A Case of Cystic Adventitial Degeneration of the Left Popliteal Artery Diagnosed by Intravascular Ultrasound

Takeshi Niizeki[1], Mitsunori Ishino[1], Tatsuro Kitahara[1], So Yamauchi[1], Eiichiro Ikeno[1] and Isao Kubota[2]

[1]Department of Cardiology, Okitama Public General Hospital, Yamagata, Japan. [2]First Department of Internal Medicine, Yamagata University School of Medicine, Yamagata, Japan.

ABSTRACT: An 87-year-old male was admitted with intermittent claudication of the left calf. We performed lower extremity angiography, which revealed stenosis of the left popliteal artery. Intravascular ultrasound (IVUS) image correctly identified the cystic appearance of visualized extravascular hypodensity, causing extrinsic compression of the lumen. We diagnosed the condition as cystic adventitial degeneration (CAD) of the popliteal artery. We operated a resection of a cyst with the artery and replaced the autovein graft (saphenous vein). After surgery, the patient was free of symptoms. CAD is a rare disease; thus, our IVUS findings may provide unique diagnostic clues in patients with CAD.

KEYWORDS: cystic adventitial degeneration, imaging invasive IVUS, peripheral artery disease

CITATION: Niizeki et al. A Case of Cystic Adventitial Degeneration of the Left Popliteal Artery Diagnosed by Intravascular Ultrasound. *Clinical Medicine Insights: Case Reports* 2016:9 11–14 doi: 10.4137/CCRep.S38175.

TYPE: Case Report

RECEIVED: December 10, 2015. **RESUBMITTED:** January 25, 2016. **ACCEPTED FOR PUBLICATION:** January 28, 2016.

ACADEMIC EDITOR: Athavale Nandkishor, Associate Editor

PEER REVIEW: Three peer reviewers contributed to the peer review report. Reviewers' reports totaled 427 words, excluding any confidential comments to the academic editor.

FUNDING: Authors disclose no external funding sources.

COMPETING INTERESTS: Authors disclose no potential conflicts of interest.

CORRESPONDENCE: takeshi.niizeki@okitama-hp.or.jp

Introduction

Cystic adventitial degeneration (CAD) is a rare disease but a serious claudication that causes vascular stenosis, usually in middle-aged men.[1,2] In ~85% of cases reported, CAD affects the popliteal artery.[1,2] Typically, duplex sonography, angiography, contrast-enhanced computed tomography, and magnetic resonance imaging (MRI) are used to establish a diagnosis.[3,4] Earlier studies on CAD are almost single case reports or small case series.[3,4] As little is known about intravascular ultrasound (IVUS) findings and their importance in patients with CAD, we report here our experience of a case in which IVUS correctly identified CAD of the popliteal artery when angiography was inconclusive. The patient has given consent for publication of this report.

Case Report

An 87-year-old male was referred to our hospital with a complaint of intermittent claudication in the left calf with a walking distance of about 100 m. His coronary risk factors were hypertension, diabetes mellitus, dyslipidemia, and chronic kidney disease (estimated glomerular filtration rate 51 mL/minute/1.73 m^2). He denied recent trauma. There was no cyanosis of the left lower limb, no ulceration, and no dystrophic signs. A routine hemogram and biochemistry data were within normal limits. He had no history of connective tissue disease, which was also excluded by physical and laboratory examinations. His ankle-brachial index (ABI) was 0.92 on the right leg and 0.46 on the left leg., and peripheral artery disease was suspected. As shown in Figure 1, digital subtraction angiography (DSA) of the left lower extremity showed that the left popliteal artery had a severe eccentric stenosis. We performed DSA using carbon dioxide contrast because of renal dysfunction. The proximal and distal reference segments had no evidence of atherosclerosis. Attempt to pass the stenotic site by a 0.014 inch chevalier floppy wire (Future Medical Design Corporation) was performed, and the guide wire was easily crossed. IVUS (ViewIT; Terumo Corporation) revealed a nonatherosclerotic lesion and cystic appearance; the area of extravascular hypodensity was clearly visualized, which was causing extrinsic compression of the lumen (Fig. 2). Because we considered a differential diagnosis, including cyst, tumor, and CAD by these IVUS findings, we stopped endovascular therapy and performed MRI. The axial view of MRI revealed cystic lesions encompassing the left popliteal artery circumferentially (Fig. 3A). In the sagittal view of MRI, these cystic lesions also exhibited high signal intensity on T2-weighted images clearly (Fig. 3B). Because of the characteristic location and clinical signs, we diagnosed CAD of the popliteal artery. Thus, we considered that IVUS may be used as an adjunctive tool to diagnose and evaluate CAD when angiography was inconclusive. Surgical exploration was recommended, and an informed consent was obtained. During surgery, a diseased segment of the popliteal artery was isolated. Gross evidence of gelatinous and

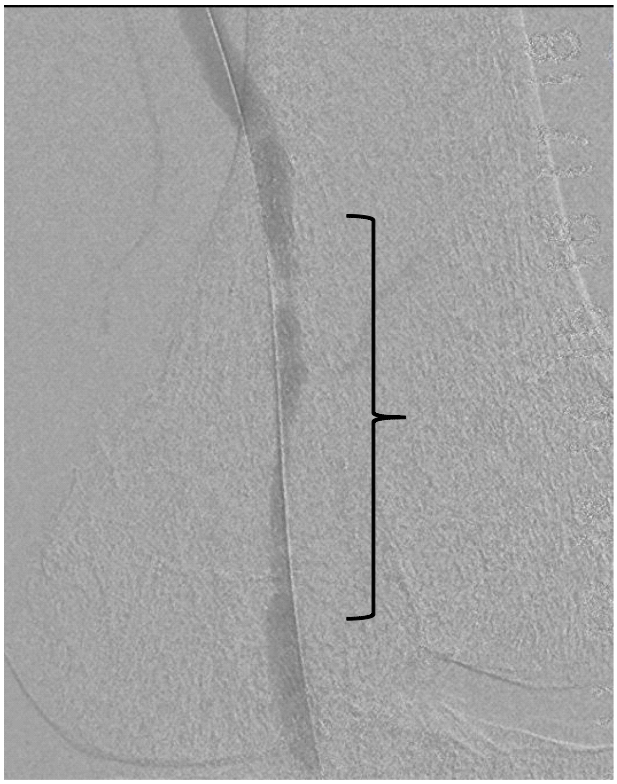

Figure 1. DSA by carbon dioxide contrast showed the stenosis of the popliteal artery and smooth proximal and distal reference segments.

mucoid degeneration was noted. Fibrosis, necrosis, calcification, and ulceration were not found around the cyst. The segment was resected, and an interposition graft was completed with a segment of reversed saphenous vein. Histopathologic analysis of the segment demonstrated myxoid change with formation of cystic spaces, primarily involving the adventitia (Fig. 4). We definitively diagnosed CAD of the popliteal artery by these findings. Spinner et al.[5] recently reported the joint connection of the middle genicular artery in patients with CAD, that is, the conduit for cyst propagation from the knee joint to the parent vessel. However, in our case, a joint connection was not identified on MRI or at operation. The postoperative course was uneventful. The follow-up ABI was normalized, and clinical success with symptom resolution was achieved.

Discussion

Here, we report a rare case in which IVUS finding was useful for diagnosis and evaluation of CAD of the popliteal artery. Although stenotic lesions in lower extremities arteries are mostly caused by arteriosclerosis obliterans, nonatherosclerotic causes such as popliteal artery entrapment syndrome, CAD, and thrombosed aneurysm must be considered in lesions of the popliteal artery as this case.

Atkins and Key[6] first reported CAD of the iliac artery in 1947 and Hiertonn and Lindberg[7] first described CAD

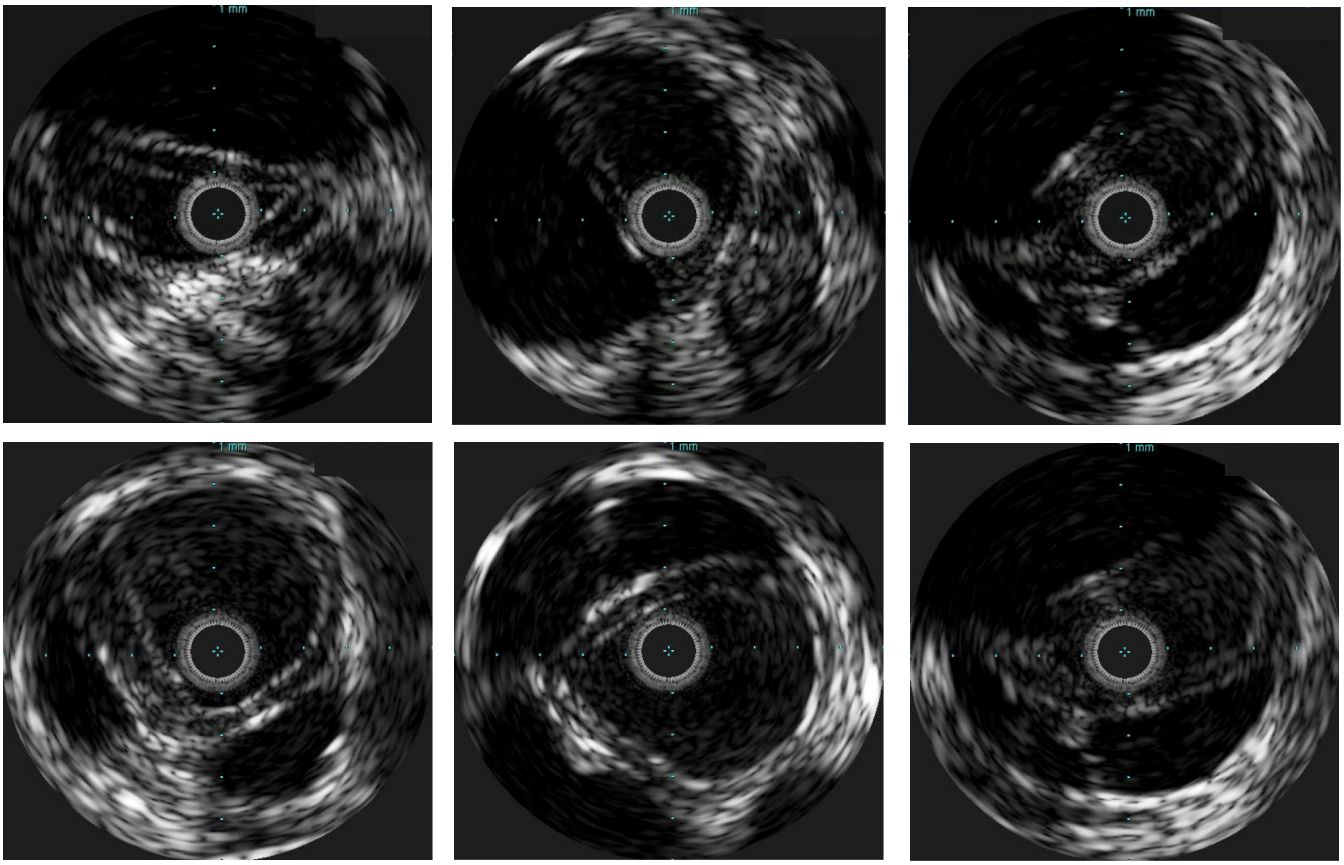

Figure 2. Intravascular ultrasound revealed the nonatherosclerotic lesion and the extrinsic compression of the lumen by the extravascular cyst. The cystic appearance (extravascular hypodensity area) can be visualized clearly.

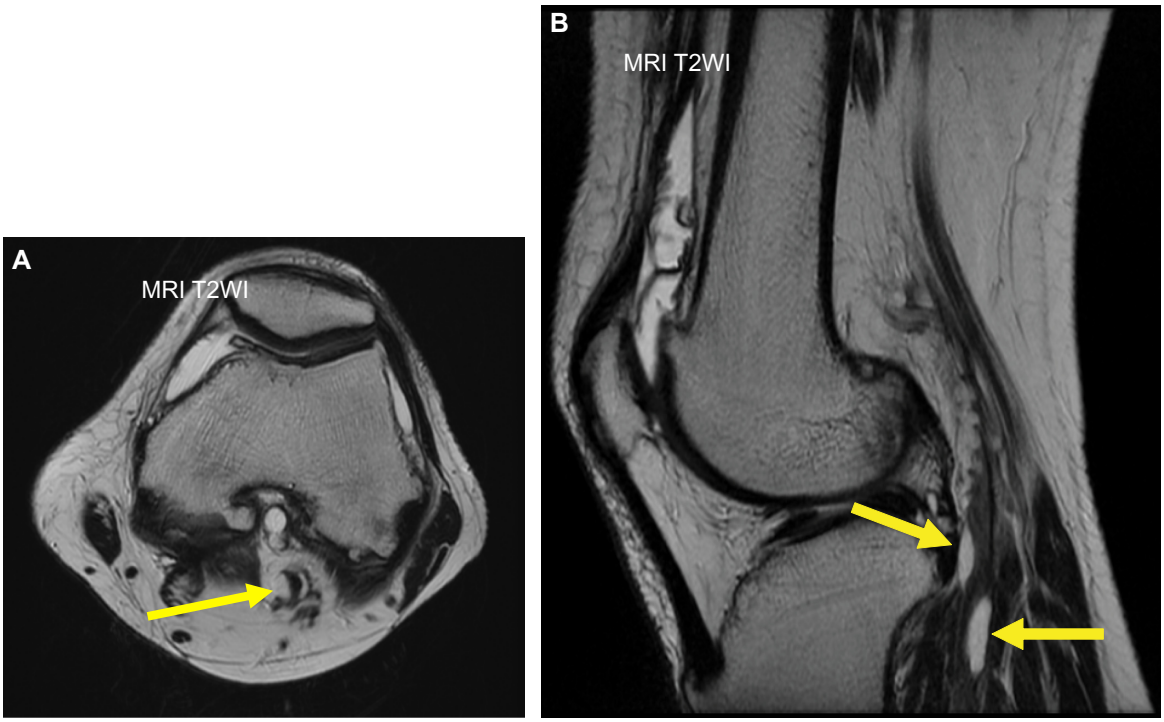

Figure 3. The axial image: MRI clearly showed cystic lesions encompassing the left popliteal artery circumferentially (**A**). The sagittal image: these cystic lesions also exhibited high signal intensity on T2-weighted images clearly (**B**).

of the popliteal artery in 1957. Ishibashi et al.[8] reported age and sex distribution of CAD: males (83.2%) and average age 44.6 years. Most of the patients with CAD are less than 50 years of age in general, and case reports of patients older than 80 years are uncommon, with only two cases. Thus, it was difficult to diagnose in our case because the patient was of advanced old age (87 years).

The detailed etiology of CAD has been unclear yet. Several hypotheses have been put forward: microtraumatic origin, repeated stretch injuries causing degeneration of the arterial adventitia,[9] embryologic origin, embryological displacement of mucin-secreting cells into the adventitia of the artery from endothelium of the knee joint,[10] and ganglion formation; cyst is a true ganglion of the vessel wall.[11]

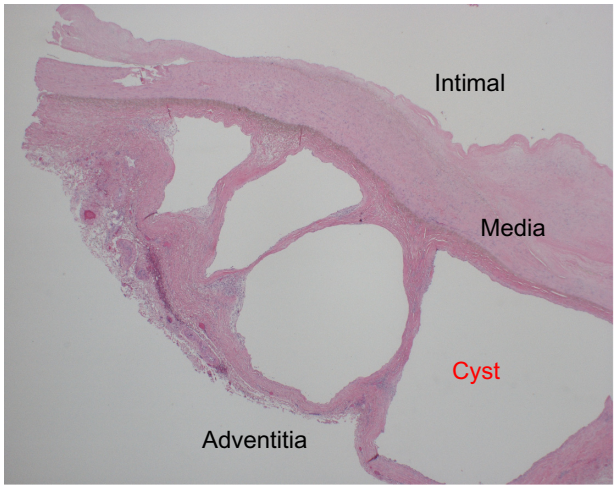

Figure 4. Hematoxylin–eosin staining showed that the cyst exists in the adventitia and the cyst wall has no endothelial cells.

Treatment of CAD typically consists of complete surgical resection of the diseased arterial segment with reversed saphenous vein graft interposition.[4] Because of both the high incidence of recurrence and the risk of dissection or rupture, percutaneous transluminal angioplasty and needle aspiration were ineffective.[12] Thus, we decided to treat by resection and autovein grafting because of the lowest recurrence rate.

Typically, duplex sonography, angiography, and MRI are used to establish a diagnosis.[3,4] Body surface sonography is useful for identifying the cyst and diagnostic imaging of CAD. Unfortunately, the specificity of duplex sonography is dependent on the skill of the technician. Angiography is also one of the useful techniques for diagnosing CAD. However, the diagnosis and degree of stenosis remain uncertain. In addition, MRI is useful for identifying and diagnostic imaging of CAD. Although IVUS is an invasive imaging device, IVUS finding might be one of the adjunctive tools as our case. IVUS excluded atherosclerosis, hematoma, aneurysm, and dissection and clearly identified the extravascular cyst that compressed the lumen in real time (Fig. 2). CAD is a rare disease; thus, IVUS findings might provide unique diagnostic clues in patients with CAD and be one of the useful and feasible methods for evaluating CAD.

Conclusion

Although uncommon, CAD should be considered as a potential cause of intermittent claudication by the lesion of the

popliteal artery. Our report raises the possibility that diagnosis of CAD supported by IVUS device is suitable.

Author Contributions

Conceived and designed the experiments: TN. Analyzed the data: TN, MI. Wrote the first draft of the manuscript: TN. Contributed to the writing of the manuscript: TN. Agree with manuscript results and conclusions: TN, TK. Jointly developed the structure and arguments for the paper: TN, SY. Made critical revisions and approved final version: TN, EI, IK. All authors reviewed and approved of the final manuscript.

REFERENCES

1. Macfarlane R, Livesey SA, Pollard S, Dunn DC. Cystic adventitial arterial disease. *Br J Surg*. 1987;74:89–90.
2. Mangialardi N, Serrao E, Tozzi A, Illuminati G, Ferri E, Colonna M. Adventitial cyst of the popliteal artery. Report of a case. *Panminerva Med*. 1996;38:117–20.
3. Altehoefer C, Blum U, Ebert D. Cystic adventitial degeneration and entrapment syndrome of the popliteal artery as a differential diagnosis of popliteal stenosis or occlusion in the younger age group. *Vasa*. 1998;27:179–82.
4. Inoue Y, Iwai T, Ohashi K, et al. A case of popliteal cystic degeneration with pathological considerations. *Ann Vasc Surg*. 1992;6:525–9.
5. Spinner RJ, Desy NM, Agarwal G, Pawlina W, Kalra M, Amrami KK. Evidence to support that adventitial cysts, analogous to intraneural ganglion cysts, are also joint-connected. *Clin Anat* 2013; 26:267–81.
6. Atkins HJ, Key JA. A case of myxomatous tumour arising in the adventitia of the left external iliac artery; case report. *Br J Surg*. 1947;34:426.
7. Hiertonn T, Lindberg K. Cystic adventitial degeneration of the popliteal artery. *Acta Chir Scand*. 1957;113:72–7.
8. Ishibashi S, Namiki K, Abe M, et al. Cystic adventitial disease of the popliteal artery – a case of young boy. *Tohoku J Exp Med*. 1995;176:173–80.
9. Hiertonn T, Lindberg K. Cystic adventitial degeneration of the popliteal artery. *Acta Chir Scand*. 1957;113:72–7.
10. di Marzo L, Della Rocca C, d'Amati G, et al. Cystic adventitial degeneration of the popliteal artery: lectin-histochemical study. *Eur J Vasc Surg*. 1994;8:16–9.
11. Briselli MF, Landers AD. Cystic adventitial degeneration of the popliteal artery. *Wis Med J*. 1982;81:22–4.
12. Schöllhorn J, Arnolds B, von Reutern GM, Schlosser V. Cystic adventitial degeneration as a cause of dynamic stenosis of the popliteal artery: a case report. *Angiology*. 1985;36:809–14.