OBJECTIVE. Our purpose was to identify the characteristic imaging features of cystic adventitial disease of the peripheral arteries.

CONCLUSION. Patients with cystic adventitial disease of the peripheral arteries often present for evaluation of soft-tissue masses involving the extremities. Noninvasive imaging reveals a characteristic appearance allowing confident diagnosis.

Cystic adventitial disease is an uncommon vascular disorder characterized by cystic degeneration of a peripheral artery. Any peripheral artery can be affected; however, the disorder has a striking proclivity for the popliteal artery and typically affects young to middle-aged men without evidence of atherosclerosis or other systemic vascular disease [1]. Affected patients typically present with sudden onset of rapidly progressive calf claudication and lower extremity pain. Physical examination often shows a soft-tissue mass, and, in a younger patient with no evidence of atherosclerosis, clinicians often focus on the soft-tissue mass, suspecting a soft-tissue sarcoma or a popliteal cyst rather than a vascular cause.

We detail our experience with seven patients with cystic adventitial disease, of whom five were referred for evaluation of a soft-tissue mass and two, for evaluation of unexplained lower extremity pain. We retrospectively reviewed imaging characteristics of this lesion to identify those features that would allow an accurate prospective diagnosis.

Materials and Methods

We retrospectively reviewed the imaging studies of seven patients with surgically and pathologically confirmed cystic adventitial disease of the peripheral arteries. The study group consisted of six men and one woman. Ages ranged from 38 to 79 years (mean, 53 years). MR imaging was performed in all patients, with IV gadolinium-enhanced MR imaging in four patients. MR imaging included spin-echo T1-weighted images (n = 6) and either conventional spin-echo dual-echo or short tau inversion recovery sequences (n = 7). Conventional angiography was performed in six patients, with three of these patients also having MR angiography. MR angiography was performed with a two-dimensional time-of-flight technique in two patients and three-dimensional contrast-enhanced MR angiography in one patient. Sonography was performed in four patients, including intraarterial sonography in one patient. Sonographically guided aspiration was performed in one patient. Conventional radiographs were available in two patients. Five patients presented for evaluation of soft-tissue masses discovered at physical examination. Two patients presented for unexplained lower extremity pain.

Radiographs were evaluated for the presence of a mass, calcification, or bone erosion. The MR images were analyzed for signal intensity, signal homogeneity, enhancement pattern, lesion size, and location. Skeletal muscle and fat were used as the reference tissue for all MR imaging sequences. Sonograms were evaluated for lesion echogenicity and Doppler flow.
Results

All cases showed involvement of a single peripheral artery. Lesions were located in the popliteal (n = 6) and radial arteries (n = 1). No patients showed evidence of diffuse atherosclerosis or other generalized arterial disease. Length of arterial involvement varied from 1.7 to 11.2 cm (mean, 4.6 cm). Nonocclusive disease was seen in three patients, and segmental occlusion in four. The length of occlusion ranged from 1.7 to 11.2 cm (mean, 5.4 cm).

Radiographs failed to show the lesions in both cases. Subtle soft-tissue prominence was seen in a single case (Fig. 1). No arterial vascular calcification suggesting atherosclerosis was present in any patient.

MR imaging showed aggregates of multiple small round or oval masses originating in the wall of the involved peripheral artery, with larger lesions having a multiloculated appearance. The individual intramural masses showed homogenous low signal intensity on T1-weighted MR images and high signal intensity on T2-weighted or fluid-sensitive sequences, consistent with a cystlike character (Fig. 2). After IV gadolinium administration, no enhancement was seen in three patients (Fig. 3). Mild peripheral enhancement was seen in one patient. There was resultant compromise of the arterial lumen by the multiloculated cystic masses, which varied in degree from minimal eccentric indentation of the arterial lumen, to more prominent crescentic stenosis, to complete occlusion. MR angiography in three patients showed well the degree of luminal compromise and the extrinsic, intramural nature of the stenosis (Fig. 3). In two patients in whom MR angiography showed occlusion, prominent collaterals were seen with reconstitution of the distal arteries. No significant vascular disease was seen proximal or distal to the lesions. Correlation with MR angiography and conventional angiography was available in two patients and was excellent.

Conventional angiography showed smooth, tapered stenoses with curvilinear or spiral narrowing of the lumina of the involved vessels (Fig. 2). The stenoses showed rounded, spiral, or hourglass morphologies with extrinsic compression. Nonocclusive segmental narrowing was seen in three patients with no significant poststenotic dilatation. Arterial occlusion was seen in four patients with formation of collaterals that reconstituted the distal arteries. Concomitant atherosclerotic changes were absent.

Sonography showed multilobulated collections of small rounded or oval, anechoic or hypoechoic masses arising in the walls of the affected arteries (Fig. 1). Crescentic indentation on the anechoic lumina of the vessels was seen with eccentric narrowing or occlusion by the cystic masses. Low-level echoes were seen in several masses indicative of debris in the contents of the cysts. Duplex Doppler sonography confirmed the absence of flow in the cystlike lesions and extrinsic narrowing of the affected arteries. Sonographically guided cyst aspiration was performed in one patient with aspiration of thick gelatinous mucinous material (Fig. 3). Intraarterial sonography showed hypoechoic masses in the wall of the popliteal artery and crescentic narrowing of the lumen (Fig. 4).

Discussion

Cystic adventitial disease is a relatively rare vascular condition with the typical patient a middle-aged man complaining of rapidly progressing claudication-like symptoms [2]. Five (71%) of our patients presented for the evaluation of a soft-tissue mass, a presentation that, to our knowledge, has not been previously emphasized. All patients had intermittent extremity pain; however, the mass was the dominant clinical finding and became the focus of the examination. The cause of cystic adventitial disease is controversial. First reported in 1954 by Ejrup and Hierton [3], cystic adventitial disease was originally described as a hematoma of the arterial wall [4]. Further studies eventually elucidated the true nature of the lesions identifying the cyst contents as grossly resembling thick mucinous gel containing varying combinations of mucopolysaccharides, hyaluronic acid, and hydroxyproline [5, 6]. The progression of cystic adventitial disease is slow, occurring over a period of several years [7], accounting for the large size and masslike configuration of many lesions, and also explaining the clinical confusion with a slowly growing soft-tissue tumor.

The MR imaging appearance of cystic adventitial disease is quite characteristic, showing multiple arterial intramural cystlike masses. The individual lesions typically show homogenous low signal intensity on T1-weighted spin-echo MR images and high signal intensity on fluid-sensitive sequences (Fig. 2). The lesions are oriented along the long axis of the vessel, and this orientation, as well as the intramural location, is well shown on high-spatial-resolution multiplanar MR imaging. Large lesions form conglomerate multilobulated masses; however, these features are still present in the smaller individual components. Individual lesions showed no enhancement or minimal marginal enhancement (Fig. 3). However, we suspect that because the basic pathophysiology of the disease process entails a myxoid degeneration in the adventitia of the vessel...
Imaging of Cystic Adventitial Disease

Although angiography has been regarded the gold standard for the diagnosis of cystic adventitial disease, we found MR angiography to be equally diagnostic (Figs. 2 and 3). Both techniques showed with exquisite detail the extrinsic compression of the affected vessel with absence of significant disease proximal and distal to the lesion. They also showed the hourglass configuration that is seen with circumferential involvement and the crescentic margins in cases of complete occlusion. MR angiography accurately showed the degree and length of stenosis or occlusion as well as the lack of vascular abnormality elsewhere in the affected extremity. In our opinion, MR angiography equaled conventional angiography for both diagnosis and surgical planning, with the obvious advantage of a noninvasive evaluation.

Sonography is also useful in showing the characteristic appearance of cystic adventitial disease. Sonography reveals the anechoic or hypoechoic masses originating in the arterial wall and can show the degree of associated vascular compromise (Fig. 1). Sonography typically reveals the lesions as multilobulated masses composed of multiple smaller cysts. Low-level echoes may be seen in the cysts, indicating the gelatinous nature of the contents of the cyst. Color and duplex Doppler evaluation of the arterial lumen is useful to depict the extent of the associated arterial stenosis or complete occlusion of the affected artery. Intraarterial sonography was performed in one patient in our series, precisely showing the cystic masses arising from the adventitia of the arterial wall.

In conclusion, cystic adventitial disease is an unusual vascular disorder that may present clinically as a slowly growing soft-tissue mass. MR imaging and MR angiography are both characteristic, with the former showing the multiple small intramural cystlike lesions in the arterial wall and the latter the eccentric, scalloped stenosis and degree of vascular compromise. Sonography also shows the lesions well and depicts the degree of arterial stenosis.

Fig. 2.—61-year-old man with cystic adventitial disease of popliteal artery who presented with unexplained lower extremity pain and soft-tissue fullness in popliteal fossa. A, Sagittal T2-weighted MR image of knee shows multiple high-signal-intensity adventitial cysts (black asterisk) arising from wall of popliteal artery with extrinsic compression of arterial lumen (white asterisk). B, Angiogram shows focal extrinsic narrowing of arterial lumen of popliteal artery (arrow) typical of cystic adventitial disease. C, Intraoperative photograph of popliteal artery reveals focal bluish-colored mass (arrow) arising from wall of artery in region of previously noted stenosis on arteriography. D, Intraoperative photograph of dissected arterial wall shows focal nodular masses arising from it (arrow). E, Microscopic evaluation reveals areas of cystic degeneration (asterisks) in adventitia of popliteal artery.
Fig. 3.—78-year-old man with popliteal soft-tissue mass.
A, MR angiogram of popliteal artery shows occlusion of segments of artery. Margins are rounded and crescentic, consistent with extrinsic nature of occlusion (arrows). Collateral vessels are seen with distal reconstitution of artery.
B, Enhanced T1-weighted sagittal image of knee with fat saturation shows multiple cystic masses arising from wall of popliteal artery consistent with cystic adventitial disease (arrows). Popliteal vein is seen posteriorly (arrowheads).
C, Enhanced T1-weighted axial image of knee depicts adventitial cyst (asterisk) with complete obliteration of arterial lumen at this level.
D, Sonogram shows sonographically guided cyst aspiration. Eighteen-gauge spinal needle (arrow) is visualized with distal tip in adventitial cyst (asterisks) arising from popliteal artery.
E, Photograph of gross specimen shows thick, mucinous, gelatinous material that was aspirated from adventitial cyst.
compromise. Although conventional angiography continues to be the gold standard, noninvasive MR angiography is diagnostic. Radiologists should be familiar with the entity and be well acquainted with its clinical presentation and diagnostic radiologic appearance.

References