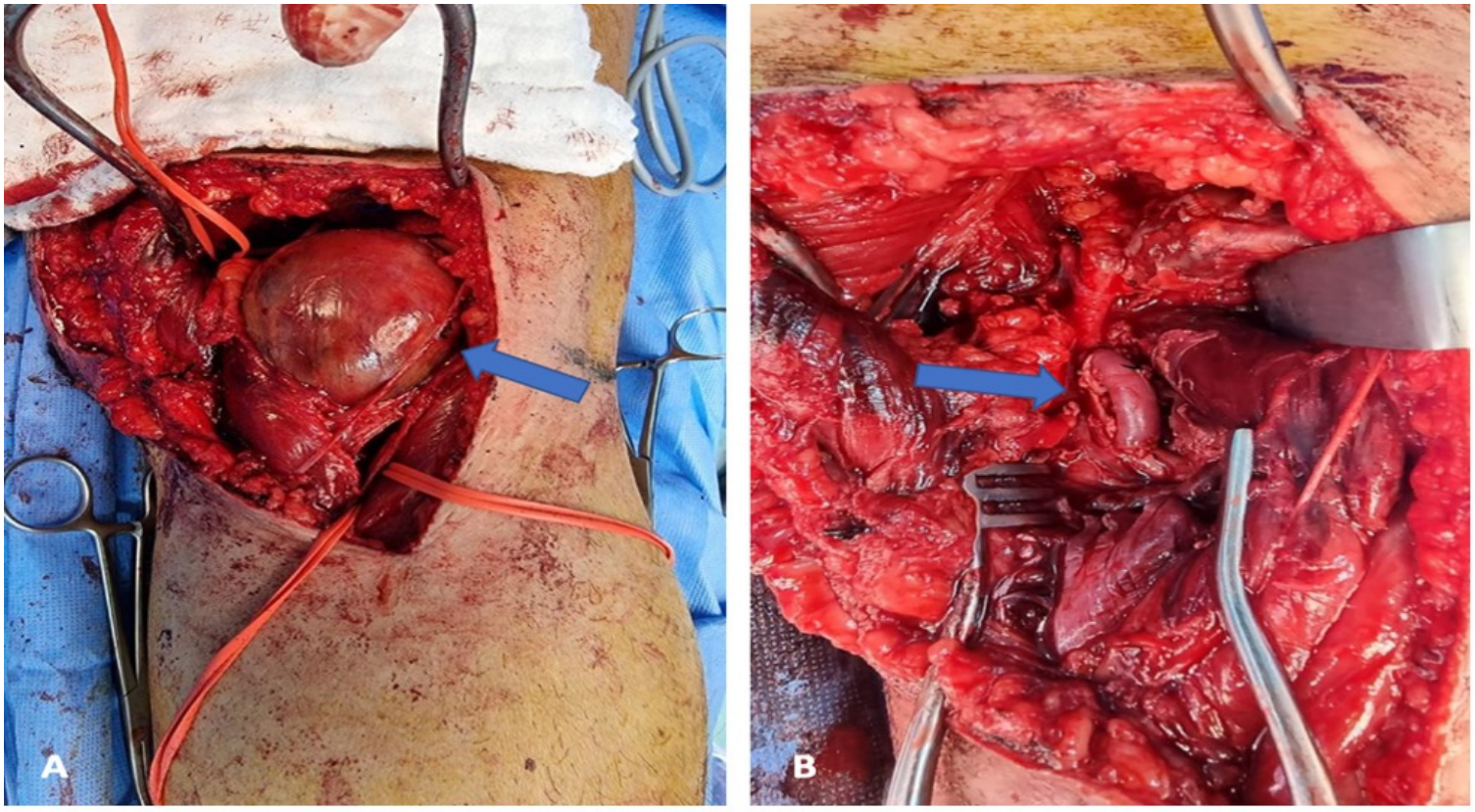

FIGURE 2. Intraoperative findings. (A) Posterior exploration of the left popliteal artery, which revealed a large aneurysm confined to the popliteal space (arrowhead). (B) Interposition vein graft end-to-end anastomosis into the transected popliteal artery was performed (arrowhead).

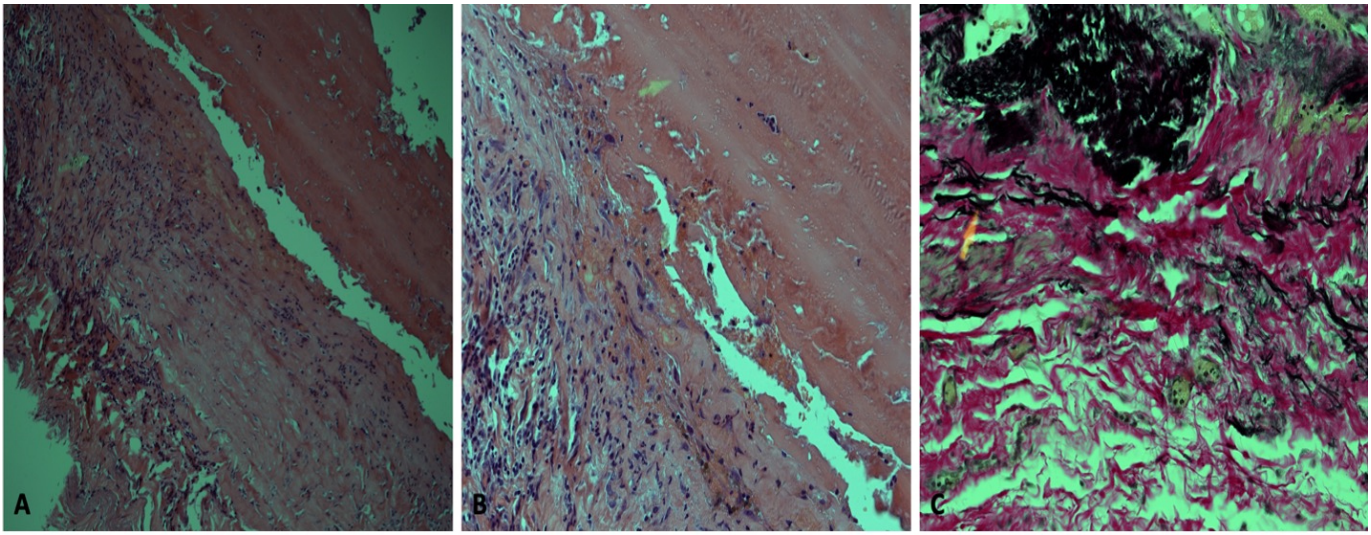

FIGURE 3. Histopathological examination. (A) Hematoxylin and eosin staining (×100 magnification) and low-power microscopic view revealing part of the aneurysm wall. Note the presence of fibrosis, scattered chronic inflammatory cells, and a hyalinized fibrin thrombus in the upper right side of this figure. (B) Hematoxylin and eosin staining (×200 magnification) and medium-power microscopic view revealing part of the aneurysm wall. Note the presence of fibrosis, scattered chronic inflammatory cells, and a hyalinized fibrin thrombus (arrowhead). (C) Elastica van Gieson staining (×200 magnification) and photomicrograph representing a section obtained from the aneurysm wall. The stain highlighted the marked fragmentation and distortion of the elastic fibers (arrowhead) with the aneurysm wall.

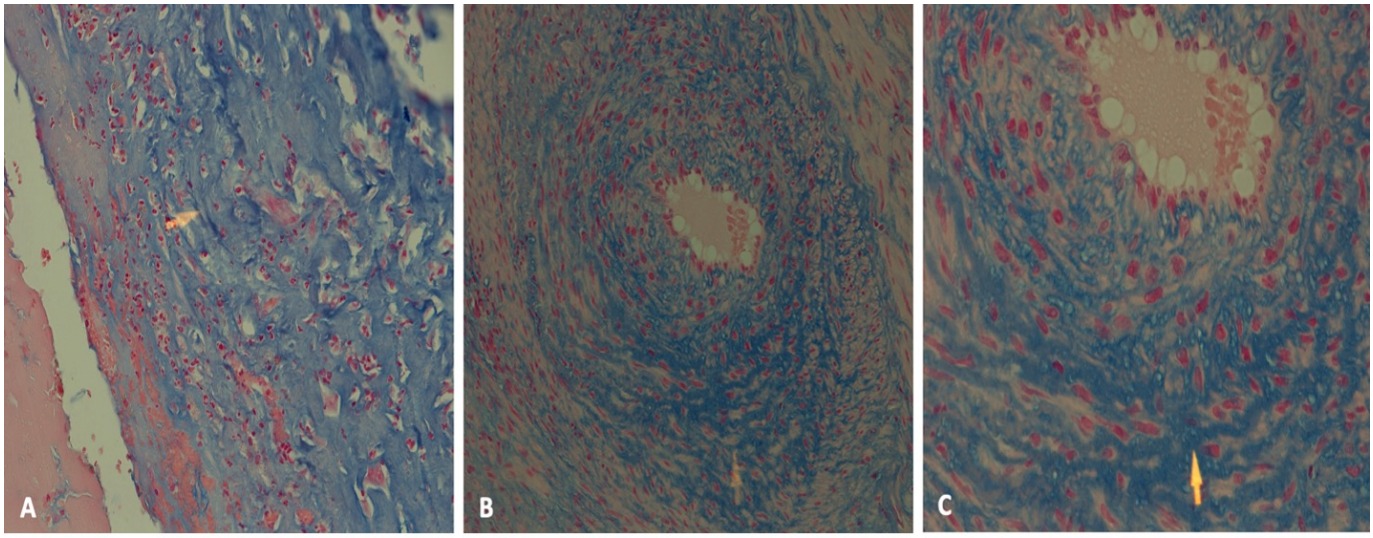

FIGURE 4. Histopathological examination using Alcian blue staining. (A) Photomicrograph of Alcian blue staining (×200 magnification) revealing a section from the aneurysm wall. Note the presence of marked mucoid material deposition in the aneurysm wall (arrowhead). (B) Photomicrograph of Alcian blue staining (×200 magnification) disclosing a medium-sized arteriole near the aneurysm. Note the presence of extensive mucoid deposition with the intima and media of the artery (arrowhead). (C) Photomicrograph of Alcian blue staining (×400 magnification) revealing a medium-sized arteriole near the aneurysm. Note the presence of extensive mucoid deposition with the intima and media of the artery (arrowhead).

and multi-organ failure attributable to shock result in poor outcomes [6].

Even though IMMD is a rare vascular condition, it should be included in the differential diagnosis of isolated popliteal artery aneurysms, particularly in younger patients and in the absence of traditional risk factors associated with nonspecific degenerative aneurysms. Open surgical repair remains the gold standard of treatment for IMMD-related aneurysms.

4. Patient perspective

Written informed consent was obtained from the patient for the 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.

ABBREVIATIONS

IMMD, Intimomedial mucoid degeneration; CTA, Computed tomography angiography.

AVAILABILITY OF DATA AND MATERIALS

The data presented in this study are available on reasonable request from the corresponding author.

AUTHOR CONTRIBUTIONS

KI—Substantial contributions to the conception, design of the work; the acquisition, analysis, interpretation of data for the work; drafting the work and revising it critically for important intellectual content; final approval of the version to be published. MYA—Substantial contributions to the design of the work; or the acquisition, analysis, or interpretation of data for the work; drafting the work or revising it critically for important intellectual content; final approval of the version to be published. SA—Substantial contributions to the design of the work; or the acquisition, analysis, or interpretation of data for the work; drafting the work or revising it critically for important intellectual content; final approval of the version to be published. MAlsa—Histological analysis of the surgical specimen, curation of the anatomopathological part of the paper; final approval of the version to be published. MAlmo—Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; final approval of the version to be published. AA—Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; drafting the work or revising it critically for important intellectual content; final approval of the version to be published.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Our work does not infringe on any rights of others, including privacy rights, and intellectual property rights. There is no human rights violation in our manuscript. The patient provided written informed consent for the publication of this case report. Our institution (College of Medicine, King Saud Medical City, King Saud University) provided an exemption for consideration since it was a case report article with permission from the patient.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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