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VELIKI HEMANGIOPERICYTOM RAMENOG POJASA: PRIKAZ SLUČAJA

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Large hemangiopericytoma of the shoulder: a case report
Veliki hemangiopericitom ramenog pojas: prikaz slučaja

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Abstract

**Introduction:** Hemangiopericytoma is a rare tumor arised from the pericytes, contractile spindle cells that surround the capillaries and post-capillary venules. The tumor was found equally among males and females.

**Case report:** We report a case of a 63-year-old female who presented a giant painful mass on her right shoulder that occurred one year before admission. Limited range of motion and a sense of tingling along the affected arm was present also. An irregular, oval-shaped mass, colored dark-red, with signs of necrosis was the maximal diameter of 14cm. Routine laboratory analyzes showed results within the range of referenced values. Lung X-ray and ultrasonographic examination of the abdomen showed no signs of secondary tumor deposits, and no osteo-muscular lesions in the affected region. Ultrasonographic examination of the neck and right axillary region showed no signs of regional metastases. The surgical excision of the entire tumor was performed, with the associated subcutaneous tissue and a part of fascia underneath. Results of the histopathological analysis confirmed the diagnosis of hemangiopericytoma. The specimen showed no signs or elements of the neoplastic tissue on the edges of the resection lines. 3 years after the operation, there was found no signs of tumor relapses, regional or systemic metastases. **Conclusion:** Considering that there are no official clinical guides and protocols for hemangiopericytoma management, as well as the occurrence of cutaneous and subcutaneous hemangiopericytomas is exceptionally rare, more extensive research in this field and more described cases are need to gain a better understanding of that issue.

**Keywords:** hemangiopericytoma, vascular tumor, mesenchimal neoplasm, surgery
Sažetak

Uvod: Hemangiopericitom je redak tumor porekla pericita, kontraktilnih vretenastih ćelija koje okružuju kapilare i postkapilarne venule. Tumor se javlja podjednako u muškoj i ženskoj populaciji.

Prikaz slučaja: Predstavljamo slučaj 63-godišnje žene sa džinovskom bolnom promenom na desnom ramenu, primećenu godinu dana pre prijema. Simptomi koji su takođe bili prisutni su ograničen opseg pokreta kao i osećaj trnjenja u zahvaćenoj ruci. Promena je bila nepravilno ovalnog oblika, tamnocrveno prebojena, sa znakovima nekroze, i maksimalnog prečnika 14cm. Rutinske laboratorijske analize pokazale su rezultate u granicama referentnih vrednosti. Rendgenski snimak pluća i ultrasonografski pregled abdomena nisu pokazali znakove sekundarnih depozita tumora, kao ni osteomišićne lezije u zahvaćenom regionu. Ultrazvučni pregled vrata i desne aksilarne regije nije pokazao znakove regionalnih metastaza. Izvršena je hirurška ekscizija celog tumora, sa pripadajućim potkožnim tkivom i delom fascije ispod. Rezultati histopatološke analize potvrdili su dijagnozu hemangiopericitoma. Uzorak nije pokazao znakove ili elemente neoplastičnog tkiva na ivicama resekcionih linija. 3 godine nakon operacije nisu pronađeni znakovi relapsa tumora, regionalnih ili sistemskih metastaza.

Zaključak: Obzirom na činjenicu da ne postoje zvanični klinički vodiči i protokoli za lečenje hemangiopericitoma, kao i da je pojava kožnih i potkožnih hemangiopericitoma izuzetno retka, potrebno je opsežnije istraživanje na ovu temu i više prikaza slučajeva da bi se steklo bolje razumevanje problema.

Ključne reči: hemangiopericitom, vaskularni tumor, mezenhimalna neoplazma, hirurgija
Introduction

First description of a hemangiopericytoma (HP) in literature appeared in a paper published by Stout and Murray in 1942. The authors stated that haemangiopericytoma (peritelioma - an older name) is an unusual mesenchymal neoplasm. It is believed that hemangiopericytoma arises from the pericytes, contractile spindle cells that surround the capillaries and post-capillary venules. Pericytes are described as modified smooth-muscle cells or resting stem cells which are capable to differentiate in myoid, fibrohistiocytic and endothelial cells. The tumor is found equally among males and females. Hemangiopericytoma is found to appear at any age, but is most common in the sixth and seventh decade. Hemangiopericytoma is primary an adult neoplasm, but occurrence in children is possible also. Considering that, adult and infantile forms of hemangiopericytoma are described in the literature. Some types of hemangiopericytoma (such as glomangiopericytoma) are associated with previous trauma, hypertension, pregnancy or steroid usage but definitive etiology is still insufficiently clarified. The probability of a tumor forming is greater wherever the tissue abounds in capillaries. It has been stated that hemangiopericytoma may occur at any anatomic site and tends to develop from subcutaneous tissue or skeletal muscle. However, in cases reported so far there are areas more affected such as the lower extremities (above the knee) and retroperitoneum, soft tissue of trunk and upper extremities, head and neck, thoracic cavity. In the head and neck region, they are usually seen in the nasal cavity, orbit, jaw, parotid gland, and oral cavity. Hemangiopericytomas also could arise intracranially within the central nervous system (CNS) and account for approximately 0.4% of all CNS tumors. The majority of hemangiopericytomas have a profound localization, inside the muscle tissue or viscera, with rare cases where the tumor infiltrates only cutaneous and subcutaneous tissue, without deeper propagation. Hemangiopericytoma of the upper extremity is rare, but in 2012 authors have reported a case of a hemangiopericytoma in the dorsal region of the hand, with only several other cases described on the upper extremity. Rare occurrence of the cutaneous hemangiopericytoma, giant size reached in our patient, and the specific localization, previously not reported in the literature, prompted us to describe our case of this unusual neoplasm in the shoulder region.
Case Report

We present a case of a 63-year old female patient who was referred to the Department of Plastic surgery with a giant painful mass on her right shoulder. The patient was presented with pain, limited range of motion and a sense of tingling along the affected arm, on the initial examination. The tumor was present one year prior to the initial manifestation of the symptoms, when the patient noticed a growth of the neoplasm with occasional bleeding. The neoplasm was described as an irregular, oval-shaped mass, colored dark-red, with signs of necrosis on different regions of the tumor and occasional bleeding and secrernation. (Figure 1) It was localized in the right supraclavicular region, with maximal diametar of 14cm, on a wide base with a vague demarcation line toward the surrounding skin. The surface of the tumor was uneven, striated with scabs and fields of secondary necrosis. Surrounding skin showed signs of reactive hyperemia. Routine laboratory analyzes and coagulation time, which were performed as a part of preoperative diagnostics, showed results within the range of referenced values. Thorough history and clinical examination of the patient did not reveal any problems concerning other organ systems. Lung X-ray and ultrasonographic examination of the abdomen showed no signs of secondary tumor deposits, and no osteo-muscular lesions in the affected region. Ultrasonographic examination of the neck and right axillary region showed no signs of regional metastases. Under general anesthesia, wide excision was performed. Entire tumor was removed, with the associated subcutaneous tissue and a part of fascia underneath. (Figure 2) Intraoperatively we found that the muscle tissue was unaffected by the tumor. Defect was closed primarily by split thickness skin graft. The resected tissue was sent to the Pathology department for histopathological analysis. Results of the histopathological analysis confirmed the diagnosis of hemangiopericytoma. (Figure 3) The specimen showed no signs or elements of the neoplastic tissue on the edges of the resection lines. The patient was on the hospital treatment for 12 days after the surgery. During the routine postoperative check-ups, which were performed regularly 3 years after the operation, we found no signs of tumor relapses, regional or systemic metastases. Functional and aesthetic result of the surgery were satisfactory. (Figure 4)
Discussion

Hemangiopericytomas manifest as slow growing, firm, and painless masses. Despite the fact that the uniform clinical presentation of hemangiopericytoma does not exist the pain is reported as a late symptom. The reason for that phenomenon is most likely compression of the neurovascular structures. Absence of direct innervation of the tumor is most likely the reason why it is not uncommon that patients do not present with symptoms until the mass reaches considerable size. Beside the pain, enlarging of the tumor in our patient was the main reason for limited range of motion and a sense of tingling along the affected arm, because the tumor invaded the cutaneous nerves in the shoulder region of our patient. Regarding the upper extremity, a case of hemangiopericytoma of the dorsal region of the hand was described in literature. Since both are localized in the upper extremity, that case could be compared with our patient. However, the difference between them is manifested by various symptoms, as the tumors affected different regions of the upper extremity. Besides the fact that it is very important to distinguish the benign or malignant morphology of the tumor, which significantly influences the course of treatment, two clinical syndromes of hemangiopericytoma (infantile and adult) have been described in the literature, depending on the age at which it occurs. Certain authors consider the infantile type of hemangiopericytoma to occur before the age of 1, while others set the limit at 5 years. The cases that appear after those age marks were defined as adult types of hemangiopericytoma. Infantile and adult types of hemangiopericytoma differ clinically and pathologically from one another. Infantile types occur more often as cutaneous and subcutaneous head and neck lesions, displaying benign behavior despite the appearance of certain histological patterns such as hypercellularity, necrosis, bleeding and increased mitotic proliferation. There have been reports in the literature of multiple infantile hemangiopericytomas in the head and neck region. The interesting fact in our case is that we found an adult form of hemangiopericytoma with cutaneous and subcutaneous localization, which is, according to the literature, characteristic of the infantile type of this tumor. Enzinger and Smith reported that 4 out of 9 infantile types of hemangiopericytoma were morphologically benign in their study of 106 cases. Depending on the localization of the tumor, a large variety of symptoms were described in the same study. For example, tumors localized in the pelvic fossa and the retroperitoneum caused urinary retention, hydrenephrosis, dysuria, nocturia, constipation and hematuria. Tumors situated at other
sites, such as upper respiratory pathways caused epistaxis, cough, dyspnea, while symptoms such as vomiting and distention occurred with digestive tract affection. The mentioned symptoms were a consequence of local tumor invasion. Furthermore, various paraneoplastic symptoms were described following the appearance of hemangiopericytoma. Hypoglycemia has been reported in about 5% of patients with hemangiopericytoma, with the most frequent localization of the tumor in the retroperitoneum and the pelvic area. Benn et al. have showed in 1990 that hypoglycemia is most probably caused by the production of the insulin-like growth factor in the tumor. The most likely proposed mechanisms were increased tumor glucose uptake, decreased hepatic output and increased glucose tissue utilization. Another described paraneoplastic manifestation associated with hemangiopericytoma is hypophosphatemic osteomalacia.

Also, there is a reported case of paraneoplastic rhinophyma-like nasal swelling with Leser-Trélat sign which was resolved post-operatively. The sign of Leser-Trélat is described in the literature as an association of eruptive, pruritic, seborrheic keratoses with occult internal malignancy and any appearance of this sign should raise suspicion of an underlying malignancy. During the preoperative diagnostics and routine postoperative check-ups during the follow-up period (3 years), our patient showed no signs of metabolic disbalance or manifestations in other organ systems, even though certain paraneoplastic symptoms are reported in the literature. Histological analysis of the tumor showed pericytoma-like vascularization, which may be seen as a secondary vascular pattern in other mesenchymal lesions, hence the differentiation of benign or malignant forms of hemangiopericytoma is made by exclusion. That is essential to stand out, because hemangiopericytoma, along with certain other neoplasms such as synovial sarcoma and solitary fibrous tumor, isn’t distinctly classified, and presents a problem in confirming the correct diagnosis due to pathologic similarities between these tumors. Histological confusion with synovial sarcoma and solitary fibrous tumor exists because of the same pericytoma-like vascularization pattern in all three neoplasms. Pathohistologically observed, benign forms of hemangiopericytoma feature bland-oval or spindle cells, immersed in a reticulin network, and arranged around an elaborate gaping vasculature, without endothelial proliferation. Perivascular hyalinization is commonly present, which was discovered and confirmed in our case, along with the other benign characteristics. On the other hand, hemangiopericytomas with aggressive, anaplastic features, such as atypia, high mitotic
activity, hemorrhage and necrosis, are defined as malignant. Possible routes of hemangiopericytoma metastasis could be explained by three pathways: direct extension, via the lymphatics, and hematogenous pathway, which is the most frequent route. Considering the difficulty to predict biological behavior of this tumor, it is useful to know that the prognostic value includes increased cellularity, anaplasia, necrosis, hemorrhage and prominent mitotic activity detected by microscope, which could be amplified using a proliferation index detected by immunohistochemical techniques. The proliferation index of 10% or greater may indicate a more aggressive type of this rare tumor. The criteria for malignancy proposed by Enzinger and Smith in classical HPC identify overtly malignant or high-grade lesions, but fail to address low-grade lesions. In their study, large-sized tumors (>5cm), increased the mitotic rate, high cellularity, presence of immature and pleomorphic tumor cells, and foci of hemorrhage and necrosis predicted a highly malignant course. Enzinger and Weiss employed the term „low-malignant potential“ for lesions with lower levels of mitotic activity, particularly if they have any degree of atypia and cellularity. Metastases were noted in approximately 30% of patients, with a 5-year survival of 71%. The most common metastatic sites were the lungs, bones and liver. Occurrence of metastases several years after the excision of the primary lesion is a frequent phenomenon regarding hemangiopericytoma and therefore long-term follow-up is essential. Current consensus regarding the treatment of hemangiopericytoma is wide, radical surgical excision with the radiotherapy follow-up if the tumor shows malignant characteristics. We performed wide surgical excision, with extirpation of the tumor in full and radical resection to clean edges, without adjuvant radiotherapy, since pathohistological analysis showed benign characteristics of the tissue. One of the main factors of increased surgical morbidity and mortality is the increased hypervascularization in the tumor region, which makes the treatment difficult. Since the tumor originates from cells which are a part of the vascular system and blood vessels, it is recommended to detect the type and extension of the circulation in the neoplasm, and if necessary perform the preoperative ligation of afferent vessels or vascular embolization, which could reduce perioperative hemorrhage and facilitate further management. During the diagnostics and treatment, it should be kept in mind to consider the possibility of complex vascular syndromes because hemangiopericytoma originates from pericytes. For example, there may occur multiple hemangiomas and multiple organ cysts such as Von Hippel-Lindau or other anomalies. In
this case, in addition to the radical surgical treatment, a multidisciplinary approach would be important.

Conclusion
Early histological diagnosis of hemangiopericytoma is essential and wide surgical excision is recommended. Considering that there are no official clinical guides and protocols for hemangiopericytoma management, more extensive research in the field and more described cases are need to gain a better understanding of the issue. The fact that the occurrence of cutaneous and subcutaneous hemangiopericytomas is exceptionally rare give this particular case report great clinical significance and further our insight in the pathology of this neoplasm.

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Figures

**Figure 1.** Large, well-circumscribed reddish growth on the right arm skin

**Figure 2.** Wide excision of the lesion and postexcisional skin and soft tissue defect
Figure 3. H&E x100; Arrow 1: Tumor cells around vascular spaces (periteliial proliferation) in short fascicular arrangements; Arrow 2: Numerous branched vascular spaces bounded by a single-row endothelium

Figure 4. Aesthetic and functional result one year after excision and skin grafting

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