**Popliteal Artery Entrapment Syndrome in Siblings**

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Popliteal artery entrapment syndrome (PAES) is a rare disease. We treated siblings with this disease. An 18-year-old male consulted our hospital for intermittent claudication of the left lower limb. Contrast-enhanced computed tomography led to a diagnosis of type II PAES. After transection of the medial head of the gastrocnemius muscle, popliteal artery bypass was performed. His younger brother (6 years younger) was also diagnosed with type II PAES, and similar surgery was performed at the age of 19. These cases suggested the involvement of genetic factors in PAES in addition to embryological factors.

**Keywords:** popliteal artery entrapment syndrome, familial occurrence, hereditary disorder

**Introduction**

The gastrocnemius muscle is primarily involved in popliteal artery entrapment syndrome (PAES). Its pathogenesis may involve positional abnormalities of medial head adhesion or chronic compression of the popliteal artery/vein and tibial nerve related to abnormal muscle bundles in the popliteal fossa. We treated siblings with PAES by popliteal artery bypass. The onset of PAES in siblings is rare. Sofar, only four case reports have been published in English. Our findings suggested the involvement of genetic factors in PAES.

**Case**

**Case 1**

An 18-year-old male was referred to our hospital for intermittent claudication (200 m). The left ankle-brachial index (ABI) was decreased to 0.59. Three-dimensional computed tomography (3D-CT) revealed occlusion of the left popliteal and anterior tibial artery. Under general anesthesia, the left popliteal artery was exposed via a posterior approach, and then, it was compressed and occluded by the medial head of the gastrocnemius muscle and the medial head of the gastrocnemius muscle was located between the artery and vein (Fig. 1). Based on these findings, a diagnosis of type II PAES was made (Rich’s classification). The anterior tibial artery was supposed to be occluded due to distal embolism. Under general anesthesia, the left popliteal artery was exposed via a posterior approach, and then, it was compressed and occluded by the medial head of the gastrocnemius muscle; therefore, the medial head of the left gastrocnemius muscle was transected and left popliteal artery bypass with a reversed great saphenous vein was performed. After surgery, the ABI recovered to 0.8 and claudication disappeared. The postoperative course was favorable, and the patient was discharged on the 8th day after the surgery. No recurrence of claudication was found during the 9-year postoperative follow-up.

**Case 2**

The younger brother (6 years younger) of Case 1 patient was referred to our hospital for left intermittent claudication (250 m) at the age of 19. The left ABI was decreased to 0.5. 3D-CT showed similar findings as Case 1, leading to a diagnosis of type II PAES. The medial head of the left gastrocnemius muscle was thicker than in Case 1. Furthermore, all arteries distal to the trifurcation in the lower leg were patent (Fig. 2). Under general anesthesia, transection of the medial head of the left gastrocnemius muscle and left popliteal artery bypass with a reversed great saphenous vein were performed, as described for Case 1. After surgery, the ABI recovered to 1.0 and claudication disappeared. The postoperative course was favorable, and the patient was discharged on the 8th day after surgery. No recurrence of claudication was found during the 1-year postoperative follow-up.
Fig. 1  Computed tomography (CT) findings. 
(a) The left popliteal artery and the anterior tibial artery were occluded (arrow). (b) On axial CT, the course of the occluded left popliteal artery (arrow) was medially shifted apart from the left popliteal vein, and the medial head of the gastrocnemius muscle was located between the occluded left popliteal artery and the patent popliteal vein.

Fig. 2  Computed tomography (CT) findings. 
(a) The left popliteal artery was occluded (arrow) with patent distal tibial arteries. (b) On axial CT, the left popliteal artery (arrow) was medially shifted and was occluded. The medial head of the gastrocnemius muscle (arrow head) was located between the occluded popliteal artery and the patent popliteal vein, as described in Case 1. The medial head of the gastrocnemius muscle was more thickened than that of Case 1.

Table 1  A summary of reports on sibling-onset popliteal artery entrapment syndrome

| Author                  | Siblings       | Age (years) | Lateratry | Type (Rich classification) | Treatment                  |
|-------------------------|----------------|-------------|-----------|---------------------------|----------------------------|
| Jikuya et al.           | Older brother  | 19          | R & L     | R:II                      | R: VG                      |
|                         | Younger brother| 19          | L         | L:II                      | L: Myotomy and VG          |
| Al-Basheer et al.       | Older brother  | 30          | L         | Unknown                   | Unknown                    |
|                         | Younger brother| 27          | R         | Unknown                   | Myotomy and VG             |
| Kfoury et al.           | Older brother  | 17          | L         | Unknown                   | VG                         |
|                         | Younger brother| 17          | R         | Unknown                   | VG                         |
| Clifford et al.         | Older brother  | 24          | R         | II                        | Myotomy and VG             |
|                         | Younger brother| 19          | L         | II                        | Myotomy and VG             |
| Orimoto et al.          | Older brother  | 18          | L         | II                        | Myotomy and VG             |
|                         | Younger brother| 19          | L         | II                        | Myotomy and VG             |

VG: vein graft
Discussion

PAES was first reported in 1879 by Stuart, a medical student in Edinburgh. Since that time, many reports have been published, but its pathophysiology and etiology remain unclear. Embryological abnormalities of the popliteal artery and medial head of the gastrocnemius muscle in the developmental process of lower limb arteries in the embryonic phase may be etiologically involved. The present cases suggest that PAES could occur in siblings. Only four reports on the sibling-onset PAES have been published in English; all of these involved its onset in brothers. The type of PAES was consistent between brothers in two reports, while it was unclear in the other two (Table 1). In the present cases, the type of PAES was consistent and the left side was affected. This suggests that genetic and environmental factors, in addition to embryological factors, are involved in the etiology of PAES. With further advances in genetic testing, the presence of the familial inheritance pattern of PAES may be explained, which may influence its diagnosis and treatment.

PAES is a rare but an important cause for intermittent claudication in young persons. It accounts for 60% of intermittent claudication in young patients. Sinha et al. conducted a meta-analysis and reported that the mean age at the time of onset was 32 years, males accounted for 83% of patients with PAES, and the bilateral sides were affected in 38%. Its incidence was reported to be 3.5% based on postmortem autopsies, which is more common than that previously reported. In the present cases (Cases 1 and 2), there were no findings suggestive of PAES on the contralateral side.

With regard to clinical symptoms, intermittent claudication is observed in most patients, as the extent of arterial occlusion frequently involves the popliteal artery alone. However, the degree of arterial occlusion and distal embolism due to aneurysm formation lead to pain at rest, toe necrosis, or ulceration in some patients. Distal embolism may be the cause of the postoperative ABI recovery of 0.8 in Case 1. PAES, as a cause of intermittent claudication at a young age, may be overlooked due to the absence of cardiovascular risk factors. No accurate diagnosis is made and inappropriate treatment is conducted for some patients. As findings, the attenuation or disappearance of arterial pulsation or Doppler sounds related to passive dorsiflexion and active plantar flexion of the ankle is observed in the phase of initial arterial compression alone. Diagnostic imaging procedures include echography, magnetic resonance angiography (MRA), CT, and angiography. However, recently, a definitive diagnosis has been made using contrast-enhanced CT or CT angiography.

For treatment, in most cases, surgery is performed. If organic lesions of the popliteal artery, including occlusion and stenosis, are absent, favorable results may be obtained by myotomy of the medial head of the gastrocnemius muscle or resection of abnormal muscle bundles alone. Early detection is important but difficult. Most patients have occlusion or marked stenosis of the popliteal artery, and angioplasty or revascularization is performed. Follow-up after revascularization is very important. However, at present, there is no consensus on long-term data, including graft patency. Present two patients had a younger brother with no ischemic symptoms in the lower limbs. PAES was evaluated using contrast-enhanced CT at the age of 17. No abnormalities of the bilateral lower limb arteries or veins, including PAES, were found. In our case, the two older brothers developed PAES, and the presence of PAES was actively investigated using CT. Recognition of familial PAES may facilitate early detection and treatment by minimally invasive myotomy of the medial head of the gastrocnemius muscle or resection of abnormal muscle bundles alone.

Conclusion

Reports on sibling-onset PAES are rare. Recognition of familial onset may lead to an early diagnosis of PAES, facilitating minimally invasive treatment.

Disclosure Statement

The authors have no conflicts of interest to declare.

Author Contributions

Study conception: YO
Data collection: YO
Investigation: YO
Writing: YO, HI
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Final approval of the article: all authors
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