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Case Report: Malignant Thigh Hemangiopericytoma With Thoracic Wall Metastasis

M. Tayfun Altınok*, Tamer Baysal*, Ramazan Kutlu*, Ahmet Sığırcı*,
Alpay Alkan*, Ömer Soysal**, Bülent Mızrak***

* Inonu University Medical Faculty, Department of Radiology, Malatya, Turkey
** Inonu University Medical Faculty, Department of Thoracic and Cardiovascular Surgery, Malatya, Turkey
*** Inonu University Medical Faculty, Department of Pathology, Malatya, Turkey

A malignant soft tissue hemangiopericytoma metastatic to the thoracic wall in a 65 year old man is reported. Although the MRI of both the primary and metastatic foci revealed complementary finding of a network of fine vessels, only the histologic examination of the specimens confirmed the malignant features of the tumor. Both the primary and metastatic foci were totally resected. The patient is alive without local recurrence and metastasis for 10 months after the operation.

Key words: Hemangiopericytoma, Metastasis, Thigh, Thoracic Wall, MRI

Olgu Sunumu: Toraks Duvarına Metastaz Gösteren Malign Yumuşak Doku Hemanjioperisitioması

65 yaşında erkek hastada toraks duvarına metastaz gösteren malign yumuşak doku hemanjioperisitioması rapor edildi. Primer ve metastatik odakların MRI incelemesinin ince damar ağını göstermesine rağmen sadece histolojik inceleme tümörün malign özelliklerini ortaya çıkardı. Primer ve metastatik odaklar tamamen çıkarıldı. Operasyondan 10 ay sonra lokal nüks veya metastaz yoktu.

Anahtar Kelimeler: Hemanjioperisitioma, Metastaz, Uyluk, Toraks Duvarı, MRI

CASE

A 65 year old man admitted to our institution with complaints of local swelling located both in the medial part of the right thigh and left lower thorax. The patient had a slow growing lesion in the thigh for almost 3 years and he noticed a thoracic wall lesion 3 months ago. Physical examination revealed that both of these lesions were mobile and painless. The lesion located in the medial part of the thigh was approximately 15x15 cm and the thoracic wall lesion was 10x15 cm in dimensions. Although both lesions were easily palpable, the margins were indefinite.

X-rays of the thigh revealed soft tissue swelling without calcification in the right thigh. CT evaluation of the lung and mediostinum was normal. On MRI imaging, the inner thigh lesion was demonstrated to be a soft tissue mass with flow voids in and around the lesion without cystic necrosis. It was located between the vastus medialis muscle and the skin, with definite margins. The lesion had intermediate signal intensity on T1 weighted images and was hyperintense on T2 weighted images. The left lower thoracic lesion was located to the anterolateral thoracic wall extending into the abdomen with any sign of invasion to the neighboring structures (Fig 1.a.b) Both lesions revealed homogeneous contrast enhancement.

Angiography demonstrated that both lesions had a network of fine vessels and the thigh lesion was fed by the vessels originating from the right superficial femoral artery (Fig.2)

The encapsulated thigh mass was resected totally and the mass in the thoracic wall was resected with one fourth of the diaphragm and two ribs (9. and 10. ribs). The diaphragm was sutured to the thoracic wall primarily and the thoracic wall defect was also closed with the surrounding tissues primarily. Histologic examination of both two

Figure 1a. MR imaging of the thigh lesion. T1 weighted images revealing that the lesion had intermediate signal intensity. The soft tissue mass was located between the vastus medialis muscle and the skin, with definite margins. There are flow voids in and around the mass which was hyperintense on T2 weighted images.

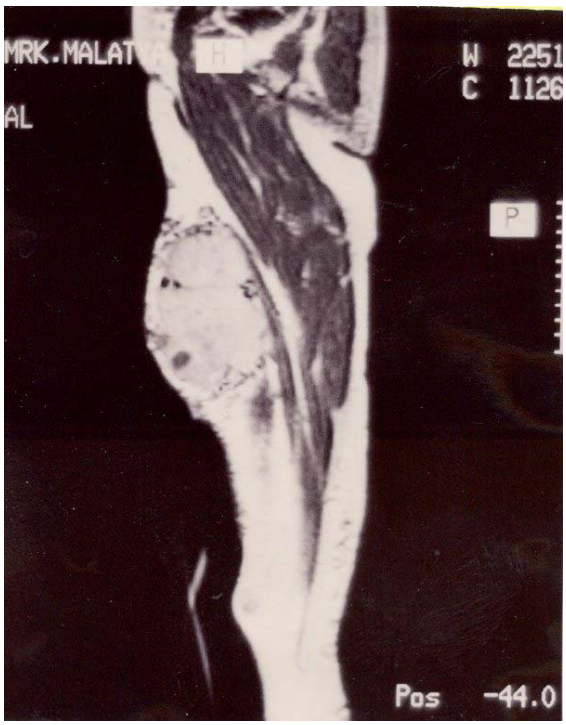
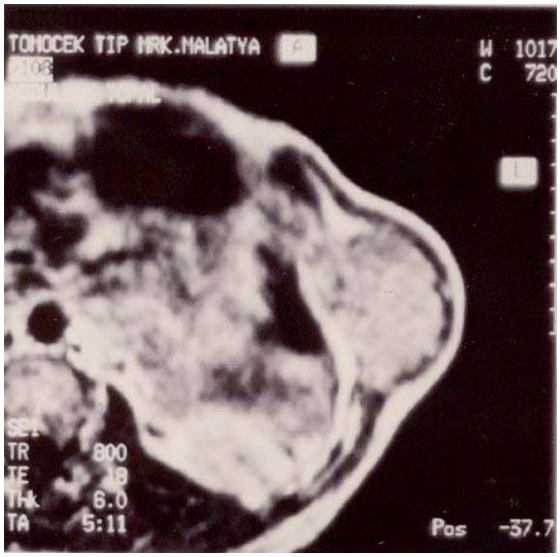


Figure 1b. T1 weighted images demonstrating that the left lower thoracic mass had intermediate signal intensity with flow voids. It was located to the anterolateral thoracic wall extending into the abdomen with any sign of invasion to the neighboring structures.



masses revealed that all tissue specimens had similar morphologic features. In large areas, compressed vascular channels were surrounded by neoplastic cells having oval to fusiform nuclei with Hematoxylin-eosin. A reticulin stain disclosed the fine reticulin network surrounding each tumor cells. The result of the thigh lesion was hemangiopericytoma (HP) with malignant features. The thoracic wall lesion was reported to be the metastasis of thigh HP (Fig.3). The patient is alive without local recurrence and metastasis for 10 months after the operation.

Figure 2. Femoral angiography. The thigh mass had a network of fine vessels and was fed by the vessels originating from the right superficial femoral artery.

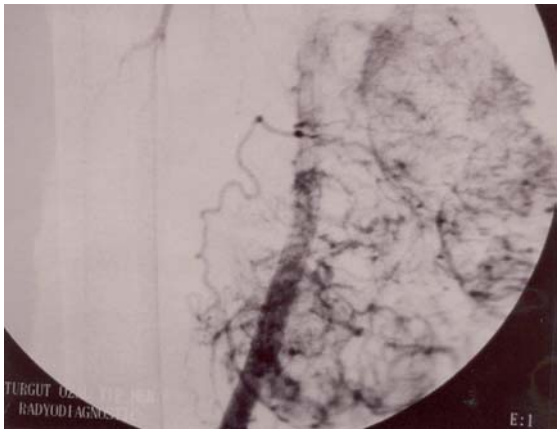
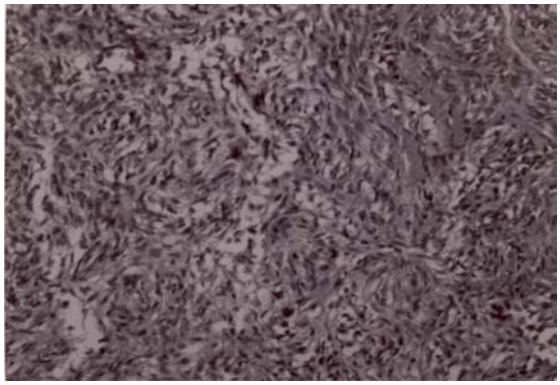


Figure 3. Histologic section of the primary tumor. Compressed vascular channels were surrounded by neoplastic cells having oval to fusiform nuclei (Hematoxylin-eosin, x 50)



DISCUSSION

HP is a rare vascular tumor originating from pericytes, the perivascular cells that are found in place of smooth muscle cells in the wall of the terminal arteriole as it becomes the precapillary and also around capillaries, external to the basement

membrane.[1-3] HP is reported to originate wherever there are capillaries.[4]

HP occurs in all age groups with a peak in the fifth and sixth decades. The sex distribution is reported to be equal (1-2). Its preferred localization is the lower limb (particularly the thigh), followed by the retroperitoneum and the pelvic region, and by the upper limb, trunk, head and neck. Most of the HP mentioned in the literature arise from soft tissues.[6] It is nearly always deeply located, within and between the muscles. The tumor, which is deep, grows slowly and the main symptom in HP of soft tissues is a painless mass.[1,2,7] The duration of symptoms prior to diagnosis may be months or years.[1,2] In our case, HP was located in the thigh and originated from soft tissue in accordance with literature data. However, it was more superficially located when compared to literature.

The radiographic picture of the tumor is not typical. MR imaging generally demonstrates a hypervascular tumor with smooth margins. It has slight intensity relative to muscle on T1 weighted images and high signal on T2 weighted images. The tumor may include cystic areas and intratumoral hemorrhage.[4,5] In our case, MR imaging revealed flow voids in and around the mass showing intermediate signal intensity on T1 weighted images and high signal intensity on T2 weighted images. There was no cystic area within the lesion. Angiography demonstrates a highly vascular tumor with one or more distinct vascular pedicles nourishing it, a rich network of peritumoral dilated and tortuous vessels. On angiography, the tumor has a rapid arteriovenous passage due to its rich capillary circulation.[4] Similarly, both lesions were highly vascular on angiography in our case.

Even though other soft tissue neoplasms are considered in differential diagnosis, definitive diagnosis can only be made after surgical excision. And also the most effective way of treatment of HP is surgical excision.[8] Recurrences which are a dominant feature of this tumor can also be resected. These tumors can be classified as benign, borderline malignant, or malignant based upon mitotic indices and cellular anaplasia. The clinical course appears to correlate well with the histologic classification.[1,2] About 20% of these tumors are benign without any tendency to local recurrence or far metastasis.[6] The borderline malignant and malignant ones tend to recur locally or to metastasize, particularly in the lungs and skeleton. Soft tissue metastasis is reported to be rare in the literature. About two third of the patients with malignant and potentially malignant tumors eventually develop distant metastases.[1,2] Although in literature it is reported that metastatic HP invariably involves the lungs.[1,2,] in our case this statement is not supported. The median duration between the first diagnosis and metastasis is reported to be 4 to 5 years, varying from 1 to 16 years.[3] In our case, soft tissue metastasis occurred after two years from HP located in the thigh.

In addition to surgical excision, radiation therapy is also advisable for local control of recurrences and metastatic disease.[1,2,8] There is little experience with chemotheraphy in HP, and it was mostly ineffective.[2,7] In the case described here is alive for 10 months after operation without administration of chemotheraphy or radiation therapy.

HP should be considered in the differential diagnosis of a vascular tumor in the thigh when MR images show the smooth contoured mass with intermediate signal intensity on T1 weighted images and flow voids within or adjacent to the tumor.[5] Angiography revealing a highly vascular tumor with rich capillary circulation supports the diagnosis. In adults, despite slow growth rate, a malignant form of HP should also be considered in the diagnosis of soft tissue tumors. The treatment of choice in HP is surgical excision. Surveillance for recurrence and metastasis is also mandatory.

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Corresponding author:
M. Tayfun Altınok M.D.
Inonu University Medical Faculty
Department os Radiodiagnostics
44069 Malatya, Turkey
Phone : 90 422 341 07 29
E-mail: taltinok@inonu.edu.tr