Persistent sciatic vessels, varicose veins, and lower limb hypertrophy: An unusual case or discrete clinical syndrome?

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Persistent sciatic artery is a rare congenital anomaly with a high incidence rate of aneurysmal degeneration and risk of thromboembolization or rupture. Despite a number of recognized associations, the presence of coexistent venous anomalies is extremely rare. We present the case of a 27-year-old woman with atypical left-sided varicose veins and soft tissue hypertrophy. Imaging showed persistence of both sciatic artery and vein. Whether these anomalies are an incidental finding or represent a discrete clinical syndrome remains unclear. We emphasize that unusual distribution varicose veins may be associated with underlying persistent sciatic vessels and recommend formal duplex scan assessment for these anomalies. (J Vasc Surg 2002;36:396-400.)

The axial artery provides the dominant blood supply to the lower limb bud during early embryologic development. Failure to involute results in a rare congenital anomaly known as persistent sciatic artery (PSA). PSA is associated with other congenital anomalies, including Mullerian and renal agenesis, arteriovenous fistulae, bone or soft tissue hypertrophy/hypotrophy, and abnormalities of the lower limb arteries. Coexistent venous anomalies, however, are extremely rare. Persistence of both sciatic vessels has been reported as an incidental finding only twice in the English-language literature. More recently, an association between PSA, varicose veins, and limb length discrepancy in young patients has been described. We present the case of a 27-year-old woman with gross left-sided telangectasia, varicose veins, and soft tissue hypertrophy. Imaging showed a persistence of both sciatic artery and vein with valvular incompetence. This combination of congenital anomalies has been previously unreported.

CASE REPORT

A 27-year-old woman was seen in the outpatient department with left-sided varicose veins. These had become progressively more prominent over the previous 20 years and ached on standing. The patient had no family history of varicose veins nor medical history of deep venous thrombosis, thrombophlebitis, or claudication. Examination revealed extensive left-sided telangectasia, reticular, and varicose veins. These were atypical in distribution, with extensive involvement of both the medial and lateral aspects of the leg (Fig 1). Although mild venous pigmentation was seen in the lower leg, no port-wine stain was visible. The left thigh and calf measured 4 cm and 2.5 cm, respectively, more than the right leg, without discrepancy in length. No clinical lymphoedema was found. The left femoral pulse was weaker than the right, but the left popliteal pulse was bounding. All other pulses were of normal volume and ankle-brachial pressure indices were 1.3 bilaterally. Hand-held Doppler scan showed an easily audible biphasic arterial signal in the popliteal fossa and evidence of venous reflux.

Ascending venography showed short saphenous varices with a venous aneurysm. The short saphenous vein drained into the popliteal vein, and filling of a single ectatic vein in the thigh was seen (Fig 2). This vein was lying in the line of the femoral axis at the level of the hip and was initially presumed to be the superficial femoral vein (SFV). Duplex scan was performed immediately afterwards, however, and showed the SFV to be hypoplastic. The ectatic thigh vein was seen to course posteriorly, running into the line of the profunda vein, and transient valvular reflux was found in the deep veins. These findings were consistent with the presence of a lower persistent sciatic vein (PSV), draining into the profunda vein and thence into the common femoral/iliac veins. Finally, the saphenopopliteal junction (SPJ) was incompetent, with sustained reflux into a grossly dilated short saphenous vein.

Arterial duplex scan showed a complete PSA, in continuity with the hypogastric and popliteal arteries. The left common iliac artery was ectatic, measuring 2 cm in diameter. The axial vessel was also ectatic, measuring up to 1.5 cm in the thigh, but without focal aneurysm formation. Magnetic resonance angiography showed arteriomegaly of the PSA in the thigh with some abnormal collaterals (Fig 3). Subsequent intraarterial digital subtraction angiography showed the typical course of the PSA in the left leg (Fig 4). Multiple abnormal arterial vessels with an increased capil-
Fig 1. Atypical distribution left-sided telangectatic, reticular, and varicose veins.

Fig 2. Ascending venography. A, Oblique view shows ectatic short saphenous vein with venous aneurysm (small arrow). B, Oblique view shows short saphenous vein (large arrow) draining into popliteal vein. C, Anteroposterior view shows filling of single ectatic vein in thigh. This can be seen as in line with femoral axis at level of hip.
lary stain were seen, possibly suggestive of shunting at capillary level, although no discrete arteriovenous malformation was visible. The patient’s arterial and venous anomalies are shown in Fig 5.

Although SPJ ligation with multiple stab avulsions may have improved the patient’s symptoms, we believed that a significant risk of recurrent varices in the presence of deep venous reflux existed. Furthermore, this procedure would not improve the appearance of many of the extensive reticular and telangiectatic veins in the thigh. After discussion of these factors, the patient opted for conservative treatment in compression hosiery. After 2 years, she remains well and has undergone serial arterial duplex scan surveillance of PSA without incident.

**DISCUSSION**

The axial artery is a dorsal branch of the primitive umbilical artery and provides the dominant blood supply to the lower limb bud in the 9-mm embryo. The femoral artery is a continuation of the external iliac artery and is visible by the 12-mm stage. As the femoral artery develops, synchronous involution of the axial system occurs. By the 22-mm stage, the familiar adult vascular tree has fully developed. Failure of the femoral artery to develop, or the axial artery to involute, results in a PSA.1,7 Although the embryologic development of the arterial system of the lower limb is well described, this is not true of the venous

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**Fig 3.** Magnetic resonance angiogram shows arteriomegaly of left-sided PSA with some abnormal primitive collateral vessels (white arrows). Superficial femoral artery is absent, and below-knee anterior tibial and peroneal arteries arise from common trunk.

**Fig 4.** Intraarterial digital subtraction angiogram shows typical course of PSA in pelvis and leg (large arrow). Multiple abnormal arterial vessels (small arrows) with increased capillary stain are seen, but no evidence is found of discrete arteriovenous fistula.
system. Within weeks of the axial artery development, a peripheral border vein provides venous outflow from the limb bud. Ultimately, the tibial continuation of this disappears and the fibula portion becomes the anterior tibial, lesser saphenous, and inferior gluteal veins. The retroperitoneal postcardinal vein gives rise to the greater saphenous vein, which gives off the femoral and posterior tibial veins." The axial artery in utero is paired with an axial venous network, which may provide a source of collateral venous outflow in the presence of deep venous obstruction. With ingrowth of the femoral vein, the axial vein normally involutes, with remnants persisting as the sciatic veins of the glutei and satellite vein of the sciatic nerve. In a manner analogous to the arterial system, failure of the femoral vein to develop, or the axial vein to involute, results in a PSV.

Morphologically, the PSA is complete if continuity between hypogastric and popliteal arteries is maintained. Arteriomegaly is frequently observed, with focal aneurysm formation in as many as 46% of patients. Acute lower limb ischemia, caused by thromboembolism, rupture, and "blue toe" syndrome are documented complications. Consequently, some investigators advocate duplex scan surveillance for all PSAs, with surgical intervention in the presence of aneurysm formation.

PSV is most commonly found in association with the Klippel-Trenaunay syndrome (KTS). Of these patients, 20% to 48% have a demonstrable PSV on magnetic resonance imaging. Of our patients, 20% to 48% have a demonstrable PSV on magnetic resonance imaging. Arteriomegaly may also occur in isolation and is a rare cause of recurrent posterior varicose veins. Although the sciatic artery and vein are paired in utero, their synchronous persistence is extremely rare. This has been reported only twice previously in the English-language literature as an incidental finding during a cadaveric study and work-up for resection of a malignant femoral nerve schwannoma. Our patient’s particular combination of congenital anomalies (ie, atypical varicosities, persistence of both sciatic vessels, and soft tissue hypertrophy) has not been previously reported.

The etiology of vascular anomalies is poorly understood. During embryogenesis, blood islands, containing angioblasts, are derived from the extraembryonic mesoderm. Vasculogenesis describes the differentiation of angioblasts into a primitive vascular plexus, and angiogenesis the remodelling and expansion of this preexisting network. The molecular basis for vascular morphogenesis is ill-defined but is likely to follow sequential changes in genetic expression. Regulatory factors implicated in these processes include vascular endothelial growth factor and its receptors, the angiopoietic system, and the ephrin-B system. The cause of our patient’s anomalies is not known. We postulate that a single genetic or environmental insult, occurring early during vascular morphogenesis, synchronously affected the development of arterial and venous systems of the lower limb. Similarly, the musculoskeletal system is also derived from mesoderm and this insult possibly may have also caused this patient’s soft tissue hypertrophy. Although the latter may be caused by hemodynamic change, we
believe these anomalies reflect a generalized mesodermal
defect.

In a young patient with atypical distribution varicose veins, diagnostic consideration should be given to other congenital anomalies, including KTS, Parkes Weber syndrome, and vascular malformations. KTS has been defined as “a rare congenital malformation characterised by a triad of (i) capillary malformation (usually a port-wine stain) (ii) atypical varicose veins - often with the persistence of embryological veins (iii) bone/soft tissue hypertrophy.”

In addition, abnormalities of the deep venous and lymphatic systems may be seen. These anomalies are variable, however, and some physicians will assign a diagnosis of KTS in the presence of two of these features. With the previous criteria, one could say that our patient has KTS with an associated PSA. Nevertheless, the patient lacks a capillary malformation, by far the most consistent feature of KTS and present in as many as 98% of patients. In addition, peripheral arterial anomalies are not a recognized feature of this syndrome. We believe our patient bares a close resemblance to the patients described by Madson et al with PSA, varicose veins, and limb length discrepancies. Whether this combination of mesodermal anomalies is an incidental finding or whether they represent a discrete clinical syndrome remains unclear. It remains plausible that, depending on the timing of the insult during embryologic development, any combination of vascular anomalies is possible. We simply wish to emphasize that atypical distribution varicose veins in young patients may be associated with persistence of both sciatic vessels. Because of the high incidence rate of sciatic artery aneurysm formation, we recommend formal duplex scan surveillance for these anomalies.

In our patient, valvar incompetence of the deep veins was the pathogenic cause of the chronic venous hypertension, with consequent SPJ incompetence and widespread reticular veins, telangiectaticas, and venous pigmentation. In the absence of a history of deep venous thrombosis, incompetence seems likely to be primary in origin, although whether this is the result of intrinsic weakness within the embryonic vein wall or valve cusp remains obscure. Excision of incompetent PSVs has been reported but was not feasible in this case because a hypoplastic SFV would not allow adequate collateral venous drainage. Although superficial venous surgery was not contraindicated, we believed that the presence of deep venous incompetence would greatly increase the risk of recurrent varicosities. Furthermore, SPJ ligation would not improve the appearance of many of the extensive reticular and telangiectatic veins in the thigh, which were nonshort saphenous in distribution. In view of these factors, the patient opted for conservative treatment in compression hosiery and remains well at 2 years. In the absence of focal aneurysm formation within the PSA, the patient will continue to undergo annual duplex scan surveillance.

In conclusion, we believe this case shows that persistence of the sciatic artery and vein may occur synchronously. We emphasize that atypical distribution varicose veins may be associated with persistence of the sciatic artery and recommend formal duplex scan assessment for these anomalies.

REFERENCES


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