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Case report
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SYNOVIAL SARCOMA OF THE HAND – CASE REPORT AND LITERATURE REVIEW

SINOVIJALNI SARKOM ŠAKE – PRIKAZ SLUČAJA I PREGLED LITERATURE

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Summary

Introduction. Synovial sarcoma is a rare malignant tumor most often localized on the lower limbs near large joints. Histopathology findings is the gold standard for making the diagnosis. However, due to the similarity with other tumors, the findings can be misinterpreted and the correct diagnosis often delayed, and the treatment is inadequate in the early stages of the disease inadequate. **Case Report.** A 15-year-old female patient came for an examination due to subcutaneous lesion in her hand. The lesion was excised on three occasions due to recurrence, and each time the histopathology findings showed a dermatofibroma. After five years, the patient returned due to the changes under the scar and unpleasant sensations. A surgical excision was performed when dermatofibroma was verified. Two years later, the patient developed necrosis of the entire finger. The finger was amputated, and synovial sarcoma was verified. The patient underwent 25 cycles of radiotherapy. In the following year and a half, the tumor developed metastases, first locoregional and then distant, in the region of the left shoulder joint and the left lung. **Conclusion.** The histopathological similarity of this tumor with other benign changes indicates the need for further evaluation and differentiation of the pathohistological characteristics from tumors with similar characteristics. Early detection of this type of tumor enables a more favorable course and outcome of the treatment of these patients. Initial radical surgical treatment of this type of tumor is necessary due to the aggressiveness they show and the high frequency of local recurrence.

Key words: Sarcoma, Synovial; Hand; Soft Tissue Neoplasms; Diagnosis, Differential; Diagnostic Errors; Neoplasms; Recurrence

Sažetak

Uvod. Sinovijalni sarkom je redak maligni tumor najčešće lokalizovan na donjim ekstremitetima u okolini velikih zglobova. Zlatni standard za postavljanje dijagnoze je patohistološki nalaz. Međutim, zbog sličnosti sa drugim tumorskim promenama, nalaz se može pogrešno protumačiti te je postavljanje prave dijagnoze često odloženo, a lečenje neadekvatno u ranim fazama bolesti. **Prikaz slučaja.** Petnaestogodišnja pacijentkinja se javila na pregled zbog promene na šaci. Promena je ekscidirana u tri navrata zbog recidiviranja i sva tri puta je patohistološki nalaz ukazivao na dermatofibrom. Nakon pet godina pacijentkinja se javlja zbog promene ispod ožiljka i neprijatnih senzacija. Urađena je hirurška ekscizija kada je patohistološkim nalazom verifikovan dermatofibrom. Dve godine kasnije kod pacijentkinje se razvila nekroza celog prsta, zbog čega se javila na kontrolni pregled. Prst je amputiran, a patohistološkom analizom je verifikovan sinovijalni sarkom. Kod pacijentkinje je potom sprovedeno 25 ciklusa radioterapije. U narednih godinu i po dana je ovaj tumor dao metastaze, prvo lokoregionalne, a potom i udaljene, u predelu zgloba levog ramena i levog plućnog krila. **Zaključak.** Patohistološka sličnost ovog tumora sa drugim benignim promenama ukazuje na potrebu za daljom evaluacijom i diferencijacijom patohistoloških karakteristika od tumora srodnih karakteristika. Rano otkrivanje ove vrste tumora omogućava povoljniji tok i ishod lečenja ovih pacijenata. Neophodno je inicijalno radikalno hirurško lečenje ove vrste tumora zbog agresivnosti koju pokazuju kao i velike učestalosti lokalnog recidiva.

Ključne reči: sinovijalni sarkom; šaka; neoplazme mekog tkiva; diferencijalna dijagnoza; dijagnostičke greške; metastaze; recidivi

Introduction

Synovial sarcoma is a relatively rare malignant tumor, most often localized around joints, tendons or bursae, but can also be found outside these areas. The prevalence of this tumor is high in patients between 10 and 40 years [1]. It is usually localized near the large joints of the extremities, especially around the knee and ankle [2,3], while localization on the hand is less common. Tumor manifestation on fingers, in contrast to the carpal region, is extremely rare, which is the case here. Diagnosing synovial sarcoma can be extremely exhausting and time-consuming. Studies show that the average period from the appearance of the first symptoms to the correct diagnosis is 2 to 5, and sometimes even up to 20 years [1, 4]. The main symptom is a slow-growing subcutaneous tumor, and in addition, there is a pulsating pain that is also insufficiently specific for making the diagnosis [1, 3]. Histopathological verification is the gold standard for determining the diagnosis. A tumoral change can rarely be seen on an X-ray image. Magnetic resonance imaging (MRI) can be used as an important tool in establishing the diagnosis, but also in determining the limits of the change and the margins of the sur-

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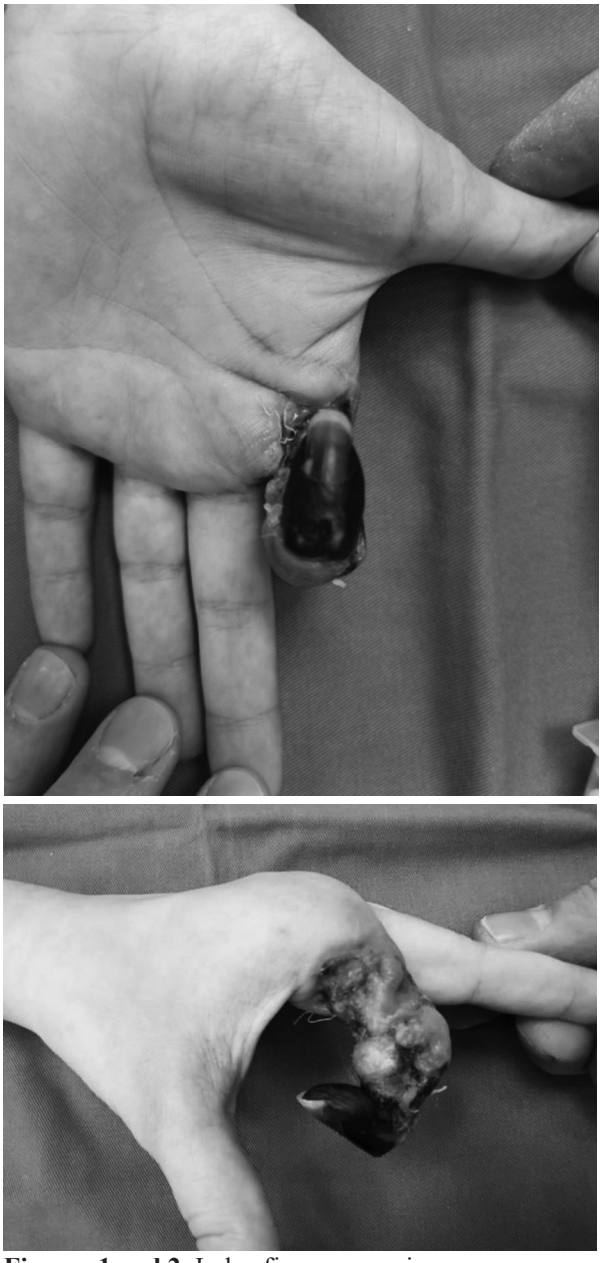
Abbreviations
MRI – magnetic resonance imaging
HP – histopathology

gical resection. Current therapy consists of multimodal treatment that includes radical surgical excision, systemic and perfusion chemotherapy, as well as radiotherapy [3,6]. This case report has a long and unusual path to the accurate diagnosis, as well as the unusual localization, and unfortunately unfavorable course too. It aims to draw the attention of clinical doctors and pathologists, to facilitate faster diagnosis of synovial sarcoma, and to make the relevant data available that could possibly be used in setting guidelines for the diagnosis and surgical resection.

Case report

A 20-year-old female patient came the first time to the outpatient department of the Clinic for Plastic and Reconstructive Surgery of the University Clinical Center of Vojvodina due to subcutaneous tumor and thickening of palmar skin at the level of the proximal interphalangeal joint of the index finger. Inspection of the medical documents revealed that the symptoms such as hardening and unusual feeling dated back 5 years, when excision with histopathology (HP) verification was performed on several occasions. Each time, the findings indicated a benign change - cutaneous fibrous histiocytoma (CFH). Re-excision of the change with HP verification was indicated in our facility, when the histopathology findings showed a dermatofibroma (plaque-like CD34+ dermatofibroma). During further treatment, the patient had complications such as wound dehiscence and infection, which were treated with local and systemic antibiotic therapy, to which an adequate response was obtained. After the wound complete healed, the patient was referred for physical therapy. A control X-ray examination of the hand and an ultrasound of the soft tissues in the scar area were advised in further follow-up. The patient failed to appear for the scheduled follow-up examination.

Two years later, the patient came for examination due to pain, with severe necrosis of the index finger and signs of local infection – purulent discharge and complete stiffness of the second finger. Active mobility in any of the three index joints was impossible **(Figures 1 and 2)**. After the examination, a decision was made to amputate the patient's index finger under general anesthesia. Intraoperative observation showed that the metacarpophalangeal joint was also affected by necrosis, and the index finger was amputated with partial resection of the head of the second metacarpal bone. The resected part was sent for HP verification, when the diagnosis of synovial sarcoma was obtained after several revisions and mutual consultations of several pathologists **(Figure 3)**, although it was initially described as a benign tumor (histiocytoma, dermatofibroma). The histopathology description read: "Tumor cells are spindle-shaped and arranged in bundles, bands and smaller swirls. Blood vessels are irregularly shaped, branched in places. The tumor tissue spreads along the hypocellular c+ connective tissue in the tendon. Immunohistochemical tissue is positive for vimentin, CKAE1/AE3, EMA and focally for CD56. Negative for SMA, S100 and CD31. The finding is in favor of a synovial sarcoma." Macroscopically, the color of the tumor was black-grey. The bone marrow of the second metacarpal bone was free of tumor elements. The free tumor margin was at least 1.5 cm. The postoperative period passed without complications. The patient



Figures 1 and 2. Index finger necrosis
Slike 1 i 2. *Nekroza kažiprsta*

Figure 3. Low magnification (10x) photomicrographs of hematoxylin and eosin-stained sections showing numerous cells packed in bundles and whorls
Slika 3. *Mikrofotografije niskog uvećanja (10x) hematoksilin i eosin obojeni delovi pokazuju brojne ćelije upakovane u vijuge i snopove*

was then presented to the Oncology Committee for Soft Tissue Tumors of the Institute of Oncology of Vojvodina, which indicated further follow-up.

Less than a year after the last operation, the patient noticed hardening in the scar area and felt pain. She was referred for an MRI, which showed a vaguely limited signal characteristic of pathologically altered tissue with propagation through the distal parts of the second metacarpal bone **(Figure 4)**, and further operative treatment was indicated. The patient underwent reamputation of the second metacarpal bone and capsulectomy of the third metacarpophalangeal joint under general anesthesia **(Figure 5)**. The wound healed properly. The patient underwent physical therapy, and disease propagation was obtained as a HP finding, with clear margins again. In the course of further diagnostics, there were no signs of dissemination of the underlying disease on the computed tomography of the chest, abdomen and pelvis, which were performed after the operation. The patient was presented again

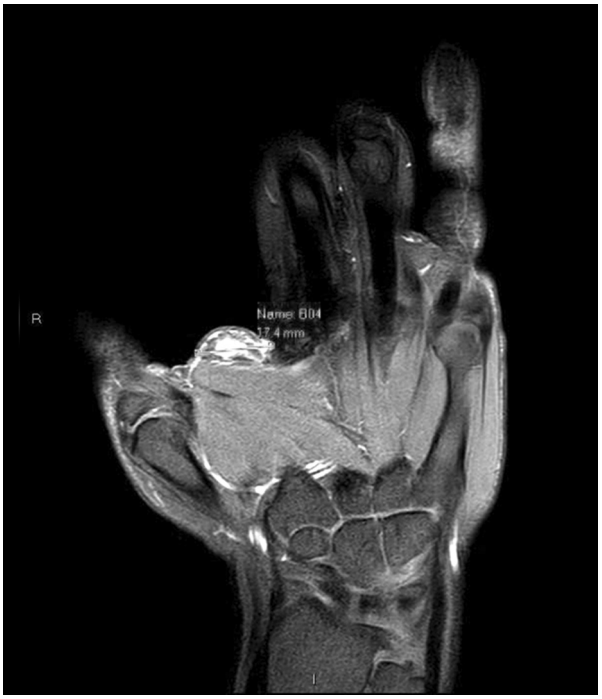


Figure 4. Disease propagation on the second metacarpal bone
Slika 4. *Propagacija bolesti na drugu metakarpalnu kost*

to the Oncology Committee for Soft Tissue Tumors, which indicated 25 cycles of radiotherapy that were fully implemented. No local recurrence or systemic dissemination of the disease was recorded in further control examinations and repeated diagnostics, and further follow-up was indicated.

Over a year after the second operation, during the control examination, small multiple skin lesions were

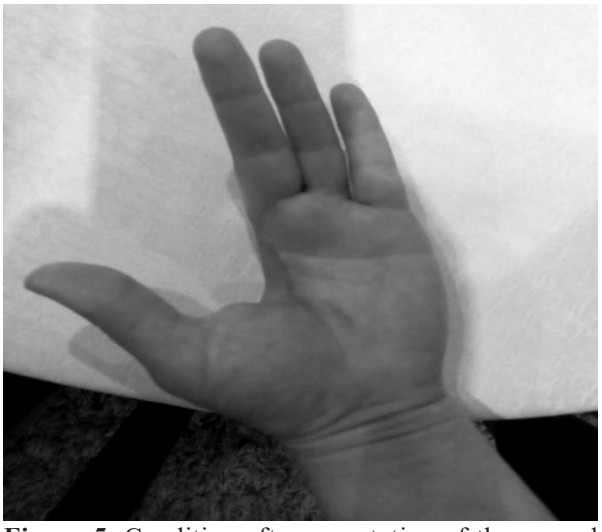


Figure 5. Condition after amputation of the second metacarpal bone and partial capsulectomy of the third metacarpophalangeal joint
Slika 5. *Stanje nakon amputacije druge metakarpalne kosti i parcijalne kapsulektomije trećeg metakarpofalangealnog zgloba*

found on the palm in the projection of the first metacarpal bone with a diameter of up to 5 mm. Similar changes were observed in the area of the left axillary region. Enlarged lymph nodes in the axilla were not observed. Biopsy of the changes under local anesthesia with HP verification was recommended, which the patient refused due to her early pregnancy. A delayed biopsy was performed three months later, and the HP showed a ring granuloma, DDx necrobiotic interstitial granulomatous dermatitis, without signs of the previously diagnosed tumor elements. Enlargement of the regional lymph nodes of the axilla was observed during the control examination. The Council of Gynecology Specialists made a decision to terminate the pregnancy due to high possibility of the paraneoplastic syndrome, which is why the patient was hospitalized at the Clinic of Gynecology and Obstetrics, University Clinical Center of Vojvodina. During her stay at the Clinic, the patient developed left arm swelling with severe pain in the shoulder joint area and a urinary infection, accompanied by high values of laboratory markers of inflammation. After discharge, an MRI of the chest with the shoulder joint was performed, where a massive tumor bony infiltration of the glenoid, the proximal humerus, clavicle, acromion and the left sternoclavicular joint was observed **(Figure 6)**. The MRI also showed ipsilateral infiltration of the lung parenchyma, as well as pathological changes and enlargement of the left axillary lymph nodes. The patient was again referred to the Oncology Committee that will decide on further systemic treatment, given that further surgical treatment is not indicated at this time.

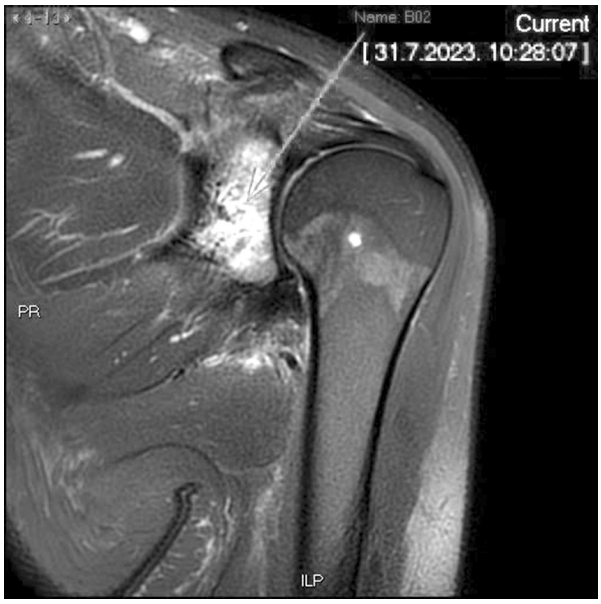


Figure 6. Massive bony infiltration of the glenoid, proximal part of the humerus, clavicle, acromion and the left sternoclavicular joint.
Slika 6. *Masivna koštana infiltracija glenoida, proksimalnog dela humerusa, klavikule, akromiona i levog sternoklavikularnog zgloba*

Discussion

Synovial sarcoma is a highly malignant tumor, rarely occurring in clinical practice, with an incidence of 1.42 per million adults in the United States [7]. Synovial sarcomas in the hand region have a very low incidence compared to other localizations, with a frequency of 4-8.5% in different studies [2, 8, 9], while fingers are affected less often than hands [10]. However, in a Serinelli S, et al. study, there was an increase in the number of synovial sarcomas of the hand, especially on the palmar side of the fingers [1]. It can be seen in the above case that the tumor was not recognized for a long time, and that the histopathology diagnosis of benign lesions combined with the patient's lack of motivation affected the unfavorable course of the disease. This case is extremely unusual given that the diagnosis of the malignant disease was made by at least three different pathologists after four histopathology analysis that showed the benignity, which can be explained by the histopathological similarity of this malignant tumor with benign changes. Benign lesions such as histiocytoma, dermatofibroma or annular granuloma may correspond to this tumor according to the pathohistological characteristics. Therefore, it is sometimes necessary to perform additional diagnostic methods, primarily immunohistochemical analysis, especially for the recurring changes, in order to establish a differential diagnosis. The study by Serinelli also shows that the tumor was initially misrecognized as a benign change such as abscess and arthritis in about 30% cases [1]. Despite their name, synovial sarcomas do not arise from synovium, their histogenesis is still unclear [11, 12]. The Naka N, et al. study suggests that the origin of this tumor may be from a multipotent mesenchymal stem cell [13].

During the excision of the change, one should consider the specificity of localization and the fact that fingers, being a functionally extremely important part of the body, have limited possibilities for skin and soft tissue reconstruction, which makes it difficult to achieve wide surgical margins [14]. Radical surgical excision was the treatment of choice in most studies. However, adjuvant radiotherapy and chemotherapy were used in 50% of cases [1], while in another study, 30% of patients received radiotherapy and only 5% received immunotherapy [8], which indicates the increasingly widespread use of adjuvant chemo- and radiotherapy after surgical excision of such changes. Damron et al. believe that standard therapy, in addition to radical excision, should always include adjuvant radiotherapy or chemotherapy [15]. Radiation therapy can improve local disease control [16]. Davis et al. observed that the choice of initial treatment affects the final oncological outcome of the disease [17]. The recurrence rate in the studies was from 33 to 80% of cases, depending on the literature [1, 6], which indicates a high degree of malignancy of this type of tumor.

Conclusion

After analyzing the clinical course of this case, it is necessary to emphasize that synovial sarcoma is very difficult to recognize in the early stages of the disease and that pathologists and clinical doctors should always suspect this type of tumor, which often gives unfavorable outcomes due to late detection. Examples from this and other mentioned papers indicate that one cannot always fully rely on histopathology verification and that the clinical course is also an important part, especially in the cases where changes recur. Early detection of this type of tumor enables a more favorable course and outcome of the treatment of these patients. It is necessary to consider the initially more radical treatment of such tumors due to the aggressiveness they show. Further evaluation and differentiation of the pathohistological characteristics of synovial sarcoma from tumors with similar characteristics and appearance in the clearest cases will allow for easier recognition of this type of malignant mesenchymal tumor in the initial stages of the disease, which will significantly affect the treatment prognosis. Also, it is necessary to emphasize the importance of health education of the general population, which could improve compliance.

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