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INTERVENTIONAL RADIOLOGY

CASE REPORT

Diffuse unilateral pediatric arteriopathy: successful treatment with repeated angioplasty

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ABSTRACT

We report a three-year-old girl exhibiting severe long-segment stenoses and occlusions with diffuse arterial involvement of the upper and lower extremities on the right side. The obstructive lesions, which caused atrophy of the right limb and chronic ulceration of the foot, were treated successfully with repeated percutaneous transluminal angioplasty. Obstructive peripheral arterial disease can cause growth retardation of the involved extremity, which can be salvaged by repeated endovascular therapy even in a small growing child.

Key words: • *percutaneous transluminal angioplasty* • *lower extremities* • *peripheral arterial disease* • *femoral artery* • *crural artery*

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The spectrum of pediatric vascular pathology differs from that in the adult population and varies greatly to include congenital and acquired disorders. Congenital arterial pathologies are not encountered frequently. These lesions represent a heterogeneous group of isolated or multiple abnormalities and are occasionally associated with vasculitis, rheumatoid disorders, or congenital syndromes.

Advances in catheter and wire technologies have made it increasingly possible for complex arterial interventions to be performed in children (1). Although several studies have established the efficacy of percutaneous transluminal angioplasty (PTA) in adults, there have been fewer and less extensive studies in children, and most have been limited to the renal and hepatic arteries. A literature search found few cases reporting other pathologies (2).

We present a pediatric patient who had an extensive congenital peripheral arterial disease of unknown origin that involved solely the extremities on the right side. We could not find a similar involvement pattern reported in the literature.

Case report

In July 2003, a three-year-old girl was referred in Başkent University Adana Medical Center due to short stature and thinning of her right leg in comparison with her left. Physical examination showed weak pulses on the right upper and lower extremities as compared with the left. There was no difference in size between the upper extremities. Color Doppler ultrasonography (CDUS) showed mild to moderate diffuse stenotic lesions of the brachial, ulnar, and radial arteries in the right arm and severe diffuse obstructive lesions of the popliteal, crural, dorsalis pedis, and posterior tibialis arteries in the right leg. Laboratory investigations showed heterozygous mutation of factor V Leiden. The patient had no coexisting genetic or acquired thrombophilic disorders.

The patient was subsequently referred to our interventional radiology unit for diagnostic angiography, with a presumptive diagnosis of congenital arterial disorder. Informed written consent for angiography was obtained from the child's parents. Following general anesthesia, a 4 F vascular sheath was placed in the left femoral artery. Digital subtraction angiography (DSA) of the pelvis and the arteries of the right lower extremity was performed. Angiography showed multiple diffuse stenoses of the distal popliteal, crural, and dorsalis pedis arteries. After reviewing the angiography images, PTA of the lesions was suggested, as this would have promoted growth of the extremity and prevented possible future occlusion. However, the parents declined the intervention at that time, and the patient was discharged on therapeutic levels of warfarin for anticoagulation. A specific diagnosis for the arterial involvement could not be reached, and a literature review did not reveal

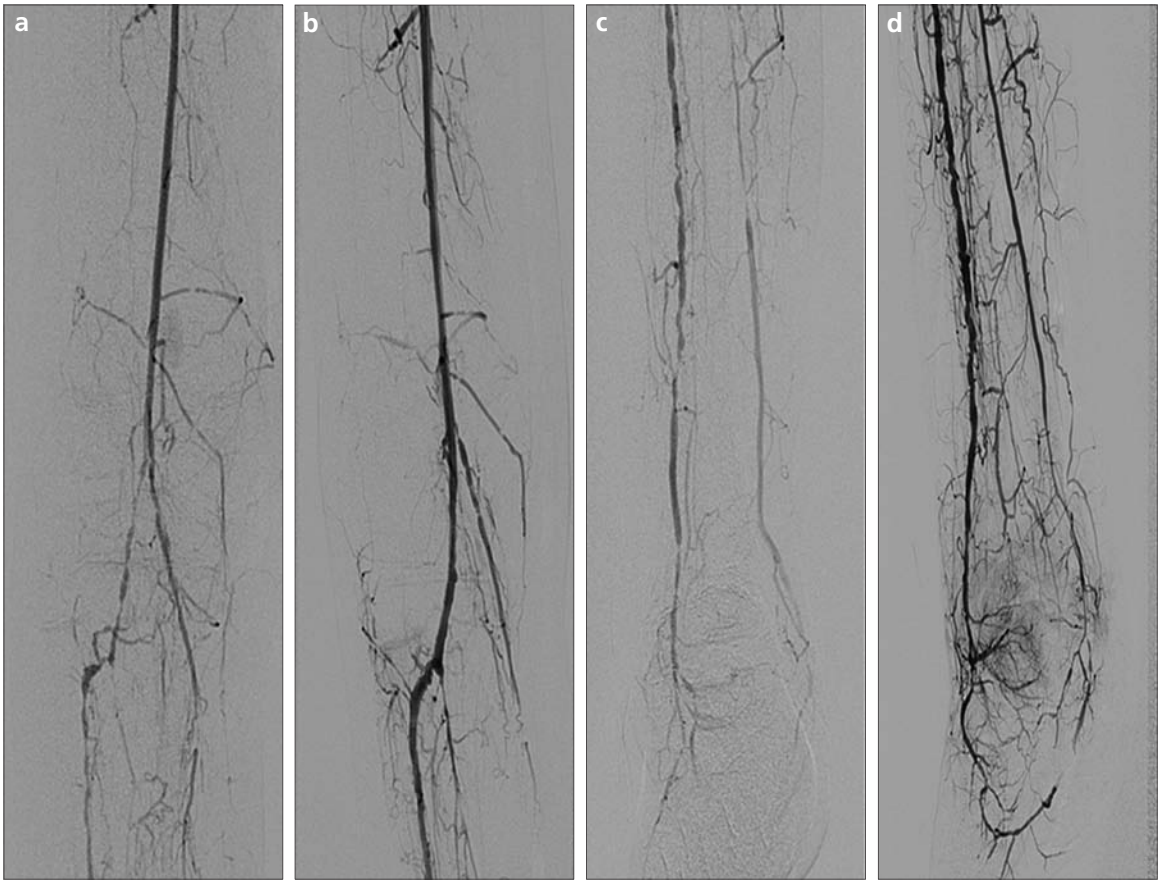

Figure. a–d. Digital subtraction angiography of the right lower arterial system (anteroposterior view) before treatment demonstrated multiple diffuse stenoses and occlusions of all crural arteries, including some muscular arteries **(a** and **c)**. After balloon angioplasty, the stenoses were successfully dilated, and blood flow was restored **(b** and **d)**.

any similar case reports. Although there was no family history of a similar vasculopathy, it was considered a congenital arterial obstructive disease involving the right-side upper and lower extremities.

The patient failed to present at follow-up and returned six years later complaining of pain in the right leg upon walking, significant shortening of the right leg compared with the left, and a large ulcer on the medial part of the right foot. It was discovered that the patient had undergone a technically successful angioplasty of the right lower extremity at another institution shortly after our initial evaluation. The intervention included balloon dilatation of the right limb arteries. Although the limb responded well with improved growth, no further intervention had been performed. This was despite symptom recurrence and increasing limb length discrepancy during the subsequent three years. An ulcer had appeared on the medial aspect of the right foot; topical treatment had been initiated, but no improvement in wound healing after six months was observed. Evaluation with CDUS revealed a diffuse long-segment stenosis and multiple occlusions all along the arteries of the right leg. Arterial flow in the ankle arteries was very weak, with an ankle brachial index of 0.25 on the right and 1.0 on the left. Angiography and PTA of the right lower limb arteries were planned. Informed consent for angiography and PTA was obtained from the child's parents.

After general anesthesia, the left femoral artery was punctured with a 21 G needle under CDUS guidance, and a 4 F vascular sheath was placed. With 4 F flush and diagnostic catheters, DSA of the pelvis and the right lower extremity arteries was performed. The lesions were diffuse and long. The arterial system proximal to the popliteal artery was near normal. The anterior tibial artery was patent with diffuse bead-like stenoses. The peroneal, posterior tibial, dorsalis pedis, medial, and lateral plantar arteries; plantar arterial arch; and lateral tarsal and arcuate arteries were occluded. The stenoses and occlusions were negotiated with straight or angled guide wires (Glide wire, Terumo, Tokyo, Japan), and a 0.018 control wire (Boston Scientific, Natick, Massachusetts, USA) was placed. After 2500 U (70 U/kg) intravenous heparin were administered, balloon dilatations were performed in the right popliteal artery with a 3.5-mm balloon, in the distal anterior and posterior tibial arteries with a 2-mm balloon, and in the proximal crural arteries with a 2.5-mm balloon (Symmetry, Boston Scientific, Galway, Ireland). Repeated dilatations were performed when there was more than 30% residual stenosis. We did not consider stent placement because of the young age of the patient, the presumed congenital etiology of the condition, and the appearance of the lesions. The procedure was successful, with restoration of near-normal flow in the dilated arteries. No complications occurred during or immediately after the treatment (Fig.).

Following six hours of bed rest, the patient was discharged the same day. A day after the treatment, pain on walking had resolved completely. The ulcer on the medial aspect of the foot started to improve within a week and entirely healed within a month. We planned three-month follow-up visits for CDUS and to repeat balloon dilatations whenever there is >50% stenosis of any artery of the leg until after puberty, at which time we predict that body growth will cease and obstructive lesions will not cause atrophy of the extremity. The patient has remained well during the first year of follow-up.

Discussion

Congenital obstructive arterial lesions of the extremities on one side of the body have not been reported before. The history of the patient and physical, laboratory, and angiography findings did not let us establish a diagnosis. This patient presumably has a congenital, non-atherosclerotic arterial disease of unknown origin. We observed the main involvement of the right crural arteries, characterized by diffuse obstructive, partially atherosclerotic and fibromuscular dysplasia-like lesions. Similar pathology is also seen in adults with fibromuscular dysplasia involving the lower limbs (3). However, the evaluation of DSA images did not demonstrate a "string of beads" pattern with multiple stenoses and aneurysmal or tubular dilatations, which are specific for the disease. Therefore, fibromuscular dysplasia was not considered in the differential diagnosis.

Major prenatal and neonatal causes of peripheral obstructive diseases, including TORCHES (toxoplasmosis, rubella, cytomegalovirus, and herpes simplex), varicella–zoster virus, HIV and parvovirus B19, drug use in pregnancy, intrauterine hypoxia, birth asphyxia, and chromosomal diseases, were excluded.

Laboratory investigations showed a heterozygous factor V Leiden mutation. The patient had no coexisting thrombophilic factors such as genetic or acquired thrombophilic disorders. Heterozygosity for factor V Leiden is relatively common and occurs in 3%–8% of the general population in the USA and Europe. Some authors have reported an effect of only a factor V Leiden mutation, even in heterozygous form, on arterial thrombotic events (4). Our patient had one factor V Leiden allele with no coexisting genetic disorder and no acquired or circumstantial thrombogenic risk factors; therefore, the clinical manifestation could not be due to the factor V Leiden mutation. In addition, the lesions were stenoses from intrinsic arterial wall disease, and not a consequence of thrombosis.

Regardless of the fact that we were unable to find a similar case with the exact etiology in the literature, our patient's clinical and angiographic findings showed some pathophysiological similarities with other diseases. These include congenital supravalvular aortic stenosis, which results in obstructive arteriopathy of varying severity, most prominently at the aortic sinutubular junction, associated with stenoses of systemic and pulmonary arteries (5); mucopolysaccharidosis I, characterized by involvement of the abdominal aorta, the occlusion of the lumbar arteries, renal arteries, and major visceral arteries, including the celiac trunk, superior, and inferior mesenteric arteries, and the narrowing of the iliac arteries (6); giant cell arteritis, which is clinically defined by upper limb involvement (7); and digital arterial occlusion associated with parvovirus B19 (8).

Obstructive involvement in the peripheral arteries of pediatric patients is very rare. To our knowledge, unilateral arteriopathy of the extremities has not been previously reported. Endovascular techniques may be a valuable therapeutic option for such vascular lesions. This continued to resolve the patient's symptoms as they recurred with growth. These recurrences may reflect non-developing parts of the body in a developing child or types of growth failure in the vessel lumen. Repeated interventions can be attempted to promote growth of the extremity without hesitation until the patient reaches adolescence.

Conflict of interest disclosure

The authors declared no conflict of interest.

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