

Two cases of hemangiopericytoma (angiographic diagnosis)

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ABSTRACT — The authors present two cases of hemangiopericytoma in which angiography showed the typical findings of this rare vascular tumor. The angiographic picture, the clinical data and the anatomopathological and histological findings are discussed.

INDEX TERMS — Hemangiopericytoma - Diagnosis - Tumors - Arteriography.

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INTRODUCTION

Hemangiopericytoma is a rare mesenchymal tumor (1% of all vascular neoplasias). It was first described in a separate classification by Stout and Murray in 1942 (14) and is so called due to its origin in the pericytes of the capillary vessels.

The absence of clinical symptoms often makes early diagnosis difficult even though the neoplasia is highly malignant.

The angiographic examination providing characteristic findings may help in suggesting early radical surgery.

CASE I

A 25-year-old man was hospitalized for a mass in the right thigh which had appeared about a month earlier, moderately painful on

palpation and after protracted strain.

He had been hospitalized elsewhere six years before for an endocranial compressive syndrome. On that occasion a neoplastic mass localized in the small wing of the left sphenoid was removed: it was histologically interpreted as a meningioma. Four years later, owing to the recurrence of neurologic symptoms, the patient was submitted to a CT scan. It showed a space-occupying recurrence in the same site and the histologic diagnosis of the removed specimen was hemangiopericytoma.

The patient, submitted subsequently to local ^{60}Co treatment, was in good health until noting a mass in the right leg.

Physical examination on admission to our Institute revealed a mass which was hard and moderately painful on palpation. The mass was as big as an orange and felt detached from the skin but attached to the adductor muscles of the right thigh.

While the hematochemical findings were normal, X-ray examination of the chest revealed two nodular parenchymal images, at the left base and in the right subclavicular

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area with radiologic features of metastatic nodules.

Right femoral arteriography showed at the site of the clinically visible mass a rounded basket-like arterial branch vascularization originating from muscular collaterals of the superficial femoral artery (Fig. 1); subsequently a thick arteriolar circle was locally seen (Fig. 2); with strong and persistent staining and early venous drainage (Fig. 3).

An apparently capsulated neoformation was surgically removed from the vastus medialis. It was 5 cm in size, weighed 40g, was of hard-elastic consistency, smooth surface, red-grayish in color. On the cut surface it was made up of homogeneous tissue of a yellow-pinkish color. Histologic examination proved it to be a highly mitotic angiosarcoma of the hemangiopericytoma type.

CASE II

A 22-year-old woman was hospitalized in our Institute for recurrent angiomas in the left leg. Past history showed at five years of age pain in the left leg particularly in the popliteal cavity and after stress. The patient underwent repeated surgical removals of angiomatous formations in the popliteal cavity, in the left lateral and medial gastrocnemius from the age of 6 to 16 years.

After a four year remission the pain in the limb reappeared, associated first with edema after prolonged stress but even at rest.

The physical findings revealed a small mass the size of a hazel-nut in the medial malleolar area, having a hard-elastic consistency, detached on deep touch, and slightly painful on palpation.

The hematochemical findings were normal.

A left femoral arteriography was performed. It showed a hypervascularized formation in the malleolar area with the same features as in the first case. A smaller

neoformation appeared in the muscular masses of the calf (fig. 4-5).

A subcutaneous nodule of 2,0x1,5x0,7 cm was surgically removed. Apparently capsulated, it was a reddish-violet mass with a vascular peduncle. It presented on the surface a non-homogeneous red-grayish tissue, histologically diagnosed as a hemangiopericytoma without atypical cells.

The second lesion, most likely a recurrence, shown by angiography in the calf, was removed at another institution after 2 years, with a histologic diagnosis of hemangiopericytoma.

No local recurrences appeared on arteriography performed on the last admission 5 years later.

DISCUSSION

Hemangiopericytomas arise in the pericytes, which are contractile cells wrapped around the capillaries, described by Zimmermann in 1922 (16) and which seem to regulate the capillary blood flow.

Due to their origin, hemangiopericytomas may be located simultaneously in many places, as can be seen in Table I. They are

TABLE I
Hemangiopericytoma sites
(by Pitluk and Conn 1979)

Site	Number	%
Lower limbs	63	36.4
Pelvis and retroperitoneum	37	21.4
Head and neck	23	13.3
Upper limbs	23	13.3
Trunk	21	12.1
Paravertebral	6	3.5
Total	173	

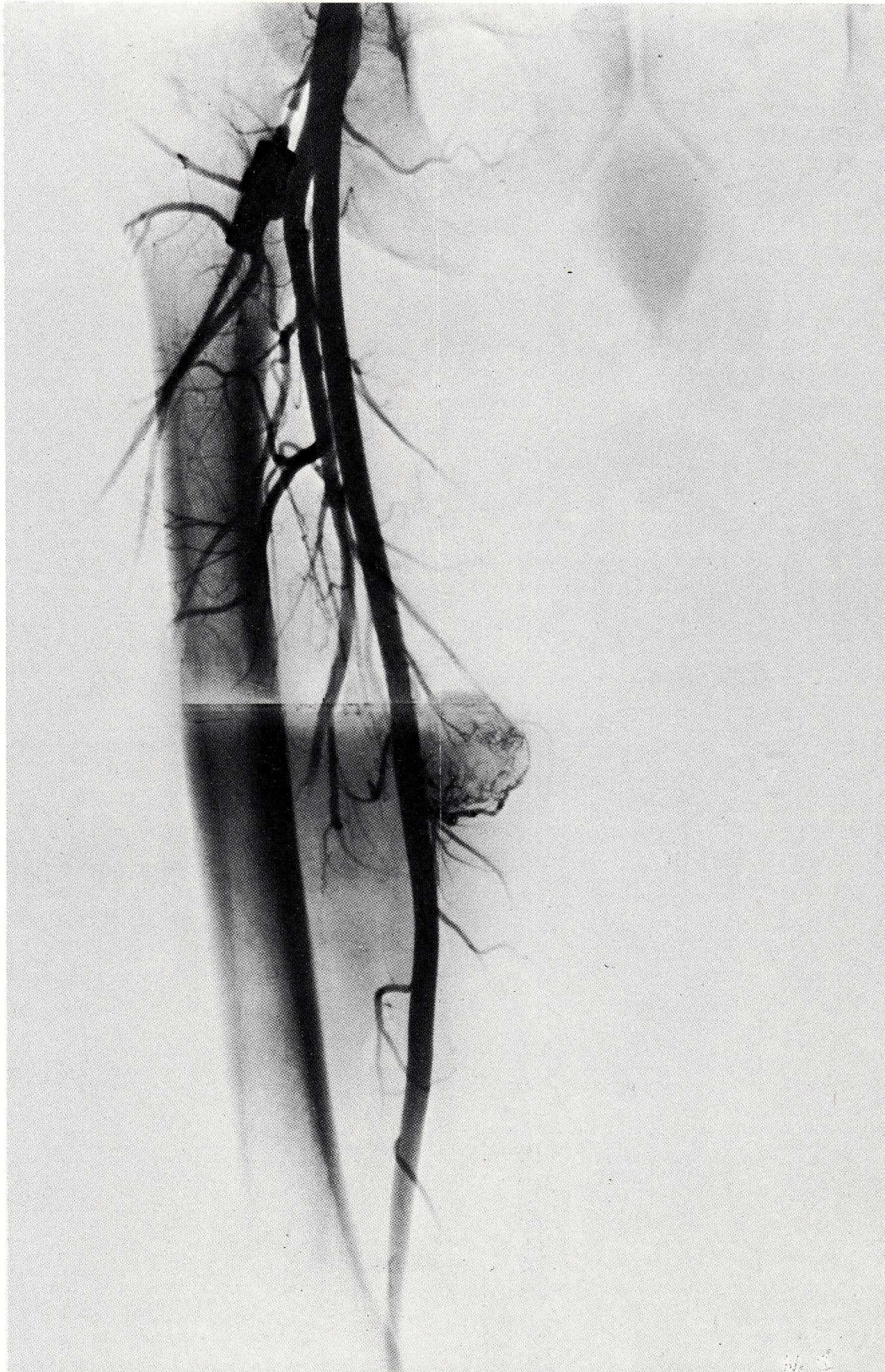


Fig. 1 - Right femoral arteriography. Early phase: basket-like branches enclosing the neof ormation.

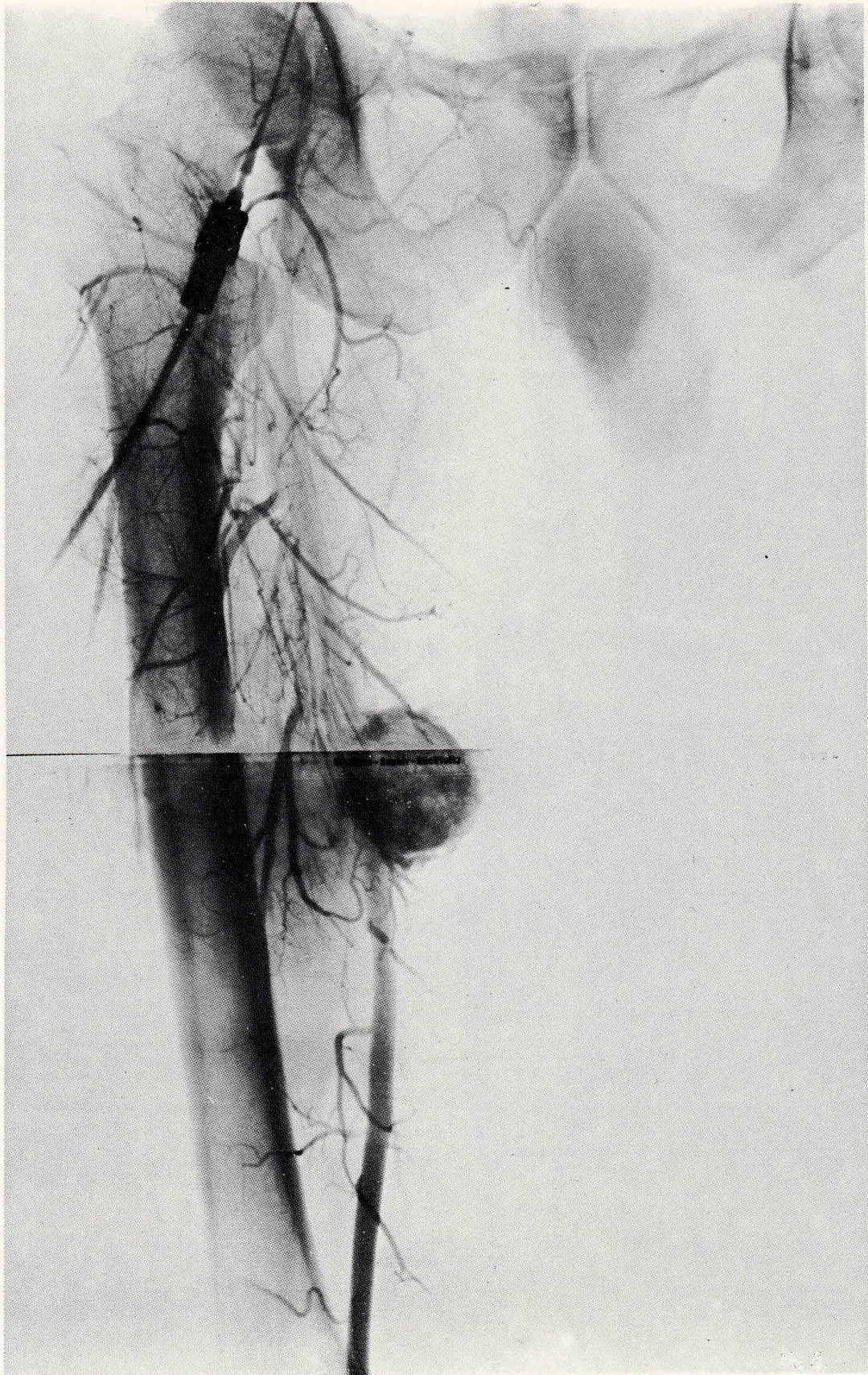


Fig. 2 - Arteriolar phase: thick arteriolar circle in the area of the mass.

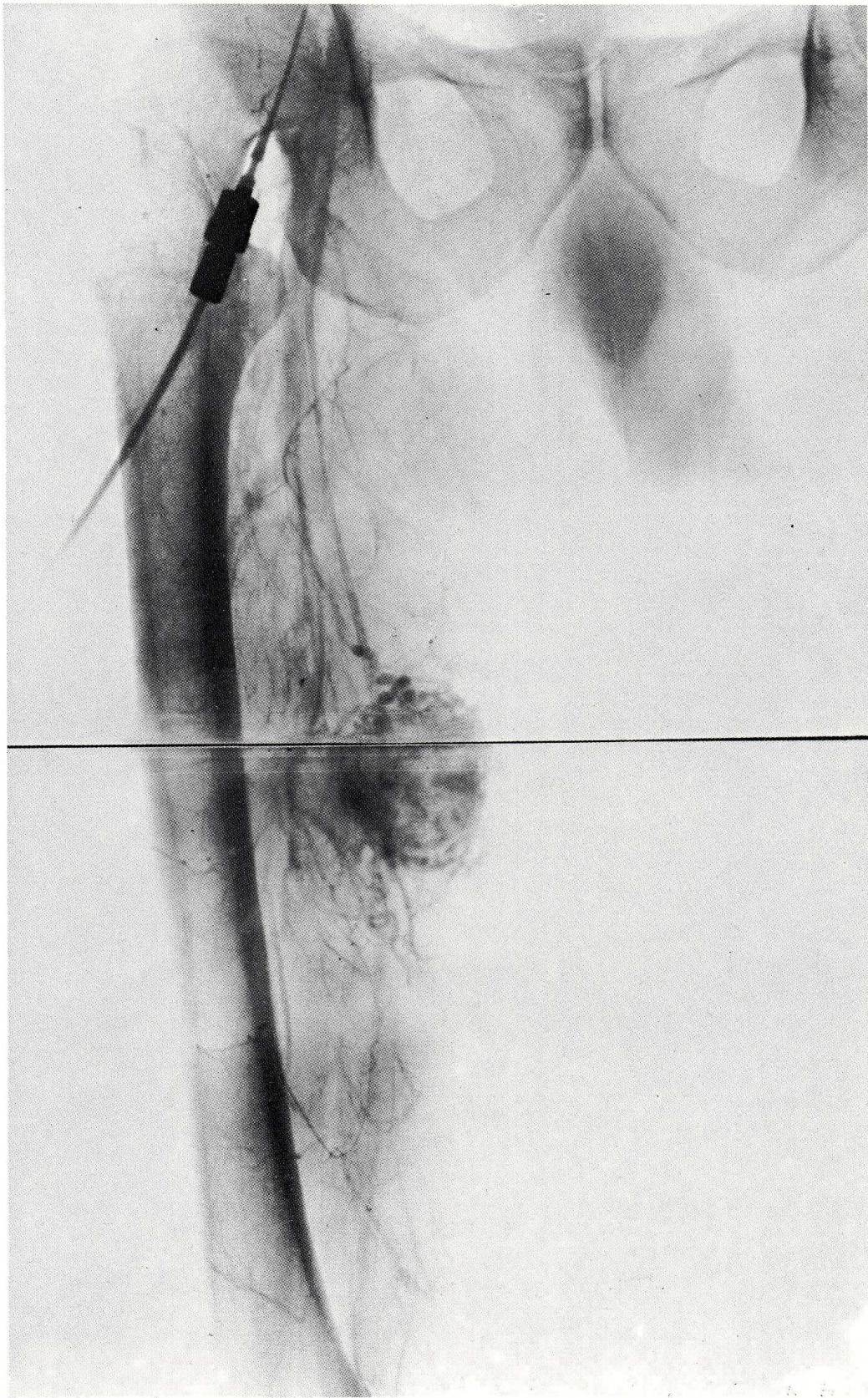


Fig. 3 - Late phase: thick, homogeneous and persistent staining of the mass which is clearly defined. The early venous drainage is evident (femoral and iliac vein).

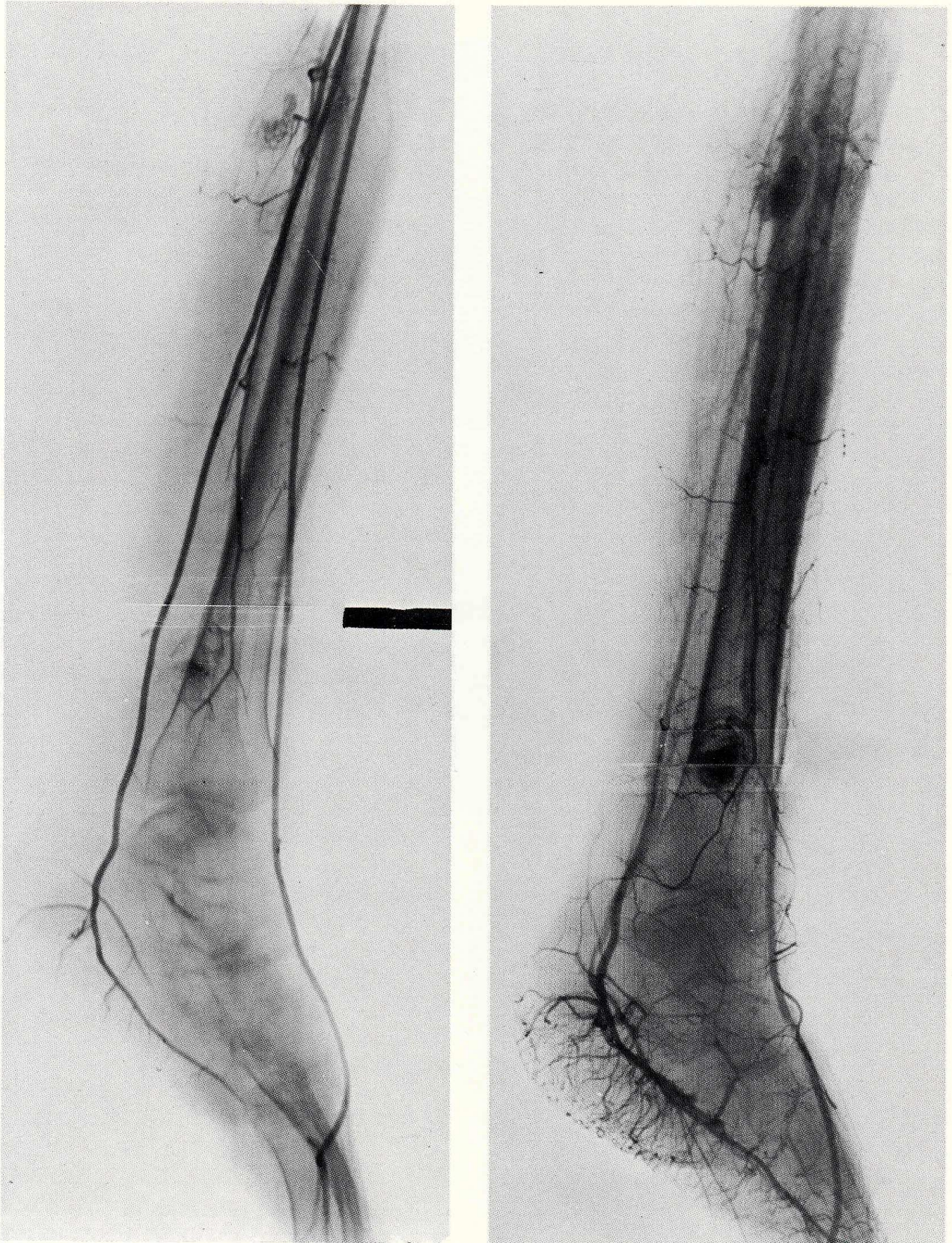


Fig. 4, 5 - Femoral arteriography: in the early arterial phase (Fig. 4) and in the later one (Fig. 5) two small hypervascularized lesions are seen. They are well-defined and have thick staining in the supramalleolar area and in the calf.
No early venous drainage.

rather rare: 1% of all vascular neoplasias. The literature up to 1978 reports 356 cases (17).

The incidence, regardless of age, is the same in the two sexes, though endocranial localizations are more frequent in the young age due to the generally early onset of compression symptoms (11).

Features of hemangiopericytomas are slow growth and high percentage of malignant forms with distant metastases (about 50% of cases) (14). Size of the neoplasm (diameter more than 5 cm), histologic findings of anaplasia, elevated number of mitoses, and small areas of necrosis are indicative of malignancy (4, 12).

However, the diagnosis of malignancy is still difficult: it is more often confirmed by distant metastases affecting mainly lungs, brain, intestine and skeleton (13).

Another feature of the neoplasm is the high incidence of local recurrences ranging from 25 to 50% in various case series (12, 13, 14).

The histologic pattern is characterized by proliferation of the typical spindle cells of pericytes lined up around the capillaries.

Clinically hemangiopericytomas appear as round, well-defined, hard neoformations which are generally asymptomatic. The possible symptoms are due to compression of the adjoining structures.

ANGIOGRAPHIC PICTURE

In the few cases subjected to angiographic examination, described in the literature (25 up to 1978) (1, 5, 8, 9, 10, 15, 17) a characteristic pattern was observed. It seemed to differentiate hemangiopericytoma from all other vascular tumors and confirms our data. In the early arterial phase an arterial peduncle is seen which divides into arch-shaped branches surrounding the mass. In the arteriolar phase the branches enclose a thick network of vessels giving rise in the

capillary phase to an area of homogeneous, thick and persistent staining.

At times, as in the case observed by us, early venous drainage occurs.

The picture is substantially the same in all the cases reported in the literature, independently of location, size and clinical evolution of the neoplasm, except for two cases where the lesion was situated within the chest: one was hypovascular and the other avascular (6, 15). The angiographic examination does not allow a reliable differential diagnosis between benign and malignant forms, which will be provided by histologic examination and by the clinical course.

Surgical removal should be extensive and radical, as the tumor is not properly capsulated, although it is generally well-defined because of atrophy of adjacent tissues due to compression (2, 12, 13, 14).

In one of the patients observed by us, the primary endocranial localization was probably metastatic, considering the size of the neoplasia removed from the thigh and the typical slow growth. The simultaneous presence of lung metastases suggests a highly malignant form.

CONCLUSIONS

The cases described have the typical features of hemangiopericytoma. It is important to stress the few clinical symptoms except in endocranial forms and the limited local aggressiveness of the neoplasia contrasting with the frequency of distant metastases. The angiographic pattern, although it is specific, does not allow a differential diagnosis of benignity or malignancy, which should rather be based on the histologic findings.

The diagnosis of malignancy requires antineoplastic chemotherapy or radiotherapy, which seem, however, scarcely to affect the course of the tumor (3, 7, 12).

RIASSUNTO

Gli Autori presentano due casi di emangiopericitoma in cui l'esame angiografico ha evidenziato reperti caratteristici di questo raro tumore vascolare.

Vengono discussi il quadro angiografico, i dati clinici ed i reperti anatomopatologici ed istologici riscontrati.

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