Giant Malignant Hemangiopericytoma of the Forearm: Case Report

ÖNKOLUN DEV MALİGN HEMANJİYOPERİSİTOMU: OLGU SUNUMU

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Abstract

Hemangiopericytoma is a rare soft tissue tumor. Large malignant hemangiopericytomas of the forearm are very rare. A patient with a malignant hemangiopericytoma of the left forearm, measuring 14 x 13 x 10 cm, is presented as a case report. The tumor was excised with wide margins to salvage the limb. Postoperative function of the hand was unimpaired and no complications occurred. Amongst the previously reported cases, our case is one of the largest noted.

Key Words: Hemangiopericytoma, malignant, forearm

Özet

Hemanjiyoperisitom nadir bir yumuşak doku tümörüdür. Önkolda yerleşimli büyük hemanjiyoperisitomlar ise çok nadirdir. Sol önkolda yerleşimli 14 x 13 x 10 cm boyutlarında malign hemanjiyoperisitomu olan hasta olgu sunumu olarak sunulmuştur. Kitle ekstremitenin korunması amacıyla geniş sınırlarla eksize edilmiştir. Ameliyat sonrasında hastanın el fonksiyonları tamdı ve herhangi bir komplikasyona rastlanmadı. Daha önce yayınlanmış olgular arasında bu olgu en büyük boyutlara sahip hemanjiyoperisitomdur.

Anahtar Kelimeler: Hemanjiyoperisitom, malign, önkol

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Hemangiopericytoma is a rare soft-tissue tumor, arising from primitive perivascular cells. It was first described by Stout and Murray in 1942. Only 14 cases of malignant hemangiopericytomas have been reported in English literature. The perivascular cells that form hemangiopericytoma are believed to be the pericytes of the capillaries, first defined by Zimmerman.[1] As hemangiopericytomas do not have nerve elements, they are painless. Most of the patients realize and apply for treatment when they suffer from pain or the masses enlarge to large measures. The nature of the hemangiopericytomas cannot be predicted. Apparent malignant forms which metastasise have been reported (up to 45% of some series). Presentation on the forearm is rare, especially for the malignant forms, and even more rare cases enlarge to bulky masses on this localisation. A rare case of a large malignant hemangiopericytoma of the forearm is reported.[2-5]

Case Report

A 42-year old male patient presented with a large mass on his left forearm. His history revealed a small subcutaneous mass, which appeared 3 years ago on his left elbow. The mass had enlarged to 4 x 3.5 x 3 cm and the tumor was excised by a general surgeon 5 months prior to presentation to our facility. Histopathologically, hemangiopericytoma was detected. Two months after the initial operation, the lesion has recurred as a red-purple mass, causing echymotic areas on the palm. On presentation, the patient had a painless, immobile, red-purple mass of 14 x 13 x 10 cm of the left forearm. Neurologic and vascular examination of the left upper extremity were normal (Figure 1).

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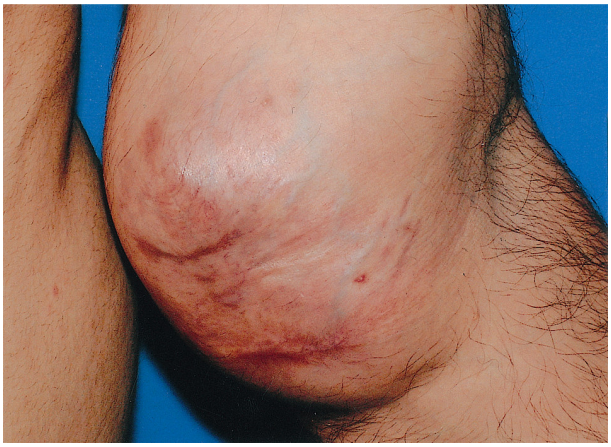


Figure 1. Preoperative anterior view.

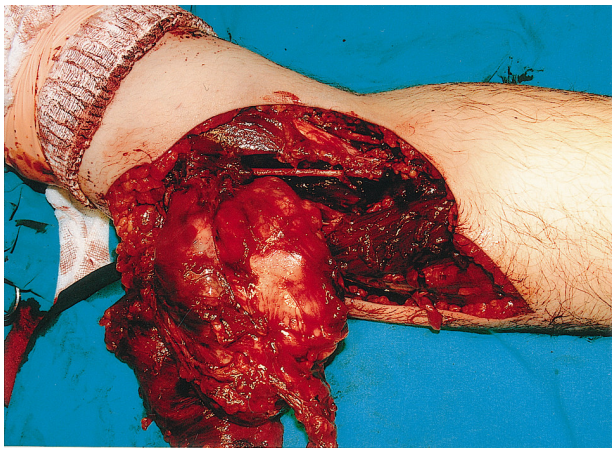
Ultrasound showed a mixed echogenicity of this soft tissue tumor, similar to hemangioma, localised in the left antecubital fossa, measuring the same as the physical examination. The left subclavian, axillary and brachial arteries and veins were patent in the colour doppler US, but, the brachial artery and vein were displaced, and the flow pattern of the artery was diminished by the mass effect of the tumor. The computerised tomography of the antecubital region revealed a discrete soft tissue mass, with necrotic and cystic degeneration areas apparent on the superior pole, causing displacement of the biceps muscle and brachial artery and vein. The tumor was nonhomogenous and had luminal opacities on the distal part, and invaded the extensor carpi radialis longus muscle. No evidence of bone destruction was seen on the tomography. Chest radiographs, bone scans, abdominal ultrasound showed no evidence of metastasis.

The patient was operated under general anaesthesia, dissection was performed after an elliptical incision of a skin island measuring 10 x 6 cm. The tumor was attached to the biceps muscle. The incisions were extended proximally and distally and the mass was seen as three lobules. The brachial artery was distorted due to the mass effect, but was not attacked. The artery and the median nerve were dissected and the lesion was resected en bloc with wide margins. Radical excision was not preferred to salvage the limb (Figure 2). The skin was closed primarily.

Histopathologic examination showed cell groups, surrounding the thin layered vascular structures. The tumor cells were uniform, round, had oval nuclei and irregular cytoplasms. A low rate of mitosis was also seen (Figure 3). Neither local recurrence nor far metastasis were seen during 1 year follow up, by the assessment of the patient with the previously mentioned preoperative imaging studies (Figure 4).

Discussion

Hemangiopericytoma of the arm is a very rare tumor. Although hemangiopericytomas may present late as very large masses because they are



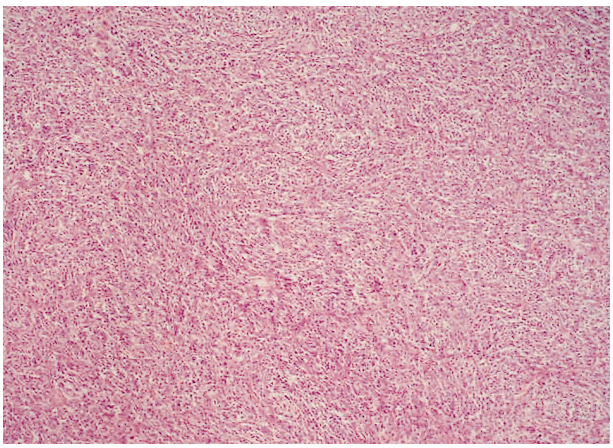


Figure 3. Histopathologic examination (H.E. x 40).

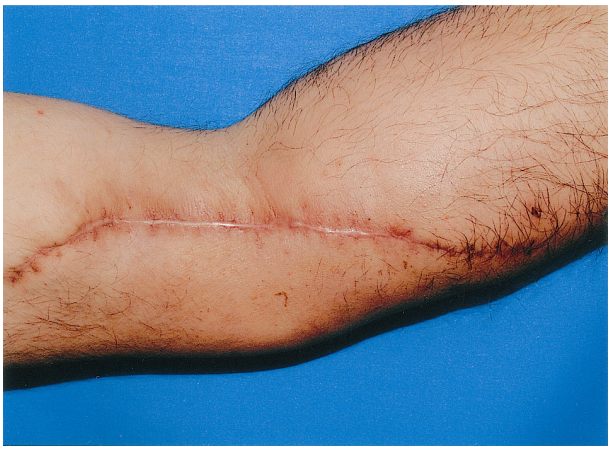


Figure 4. Postoperative 1 year.

painless and go unnoticed, malignant forms, exceeding 10 cm in measurement, located on the extremities are even more rare. Depending on these aspects and compared to the previously published case reports, our case seems to be a very rare and one of the largest hemangiopericytomas.[3] The largest malignant hemangiopericytoma ever reported measures 9 x 10 x 15 cm.[6]

Differential diagnosis includes hemangioendothelioma, glomus tumor, snovial sarcoma and fibrous histiocytoma. Histopathologic assessment is essential for the accurate diagnosis.[5]

Malignant hemangiopericytomas often metastasise, especially to lungs and the bones, but even when no metastatic lesion is detected, increased rates of mitosis and cellularity; focal necrosis and haemorrhage indicates malignancy.[7] Our case also had no distant metastatic lesions. The high metastatic potential of hemangiopericytoma is due to the close relationship of hemangiopericytoma with the capillaries.[3,8,9]

The most appropriate treatment modality for hemangiopericytoma is radical surgical excision. The reports about the role of radiotherapy and chemotherapy for the treatment of hemangiopericytoma are controversial.[3] O'Brien and Brasfield first reported that the tumor was resistant to radiotherapy and chemotherapy, like Kinnard et al and recommended wide excision.[10,11] However later studies documented tumor response to doses greater than 45 Gy.[12-14] Dube and Paulson and Jha et al claimed success of radiotherapy for total cure following surgical excision, especially for the metastatic foci.[12,15] Though chemotherapy is accepted to be ineffective, partial or short-term remission of metastatic lesions with doxyrubicin and successful control of metastatic foci in a child using actinomycin D, cyclophosphamide, vincristine and methotrexate have been reported.[16,17]

Local recurrence of epidural and retroperitoneal tumors after curative excision is reported to be high, on the contrary to the ones located on the extremities. Five-year survival rates are reported to be 71% totally, though; no recurrence has been reported, for the 5-year follow-up, for the cases that had undergone radical excision of the tumor, located on the forearm. No additional therapy is planned for our case after the surgical intervention, and recurrence has not occurred during the last 1-year follow up. As the biological nature of this tumor is unpredictable, and recurrences after 5-year tumor-free follow up have been reported, close clinical follow up should exceed 10 years.[2,3,8,9]

The presented case in here is the largest hemangiopericytoma of the forearm. Though radical surgical excision is the optimal treatment, wide marginal excision was performed to salvage the limb with all the functions intact. The previous studies indicate that although there is a high rate of metastasis, metastatic lesions can be cured easily with radiotherapy. The patient has no recurrence at the end of the first postoperative year, and full functions of his hand; but still under close follow-up.

REFERENCES

1. Zimmerman KW. Der feinere bau der blutcapillaren. Z. Anat Entwicklungsg 1923;68:29-48.
2. Bremner CG, Levy JI, Proctor NS. Hemangiopericytoma of the thigh. S Afr Med J 1975;49(6):197-200.
3. Kasdan ML, Stallings SP. Malignant hemangiopericytoma of the forearm. Plast Reconstr Surg 1993;91(3):533-6.
4. Nunnery EW, Kahn LB, Reddick RL, Lipper S. Hemangiopericytoma: A light microscopic and ultrastructural study. Cancer 1981;47(5):906-14.
5. Sen R. Hemangiopericytoma of the hand. Plast Reconstr Surg 1976;57(6):746-8.

6. Jenkins JJ. Congenital malignant hemangiopericytoma. Pediatr Pathol 1987;7:119-22.
7. Enzinger FM, Smith BH. Hemangiopericytoma. An analysis of 106 cases. Hum Pathol 1976;7(1):61-82.
8. Pandey M, Kothari KC, Patel DD. Hemangiopericytoma: Current status, diagnosis and management. Eur J Surg Oncol 1997;23(4):282-5.
9. Perugia D, Basile A, Massoni C, Barletta V. Haemangiopericytoma in the distal third of the arm. Int Orthop 1999;23(3):184-6.
10. O'Brien P, Brasfield RD. Hemangiopercytoma. Cancer 1965;18:249-52.
11. Kinnard P, Likongo Y, Echave V, Masse S. Hemangiopericytoma: Case reports. Can J Surg 1984;27:145-6.
12. Jha N, McNeese M, Barkley HT Jr, Kong J. Does radiotherapy have a role in hemangiopericytoma management? Report of 14 new cases and review of literature. Int J Radiat Oncol Biol Phys 1987;13(9):1399-402.
13. Staples JJ, Robinson RA, Wen BC, Hussey DH. Hemangiopericytoma, the role of radiotherapy. Int J Radiat Oncol Biol Phys 1990;19:445-51.
14. Mira JG, Chu FC, Fortner JG. The role of radiotherapy in the management of malignant hemangiopericytoma: Report of eleven new cases and review of the literature. Cancer 1977;39:1254-9.
15. Dube VE, Paulson JF. Metastatic hemangiopericytoma cured by radiotherapy. J Bone Joint Surg 1974;56(4):833-5.
16. Wong PP, Yagoda A. Chemotherapy of malignant hemangiopericytoma. Cancer 1978;41:1256-60.
17. Ortega JA, Finklestein JZ, Isaacs H Jr, Hittle R, Hastings N. Chemotherapy of malignant hemangiopericytoma of childhood. Cancer 1971;27:730-5.