 Peripheral Vascular Malformation (Servelle-Martorell)

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A 38-year-old man from India presented for evaluation of a painful vascular malformation of the right hand and forearm that limited the use of his right hand (Figure 1). The malformation had existed since childhood. Other parts of the body showed no abnormalities. Several operations (details not known) had been performed in India without success. The vascular malformation caused a soft-tissue swelling characteristic of venous ectasias. The right arm and hand were slightly shorter than the left arm. The peripheral pulses were symmetrically palpable, and no bruits or thrills indicative of arteriovenous shunts were found. No temperature difference was observed. Radiographs showed hypotrophy of the bone, soft-tissue swelling, and multiple opacities consistent with phleboliths in the affected hand (Figure 2). Further diagnostic procedures were denied.

The history and clinical presentation in this case are diagnostic of an angio-osteohypotrophic syndrome known as Servelle-Martorell angiodysplasia. This syndrome describes the association of venous and rarely, arterial malformations with skeletal abnormalities. In the deep venous system, an abnormal vein location, partial or complete lack of valves, and/or venous hypoplasia or aplasia can be observed. The ectasia and aneurysmal dilatation of the superficial veins may result in a monstrous deformity of the extremity. Intraosseous vascular malformations may lead to hypotrophy of bone with destruction of spongiosa and cortical bone, resulting in shortening of the limb. Intraosseous vascular ectasias can induce cystic deformation of the medullary cavity and thinning of the cortex, which may result in joint destruction. Multiple phleboliths can be found in venous ectasias on the radiograph. The prognosis of this disorder is uncertain. Therapy is predominantly conservative. In the presence of aneurysmal complications or severe shunting, surgery may be indicated.

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Circulation encourages readers to submit cardiovascular images to Dr Hugh A. McAllister, Jr, St Luke’s Episcopal Hospital and Texas Heart Institute, 6720 Bertner Ave, MC1-267, Houston, TX 77030.

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Figure 1. Right hand: extensive deformity caused by venous ectasias.

Figure 2. Right hand: hypotrophy of bone with shortening of the phalanges, cystic deformation, soft tissue swelling corresponding to venous malformations, and multiple phleboliths. Left hand: normal findings.
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