Cross-Sectional Imaging in a Case of Adventitial Cystic Disease of the Popliteal Artery

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Abstract

Adventitial cystic disease of the popliteal artery is an unusual condition of uncertain etiology, in which a mucin-containing cyst forms in the wall of the artery and produces lower extremity claudication, typically in young and middle-aged men. A diagnosis of adventitial cystic disease of the popliteal artery was made preoperatively in a 47-year-old man by means of several imaging modalities, including angiography, magnetic resonance imaging, and ultrasound. The pathological findings confirmed the suggested diagnosis.

Key words: Adventitial cystic disease—Arteries, popliteal—Angiography—MRI—Ultrasound

Adventitial cystic disease of the popliteal artery is an uncommon condition producing lower extremity claudication, usually in young and middle-aged men [1, 2]. A cystic collection of mucinous material within the adventitia compresses the popliteal artery lumen, typically at the level of the femoral condyles. Calf claudication is often severe, but prognosis has been excellent after surgical evacuation of the cyst or bypass grafting [3, 4]. Spontaneous rupture is extremely rare, with only three reported cases in the literature to our knowledge [5].

We present here a patient with clinical findings of high-grade popliteal artery stenosis produced by two adjoining adventitial cysts, clearly depicted by color Doppler imaging (CDI) and magnetic resonance imaging (MRI), after doubtful angiographic findings.

Case Report

A 47-year-old man presented with progressive onset of severe right calf claudication after walking 100 m; the left side was asymptomatic. Ankle pulses were clearly reduced on the right side. The right ankle–brachial index (ABI) was 0.67 at rest and dropped to 0.48 after treadmill exercise. The patient was a heavy smoker (more than 20 cigarettes a day) and was an alcoholic.

Arteriography showed a high-grade focal stenosis (90%) of the popliteal artery at the level of the femoral condyles, with a smooth tapered appearance in lateral view (Fig. 1). There was no deviation of the popliteal artery in flexion and extension views, suggesting a muscular entrapment syndrome. There was no angiographic evidence of atherosclerosis. The contralateral popliteal artery appeared normal. The angiographic appearance was highly suggestive of extravascular compression of the popliteal artery (Baker's cyst).

MR images, obtained 2 days later, did not demonstrate any extravascular compression. The MRI examination was performed with a low-field (0.2-T) dedicated magnet (Artoscan, Esaote, Genoa, Italy), using T1-weighted (TR 680, TE 24, NEX 3, 13 slices, 2-mm thickness) and T2-weighted sequences (TR 2080, TE 24, NEX 1, 13 slices, 2-mm thickness) in axial, coronal, and sagittal planes. The popliteal artery presented a segmental dilation, about 7 cm in length, with a thin septum, characterized by high signal intensity in spin echo T1-weighted images, outlining lacunar spaces of intermediate signal intensity (Fig. 2A). This appearance suggested a dissected popliteal aneurysm, assuming the lacunar spaces to be due to a false lumen. The presence of an intravascular well-delimited hyperintense mass on T2-weighted images suggested adventitial cystic disease (Fig. 2B).

Ultrasound (US) performed immediately after MRI (7.5–10 MHz linear probe; AU4 Idea ACT, Esaote) confirmed the absence of vascular compression, and showed an enlarged popliteal artery with evidence of two intravascular adjoining anechoic masses, about 10 mm in diameter, with smooth borders. The upper mass was localized on the anterior wall, while the lower was on the posterior wall, thus defining a thin residual lumen, clearly depicted by CDI (Fig. 3). The duplex Doppler evaluation showed a high-grade stenosis (90%) at this level. The vessel was normal above and below this focal lesion.

The patient underwent surgery a week later. After harvesting a segment of the greater saphenous vein, the patient was placed in a prone position. The popliteal artery was exposed from a posterior approach through a linear incision. A 10-cm length of the artery was grossly enlarged, with cystic lesions in the subadventitial plane. The abnormal segment of the popliteal artery was excised, and a reversed vein graft was interposed.

The surgical specimen, consisting of a 6.3-cm segment of popliteal artery, was fixed in 4% formaldehyde and routinely processed for paraffin embedding. Sections were stained with hematoxylin–eosin, van Gieson–Weigert and alcian blue. An immunohistochemical study was carried out with vimentin, epithelial membrane antigen (EMA), and cytokeratins. All the immunohistochemical analyses were carried out with the peroxidase–antiperoxidase method, and diaminobenzidine tetrahydrochloride was used as chromogen.

Pathological examination of the surgical specimen revealed grossly multiloculated intramural cysts, filled with gelatinous material, narrowing the arterial lumen. The cystic spaces were histologically located both between the media and the adventitia and in the intima. The cysts partially lacked an epithelial lining and were partially lined by a layer of cylindrical cells, which were positive for vimentin. The material inside the cystic space

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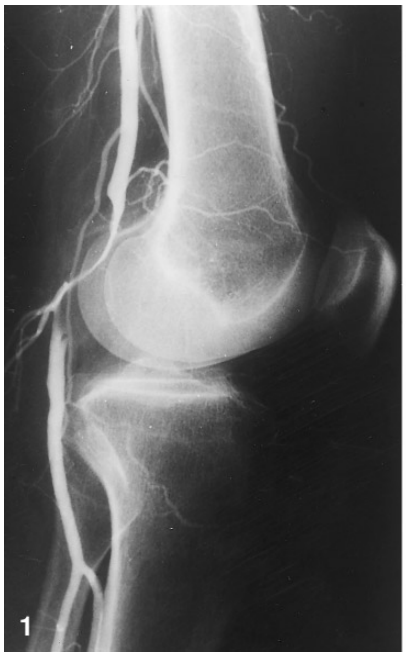

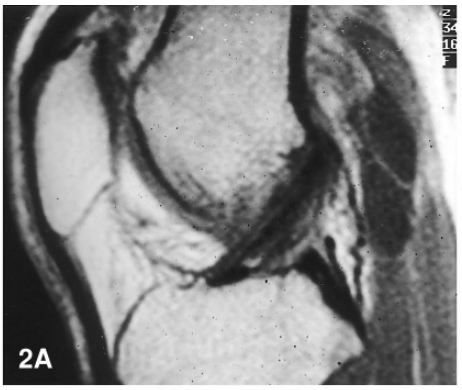

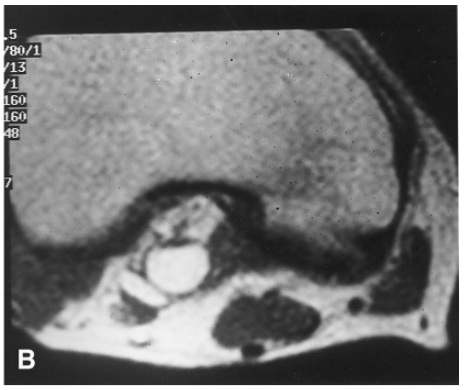

Fig. 1. The conventional arteriogram shows a high-grade focal stenosis (90%) of the popliteal artery at the level of the femoral condyles, with a smooth tapered appearance in lateral view.

Fig. 2. On MRI the popliteal artery presented a segmental dilation, about 7 cm long, with a thin septum **(A)**, characterized by high signal intensity in spin echo T1-weighted images (TR 680, TE 24, NEX 3), enclosing lacunar spaces of intermediate signal intensity. The cyst-like mass was hyperintense **(B)** on T2-weighted images (TR 2110, TE 80, NEX 1).

was positive for alcian blue, thus showing its acid mucopolysaccharide nature. The pathological diagnosis was adventitial cystic disease of the popliteal artery (Fig. 4).

Discussion

Most patients with intermittent claudication of the calf have atherosclerotic disease, usually with a typical clinical history. The most common non-atherosclerotic causes of popliteal artery disease are embolism, thrombosed aneurysm, entrapment syndrome, ruptured Baker cyst, and adventitial cystic disease (ACD) [6]. In clinical practice ACD is rarely found, with an estimated prevalence of 1 : 1200 cases of claudication [2]. The first case involving the external iliac artery was described by Atkins and Key in 1947 [7], while involvement of the popliteal artery was described by Ejrup and Hiertonn in 1954 [1].

Possible causes include trauma to the adventitia due to repetitive flexion injuries, myxoid degeneration, and inclusion of remnants of mucus-secreting synovial cells in the adventitia. Popliteal cysts are always unilateral and are usually small and unpalpable. They primarily involve the adventitia, but may eventually erode into the media or produce popliteal artery thrombosis.

There are at least two plausible etiopathogenetic theories for ACD. These adventitial cysts often contain crystal-clear fluid, similar in chemical content to the fluid in ganglion cysts, the main contents being mucoproteins or mucopolysaccharides. The presence of mucin-secreting cells may theoretically result from either aberrant developmental inclusion within the adventitia, or the tracking of capsular synovial cysts from a neighboring joint along a genicular artery, involving the adventitia. The first theory, proposed by Lewis et al. in 1967 [8], was named the ''developmental inclusion theory''. As far as the second theoretical approach is concerned, Chevrier [9] suggested that adventitial cysts are true ganglia of the vessels, and Powis et al. [10], in their ''traumatic implantation theory'', suggested a relation with acquired trauma. Both proposals are supported by surgical evidence of communication between these cystic lesions and the joint capsule of the knee [2].

Typically ACD causes relatively sudden onset of claudication without a threatened limb. Most patients are men, commonly in their fourth and fifth decades, and they often have claudication after walking very short distances. Waxing and waning of symptoms is the rule. Stenosis rather than occlusion is the usual mode of presentation.

Practically all reported cases have been surgically treated. Cyst excision is associated with an excellent prognosis. Bypass grafting is necessary when secondary intimal ulceration or thrombotic occlusion has occurred, or in the case of large lesions. An overall success rate of 91% has been reported for the combination of total excision of the involved vessel and bypass grafting [3, 4]. CT-directed percutaneous aspiration has been performed, but has been associated with recurrences [11]. Spontaneous rupture of adventitial cyst is distinctly unusual, with only three cases reported in the literature [5].

The characteristic arteriographic finding is a focal, smoothly tapered stenosis of the popliteal artery at the level of the femoral condyles. In 1961 Ishikawa et al. [12] described an hourglass appearance of ACD, due to concentric compression, and Harris and Jepson [13] a scimitar deformation due to eccentric compression. Both signs can be noted when the artery is stenotic, but they are obviously absent if it is occluded. Typically, significant disease is absent above and below the involved segment. Even in the absence of classic angiographic findings related to the popliteal artery, stenosis or occlusion of this vessel without significant disease elsewhere should prompt suspicion of ACD. Differentiation from popliteal artery entrapment is important. A decrease in pedal pulses is noted in entrapment with active plantar flexion. Angiographically, medial deviation of the popliteal artery may be present in entrapment, and narrowing or occlusion of the artery may be obtained during active plantar flexion. In ACD otherwise there is loss of ankle pulses on sharp flexion of the knee.

In our patient the diagnosis was initially missed because the scimitar sign, valuable only in the lateral view, was interpreted as extrinsic compression due to, for example, a Baker's cyst.

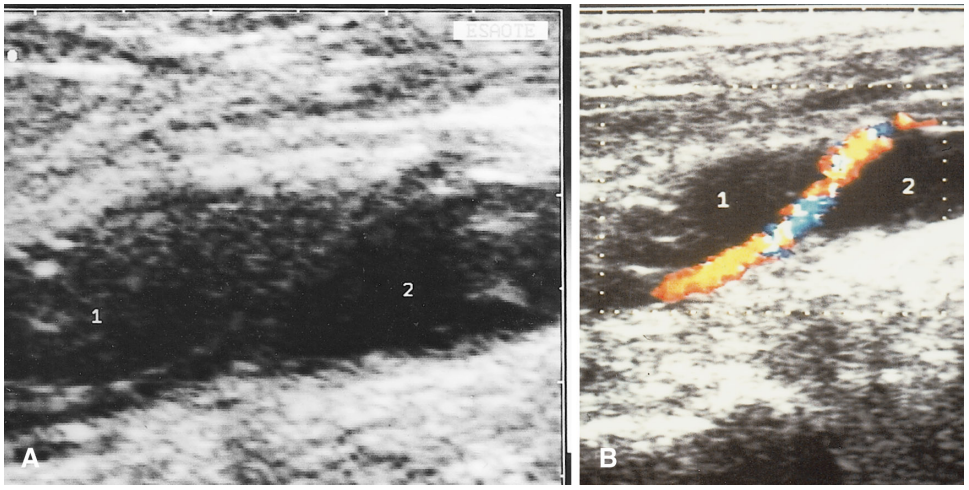

Fig. 3. US illustrates an enlarged popliteal artery, with evidence of two intravascular adjoining anechoic masses with smooth borders **(A)**, defining a thin residual lumen, clearly depicted by color Doppler imaging **(B)**.

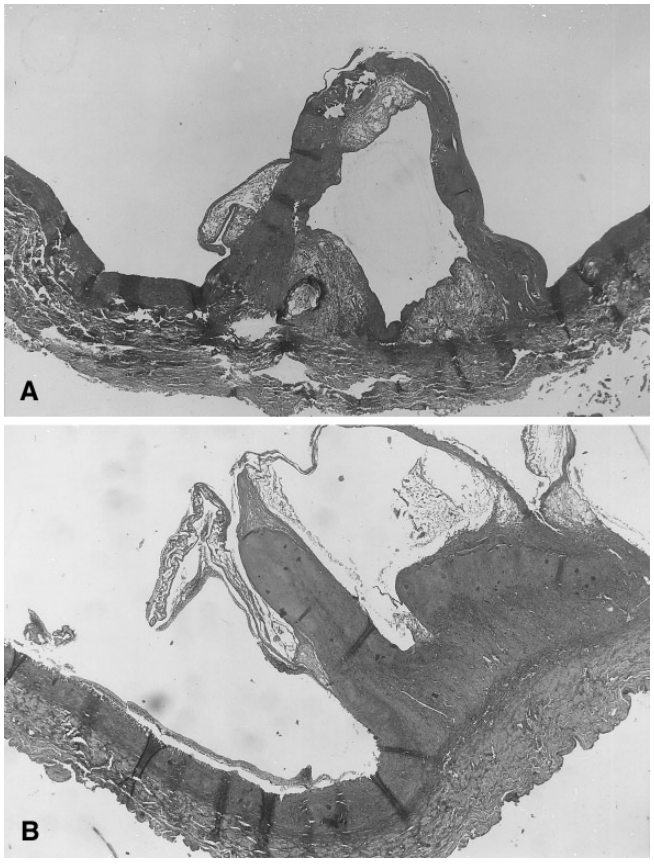

Fig. 4. Gross pathology. **A** Cross-section of the popliteal artery. An intramural cyst between the media and the adventitia protrudes in the lumen (hematoxylin–eosin, ×4). **B** Cross-section of the popliteal artery. The cystic spaces, filled with a mucous substance, are located in the intima above the internal elastic lamina (hematoxylin–eosin, ×4).

CT may assist in the diagnosis of ACD as well as in percutaneous aspiration[3, 11, 14]. On contrast-enhanced CT scans ACD may be seen as a non-enhancing cyst-like mass causing extrinsic compression of an enhancing, crescentic arterial lumen.

There are two reports in the literature on the role of MRI in the evaluation of ACD and its complications. These suggest that MRI may facilitate diagnosis of ACD by revealing cyst-like structures closely invested in a compressed artery, based on the differential signal intensity of cyst contents and flowing blood. MR angiography of the peripheral vascular system could be a valuable noninvasive means of studying the caliber of the popliteal artery. The MR angiographic appearance of the tibiopopliteal vessels has been found to be closely related to that on conventional angiography [15, 16]. At present our scanner has no hardware for MR angiography.

The contribution of CDI to the preoperative diagnosis was fundamental in our case. Although the sonographic appearance of an excised popliteal adventitial cyst has been described by Bunker et al. [17], we are not aware of any previous paper on the role of US and color Doppler. US allows the identification of the intravascular masses as cyst-like masses, with low-level echoes and a thin wall, protruding into the lumen, and the exclusion of any external compression. In our experience the main role of CDI is in the clear depiction of the residual lumen, thus allowing the differentiation among intrinsic or extrinsic compression, aneurysm, or dissection.

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Congenital Absence of the Internal Carotid Artery

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Abstract

We report three cases of congenital absence of an internal carotid artery (ICA), diagnosed incidentally by digital subtraction angiography. The analysis of the cases is based on the classification of segmental ICA agenesis proposed by Lasjaunias and Berenstein. Usually the patients with this rare vascular anomaly are asymptomatic; some may have symptoms related to cerebrovascular insufficiency, compression by enlarged intracranial collateral vessels, or complications associated with cerebral aneurysms. Diagnosis of congenital absence of ICA is made by skull base computed tomography (CT) scan, CT and magnetic resonance angiography, and conventional or digital subtraction angiography.

Key words: Agenesis—Internal carotid artery—Cerebrovascular insufficiency—Intracranial aneurysm

Congenital absence of the internal carotid artery (ICA) is a very rare vascular anomaly. Agenesis, aplasia, or hypoplasia are the three conditions responsible for an absent ICA and reflect different developmental failures. This anomaly may be completely asymptomatic and detected occasionally by skull base computed tomography (CT) scan, magnetic resonance imaging (MRI), angiography, or at autopsy or surgical procedure. The symptoms are related to cerebrovascular insufficiency, compression by enlarged intracranial vessels, and associated cerebral aneurysms. Diagnosis is important to explain the clinical and surgical problems related to this condition.

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We report three cases of congenital absence of the ICA. Patients 1 and 2 showed absence of one ICA with ipsilateral cerebral circulation supplied by the anterior communicating artery and/or posterior communicating artery. Patient 3 is an example of hypoplasia of the cervical portion of the ICA with the precavernous portion formed by a rete mirabile of vessels through the skull base. We discuss the clinical symptoms, diagnostic methods, and clinical and surgical implications of this condition.

Case Reports

Case 1

A 62-year-old woman was admitted to our hospital for acute onset of severe occipital-nuchal headache and loss of consciousness. The neurological examination revealed nuchal rigidity without focal neurological deficits. CT scan showed subarachnoid hemorrhage. Emergent cerebral angiography demonstrated an anterior communicating artery aneurysm and absence of the left ICA. The omolateral anterior cerebral artery was supplied through the anterior communicating artery; the middle cerebral artery was supplied through a hypertrophic posterior communicating artery arising from the contralateral posterior cerebral artery (Fig. 1). The patient underwent craniotomy and clipping of the aneurysm. The postoperative period was complicated by bilateral middle cerebral artery vasospasm; post-hemorrhagic hydrocephalus was treated with ventriculoperitoneal shunting. The patient has no neurological deficits.

Case 2

A 28-year old woman, treated surgically in 1988 and 1992 for an anaplastic astrocytoma in the right parietal region, was admitted to our hospital for a recurrence of the neoplasm. The neurological examination showed a left hemiparesis. MRI of the brain revealed a recurrence of the tumor with contralateral parietal extension. Surgical treatment was excluded. Digital