CASE REPORT
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Large hemangiopericytoma of the shoulder: A case report

Veliki hemangiopericitom ramenog pojasa

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Abstract

Introduction. Hemangiopericytoma is a rare tumour derived from the pericytes, contractile spindle cells that surround the capillaries and postcapillary venules. The tumour is found equally among males and females. **Case report.** We reported 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, dark red in colour, with signs of necrosis, was 14 cm in maximum diameter. Routine laboratory analysis showed results within the range of referenced values. Lung X-ray and ultrasonographic examination of the abdomen showed no signs of secondary tumour deposits, and no osteomuscular 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 tumour 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. Three years after the operation, there were no signs of tumour 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 needed to gain a better understanding of the issue.

Key words:
diagnosis; hemangiopericytoma; histological techniques; reconstructive surgical procedures; shoulder; ultrasonography.

Apstrakt

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 bolesnika.** Prikazana je 63-godišnja bolesnica sa džinovskom bolnom promenom na desnom ramenu, primećenu godinu dana pre prijema. Žalila se na ograničen opseg pokreta kao i osećaj trnjenja u zahvaćenoj ruci. Promena je bila nepravilno ovalnog oblika, tamnocrvene boje, sa znakovima nekroze, maksimalnog prečnika 14 cm. 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, a 3 godine nakon operacije nisu bilo znakova relapsa tumora, regionalnih ili sistemskih metastaza. **Zaključak.** S 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:
dijagnoza; hemangiopericitom; histološke tehnike; hirurgija, rekonstruktivna, procedure; rame; ultrasonografija.

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Introduction

First description of a hemangiopericytoma (HP) in literature appeared in a paper published by Stout and Murray [1] in 1942. The authors stated that HP (peritelioma – an older name) is an unusual mesenchymal neoplasm. It is believed that HP arises from the pericytes, contractile spindle cells that surround the capillaries and postcapillary venules [2]. Pericytes are described as modified smooth muscle cells or resting stem cells which are capable of differentiating in myoid, fibrohistiocytic and endothelial cells. The tumour is found equally among males and females [3]. HP is found to appear at any age but is most common in the sixth and seventh decade [4]. HP is primary an adult neoplasm, but occurrence in children is possible also. Considering that, adult and infantile forms of HP are described in the literature [5]. Some types of HP (such as glomangiopericytoma) are associated with previous trauma, hypertension, pregnancy or steroid usage but definitive aetiology is still insufficiently clarified [6]. The probability of a tumour forming is greater wherever the tissue abounds in capillaries. It has been stated that HP 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 [7], soft tissue of trunk and upper extremities [8], head and neck [9], thoracic cavity [10]. In the head and neck region, they are usually seen in the nasal cavity, orbit, jaw, parotid gland, and oral cavity [11]. HP also could arise intracranially within the central nervous system (CNS) and account for approximately 0.4% of all CNS tumours [12]. The majority of HPs have a profound localization, inside the muscle tissue or viscera, with rare cases where the tumour infiltrates only cutaneous and subcutaneous tissue, without deeper propagation [5]. HP of the upper extremity is rare, but in 2012 authors have reported a case of a HP in the dorsal region of the hand [13], with only several other cases described on the upper extremity. Rare occurrence of the cutaneous HP, 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 tumour 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, coloured dark red, with signs of necrosis on different regions of the tumour and occasional bleeding and secretion (Figure 1). It was localized in the right supraclavicular region, with maximal diameter of 14 cm, on a wide base with a vague demarcation line toward the surrounding skin. The surface of the tumour was uneven, striated with scabs and fields of secondary necrosis. Surrounding skin showed signs of reactive hyperaemia. Routine laboratory analysis 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 tumour deposits, and no osteomuscular lesions in the affected region. Ultrasonographic examination of the neck and right axillary region showed no signs of regional metastases. Under general anaesthesia, wide excision was performed. Entire tumour 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 tumour. Defect was closed primarily by split thickness skin graft. The resected tissue was sent to the Department of Pathology for histopathological analysis. Results of the histopathological analysis confirmed the diagnosis of HP (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



Fig. 1 – Large, well-circumscribed reddish growth on the right arm skin.

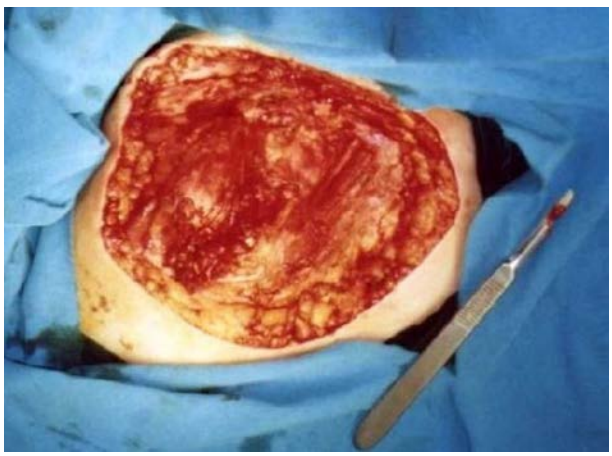


Fig. 2 – Wide excision of the lesion and post excisional skin and soft tissue defect.

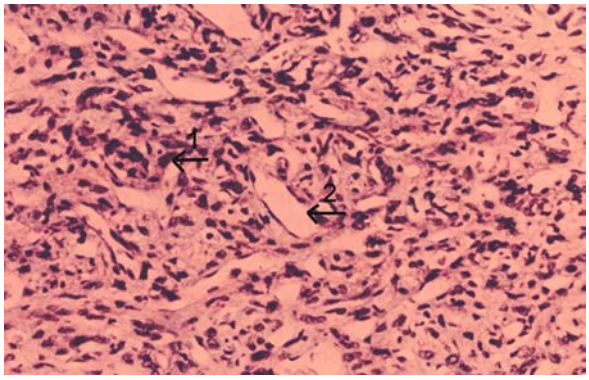


Fig. 3 –Arrow 1: tumour cells around vascular spaces (peritelial proliferation) in short fascicular arrangements; Arrow 2: numerous branched vascular spaces bounded by a single-row endothelium (haematoxylin and eosin staining, x10).

postoperative check-ups, which were performed regularly 3 years after the operation, we found no signs of tumour relapses, regional or systemic metastases. Functional and aesthetic result of the surgery were satisfactory (Figure 4).



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

Discussion

HP manifest as slow growing, firm, and painless masses. Despite the fact that the uniform clinical presentation of HP does not exists, the pain is reported as a late symptom [14]. The reason for that phenomenon is most likely compression of the neurovascular structures. Absence of direct innervation of the tumour 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 tumour in our patient was the main reason for limited range of motion and a sense of tingling along the affected arm, because the tumour invaded the cutaneous nerves in the shoulder region of our patient. Regarding the upper extremity, a case of HP of the dorsal region of the hand was described in literature [13]. 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 tumours affected different regions of the upper extremity. Besides the fact that it is very important to distinguish the benign or malignant morphology of the tumour, which significantly influences the course of treatment, two clinical syndromes of HP (infantile and adult) have been described in the literature, depending on the age at which it occurs. Certain authors consider the infantile type of HP 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 HP [5]. Infantile and adult types of HP differ clinically and pathologically from one another. Infantile types occur more often as cutaneous and subcutaneous head and neck lesions, displaying benign behaviour 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 HPs in the head and neck region [15]. The interesting fact in our case is that we found an adult form of HP with cutaneous and subcutaneous localization, which is, according to the literature, characteristic of the infantile type of this tumour. Enzinger and Smith [16] reported that 4 out of 9 infantile types of HP were morphologically benign in their study of 106 cases. Depending on the localization of the tumour, a large variety of symptoms were described in the same study. For example, tumours localized in the pelvic fossa and the retroperitoneum caused urinary retention, hydronephrosis, dysuria, nocturia, constipation and haematuria. Tumours situated at other sites, such as upper respiratory pathways caused epistaxis, cough, dyspnoea, while symptoms such as vomiting and distention occurred with digestive tract affection. The mentioned symptoms were a consequence of local tumour invasion. Furthermore, various paraneoplastic symptoms were described following the appearance of HP. Hypoglycaemia has been reported in about 5% of patients with HP, with the most frequent localization of the tumour in the retroperitoneum and the pelvic area. Benn et al. [17] have showed in 1990 that hypoglycaemia is most probably caused by the production of the insulin-like growth factor in the tumour. The most likely proposed mechanisms were increased tumour glucose uptake, decreased hepatic output and increased glucose tissue utilization. Another described paraneoplastic manifestation associated with HP is hypophosphatemic osteomalacia [18]. Also, there is a reported case of paraneoplastic rhinophyma-like nasal swelling with Leser-Trélat sign which was resolved postoperatively [19]. 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 (three 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 tumour 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 HP is made by exclusion [20]. That is essential to stand out, because HP, along with certain other neoplasms such as synovial sarcoma and solitary fibrous tumour, is not distinctly classified, and presents a problem in confirming the correct diagnosis due to pathologic similarities between these tumours. Histological confusion with synovial sarcoma and solitary fibrous tumour exists because of the same pericytoma-like vascularization pattern in all three neoplasms [1]. Histopathologically observed, benign forms of HP feature bland-oval or spindle cells, immersed in a reticulin network, and arranged around an elaborate gaping vasculature, without endothelial proliferation [20]. Perivascular hyalinization is commonly present, which was discovered and confirmed in our case, along with the other benign characteristics. On the other hand, HPs with aggressive, anaplastic features, such as atypia, high mitotic activity, haemorrhage and necrosis, are defined as malignant. Possible routes of HP metastasis could be explained by three pathways: direct extension, via the lymphatics, and hematogenous pathway, which is the most frequent route [21]. Considering the difficulty to predict biological behaviour of this tumour, it is useful to know that the prognostic value includes increased cellularity, anaplasia, necrosis, haemorrhage 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 tumour [15]. The criteria for malignancy proposed by Enzinger and Smith [16] in classical HP identify overtly malignant or high-grade lesions but fail to address low-grade lesions. In their study, large-sized tumours (> 5 cm), increased the mitotic rate, high cellularity, presence of immature and pleomorphic tumour cells, and foci of haemorrhage and necrosis predicted a highly malignant course. Enzinger and Weiss [16] employed the term ”low-malignant potential“ for lesions with lower levels of mitotic activity, particularly if they have any degree of atypia and cellularity [22]. 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 [23]. Occurrence of metastases several years after the excision of the primary lesion is a frequent phenomenon regarding HP and therefore long-term follow-up is essential [24]. Current consensus regarding the treatment of HP is wide, radical surgical excision with the radiotherapy follow-up if the tumour shows malignant characteristics [25]. We performed wide surgical excision, with extirpation of the tumour in full and radical resection to clean edges, without adjuvant radiotherapy, since histopathological analysis showed benign characteristics of the tissue.

One of the main factors of increased surgical morbidity and mortality is the increased hypervascularization in the tumour region, which makes the treatment difficult. Since the tumour 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 haemorrhage and facilitate further management [5]. During the diagnostics and treatment, it should be kept in mind to consider the possibility of complex vascular syndromes because HP originates from pericytes. For example, there may occur multiple haemangiomas 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 HP is essential and wide surgical excision is recommended. Considering that there are no official clinical guides and protocols for HP management, more extensive research in the field and more described cases are needed to gain a better understanding of the issue. The fact that the occurrence of cutaneous and subcutaneous HP 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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