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.
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.
Peripheral Vascular Malformation (Servelle-Martorell)
Thomas Weiss, Ulrich Mädler, Heike Oberwittler, Birgit Kahle, Claus Weiss and Wolfgang Kübler

_Circulation_. 2000;101:e82-e83
doi: 10.1161/01.CIR.101.7.e82

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2000 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/101/7/e82

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org/subscriptions/