

Adventitial Cystic Disease of the Iliac Artery with a Connection to the Hip Joint

Hyung-Keel Kim, Sujin Park, Deokbi Hwang, and Seung Huh

Division of Vascular Surgery, Department of Surgery, School of Medicine, Kyungpook National University, Daegu, Korea

More than 700 adventitial cystic diseases (ACDs) have been reported in the literature, with most cases affecting the popliteal artery in young men. Here, we describe our treatment and etiologic consideration of a patient who presented with an ACD of the external iliac artery, known to be an extremely rare location. On preoperative imaging, the ACD had a connection to the nearby hip joint and was treated with resection of the affected segment, including ligation of the joint connection and interposition with a prosthetic graft. The pathogenesis of ACDs is not fully understood; however, we believe that joint connections are important in their development and treatment.

Key Words: Adventitia, Cyst, Iliac artery, Hip joint

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Corresponding author: Seung Huh

Division of Vascular Surgery, Department of Surgery, School of Medicine, Kyungpook National University, 130 Dongdeok-ro, Jung-gu, Daegu 41944, Korea

Tel: 82-53-420-6520

Fax: 82-53-421-0510

E-mail: shuh@knu.ac.kr

<http://orcid.org/0000-0002-0275-4960>

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INTRODUCTION

Adventitial cystic disease (ACD) is known to be a rare nonatherosclerotic cause of peripheral arterial occlusive disease, mainly affecting young men [1,2]. In general, cystic lesions can occur around any of the joints or tendon sheaths in the body and more than 700 ACDs have been reported in the literature with the majority of cases affecting the popliteal artery [2,3]. However, an ACD involving the iliac artery is extremely rare.

The etiology of ACDs remains uncertain, but various causes, such as degenerative processes, repeated trauma, involvement of ganglia, and abnormal development have been proposed in the literature [4]. Links between the formation of ACDs and adjacent joints have been proposed and have been found in a significant proportion of patients using high-resolution imaging methods, such as magnetic resonance imaging (MRI), and have been used to support the two most popular theories (developmental and ganglion anomalies) concerning the pathogenesis of ACDs [5,6]. Here, we describe our treatment and etiologic consideration of

a patient who presented with an ACD of the external iliac artery, known to be an extremely rare location.

CASE

A 56-year-old male construction worker complained of a claudication that had persisted for 1 month. His medical history was unremarkable except for hyperlipidemia and a history of smoking for 20 years. On physical examination, the right femoral and ankle pulses were weakly palpable. The resting ankle-brachial index (ABI) was measured as 0.86 on the right side and 1.17 on the left side. However, the exercise ABI using a treadmill protocol for 5 min at a speed of 3.2 km/h and an incline of 10° remained normal on the left, but dropped to 0.25 on the right side. Computed tomography (CT) scans revealed a cystic mass located superiorly to the right external iliac artery and severe stenosis of the distal external iliac artery as a “scimitar sign” (Fig. 1A). MRI also showed a cystic lesion of the external iliac artery and demonstrated a communication with the right hip joint (Fig. 1B).

The patient underwent surgical exploration using a

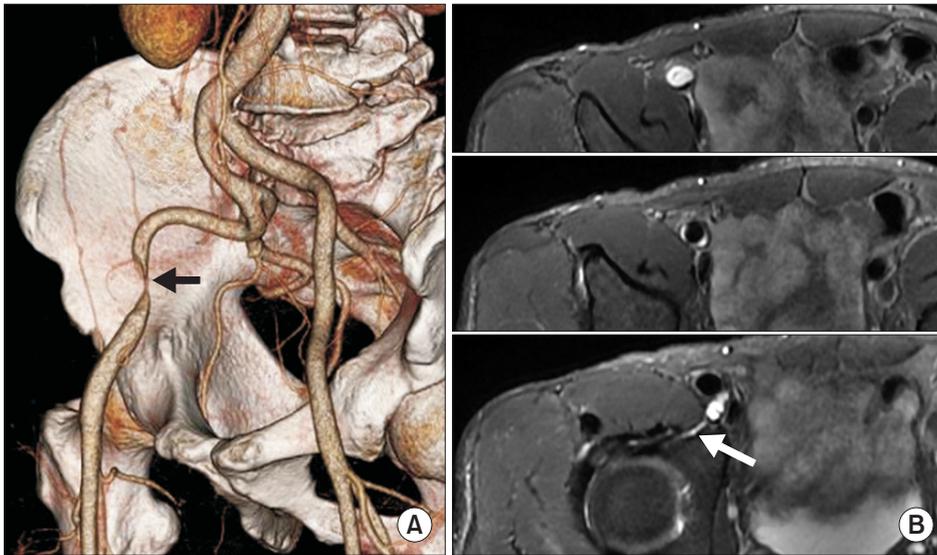


Fig. 1. Preoperative images of adventitial cystic disease. (A) Multidetector row computed tomography with 3-dimensional volume rendering showed the "scimitar sign" (black arrow) without features of atherosclerosis. (B) Axial T2-weighted magnetic resonance imaging showed a cystic mass arising from the right hip joint with a connection to it (white arrow).



Fig. 2. Intraoperative and histological images. (A) Intraoperative photograph showed multiple cystic masses at the anterior aspect of the right external iliac artery (black arrow). (B) The resected specimen demonstrated compression of the arterial lumen by a multiloculated cyst divided by septa within the adventitia and a connection from the right hip joint (white arrows).

retroperitoneal approach. Intraoperative findings were consistent with previous images, presenting mucinous-filled multiple cysts adjacent to the anterior aspect of the right external iliac artery (Fig. 2A). The affected segment of the external iliac artery was resected and interposed with a prosthetic graft. A connection to the nearby hip joint was identified and ligated with nonabsorbable sutures. His postoperative course was uneventful; the follow-up resting ABI returned to 1.12 on the right side and his claudication disappeared. The resected specimen demonstrated compression of arterial lumen by multiple cysts divided by septa within the adventitia (Fig. 2B). The patient underwent CT scan 1 year after the procedure, showing no signs of recurrence.

DISCUSSION

Our patient presented an ACD of external iliac artery and imaging studies showed a connection between the ACD and the hip joint. We have already published a case of an

ACD in the common femoral artery with a connection to hip joint [7]. The patient in this case also had a connection to hip joint and received an interposition graft after resection of affected segment.

Currently, more than 600 arterial ACDs have been reported in the literature, with nearly 90% of cases affecting the popliteal artery with symptoms of claudication in young men lacking other vascular system risk factors [2]. Among arterial ACDs, femoral artery is the second most common location and approximately 37 cases have been reported to date. An iliac arterial ACD is extremely rare and only eight cases have been reported [2]. The exact etiology of ACDs remains uncertain, but various hypotheses have been proposed. Levien and Benn [4] discussed four theories: (1) the degenerative theory with a mucinous or myxomatous systemic degenerative condition associated with a generalized disorder; (2) repeated trauma causing destruction and cystic degeneration of the adventitia of the adjacent vessel; (3) adventitial cysts arising as capsular ganglionic structures that then enlarge and track along vascular branches

to involve the adventitia of the adjacent major vessel; and (4) mucin-secreting cells derived from the mesenchyme of the adjacent joint are incorrectly placed in the vessel wall during the development of the disease. However, the hypotheses supported by most authors involve ganglionic and developmental anomalies [4,5]. Connections between ACDs and adjacent joints, as in this case and our previous report [7], have been used to support these two most popular theories concerning the pathogenesis of ACD. Proponents of the “developmental” theory consider this connection to adjacent joints to be a residue of embryogenesis. In contrast, proponents of the “ganglion” theory see the anatomical connection as resulting from herniation of the synovium through a breach in the adjacent articulation [8]. In one recent literature review of 746 ACDs, all of the cysts were para-articular, suggestive of an association between ACD formation and neighboring joints, supporting both the developmental and/or ganglion theories [2]. A joint connection was identified in 122 cases (16.4%) of 746 ACDs on imaging, during surgery, or both [2].

This connection can be an important issue in the management of these conditions. In the literature, ACDs have been managed by nonresectional and resectional methods. Although cyst resection with vascular reconstruction has been the most popular method of treatment, percutaneous aspiration and cyst evacuation with removal of the cystic wall have been used as a treatment option [9,10]. However, the recurrence was as high as to 41% in cases with percutaneous aspiration and 11% in cyst evacuation with cystic wall excision [2]. In many ACDs, as in our case, the lesions are usually multiloculated with septa delineating multiple smaller cysts and the cystic fluid has high viscosity. There-

fore, aspiration of such multiloculated cysts can result in incomplete aspiration of the contents with failure of treatment. In addition, in patients with connection to a nearby joint, the cystic fluid can reaccumulate within or near the ACDs with resultant recurrence of arterial compression if the connection is not ligated.

In cases of resectional methods for treatment, recurrence has also been reported in a few patients [11]. These cases can also be explained by unresected multiloculated cyst contents presented as in our case, or reaccumulation of cystic fluid around the treated artery from the remaining unligated connection to the nearby joint. In this case and our previous report in patient with femoral ACD [7], the joint connection could be found in preoperative MRI scans and such connections can be ligated. As a result, recurrence did not occur during short-term follow-up periods. Spinner et al. [5] also demonstrated the efficacy of high-resolution MRI, and a joint connection could be found in all of the five patients for whom MRI was available. Therefore, recognition of such connections and proper management of them will lead to better and more robust outcomes in the treatment of these patients.

In conclusion, we have described a rare case of an iliac arterial ACD and have discussed its possible etiology together with our previous report of a patient suffering from an ACD of the common femoral artery. Connections to the hip joint were identified in both patients on preoperative MRI, and these were ligated successfully during surgery. This case reemphasizes the role of joint connections in the development of ACDs, and the need for proper evaluation and management of such connections for preventing any recurrence.

REFERENCES

- 1) Paravastu SC, Regi JM, Turner DR, Gaines PA. A contemporary review of cystic adventitial disease. *Vasc Endovasc Surg* 2012;46:5-14.
- 2) Lejay A, Ohana M, Delay C, Georg Y, Girsowicz E, Thaveau F, et al. Cystic adventitial pathology as an entity in peripheral arterial disease. *J Cardiovasc Surg (Torino)* 2016;57:282-291.
- 3) Yukata K, Nakai S, Goto T, Ikeda Y, Shimaoka Y, Yamanaka I, et al. Cystic lesion around the hip joint. *World J Orthop* 2015;6:688-704.
- 4) Levien LJ, Benn CA. Adventitial cystic disease: a unifying hypothesis. *J Vasc Surg* 1998;28:193-205.
- 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;26:267-281.
- 6) Tsilimparis N, Hanack U, Yousefi S, Alievizakos P, Rückert RI. Cystic adventitial disease of the popliteal artery: an argument for the developmental theory. *J Vasc Surg* 2007;45:1249-1252.
- 7) Kim HK, Hwang D, Park S, Jeong WJ, Seo AN, Huh S. Cystic disease of the groin presenting as compression of a femoral vessel. *Vasc Spec Int* 2016;32:124-128.
- 8) Ortmann J, Widmer MK, Gretener S, Do DD, Willenberg T, Daliri A, et al. Cystic adventitial degeneration: ectopic ganglia from adjacent joint capsules. *Vasa* 2009;38:374-377.
- 9) Do DD, Braunschweig M, Baumgartner I, Furrer M, Mahler F. Adventitial cystic disease of the popliteal artery: percutaneous US-guided aspiration.

- Radiology 1997;203:743-746.
- 10) Kim HK, Choi HH, Huh S. Cystic adventitial disease of the popliteal artery: a recurrent case after cyst wall excision. *Int Angiol* 2012;31:198-201.
- 11) Flessenkaemper I, Müller KM. Early recurrence of cystic adventitial disease in a vein graft after complete resection of the popliteal artery. *Vasa* 2014;43:69-72.