

MANIFESTATION OF AN EXTRASKELETAL CHONDROSARCOMA IN THE FOOT

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INTRODUCTION

The first case of a chondrosarcoma of the foot was published in 1933 by Schreiner and Wehr (1). The majority of soft-tissue masses of the foot are of either inflammatory or reactive origin and very rarely are they real neoplasms. The chondrosarcoma represents less than 10% of all primary and secondary malignant bone tumors. The extraskeletal manifestation is even less common, although exact data for the incidence are not available. According to histological appearance the tumors are subdivided into a well differentiated mesenchymal and myxoid form. The tumors seem to develop *de novo* in the soft tissue of the extremity (2), however, other localizations such as synovia (3), heart (4), lung (5), and meninges (6), have been described. Undifferentiated and clear-cell chondrosarcoma are not found in the soft tissue outside of the bone (7, 8).

Extraskeletal myxoid chondrosarcoma are detected frequently in men older than 40 years. The tumor is most frequently located in the soft tissues of the extremities and is characterized by small round or elongated cells in a beaded positioning, surrounded by plenty of myxoid matrix (9).

Patients with an extraskeletal chondrosarcoma show symptoms very late, because the tumors are growing slowly (10). As long as the tumors do not infiltrate the bone or metastasize, pain develops very late. Radiographic evaluation

does not show a specific pattern and only sometimes demonstrates calcified soft tissue masses. Additional investigations by computed tomography or magnetic resonance imaging (MRI) are critical.

CASE REPORT

A 38-year-old woman presented with a subcutaneous swelling between the fourth and fifth metatarsals. The swelling had been increasing for 4 months. No other pathological findings were present. The MRI, which was taken at the first office visit revealed a cystic and septed process with a dimension of 1.3 x 2.0 x 0.8 inches. It was located between the shafts of the fourth and fifth metatarsals, and reached from dorsal to plantar without infiltration of the soft tissues or the bone (Figures 1-3).

A needle puncture revealed a gelatinous mass and the patient was advised to have the "ganglion" taken out if pain occurred. Two days later, the patient returned and requested surgical removal. In surgery, the soft tissue mass was resected en bloc, including the puncture site and the pseudocapsule. Clinically, the mass appeared to be more solid than a ganglion (Figures 4 and 5). The histopathological report described an extraskeletal myxoid chondrosarcoma of the foot. It was graded low-malignant (grade 1) (Figures 6 and 7).

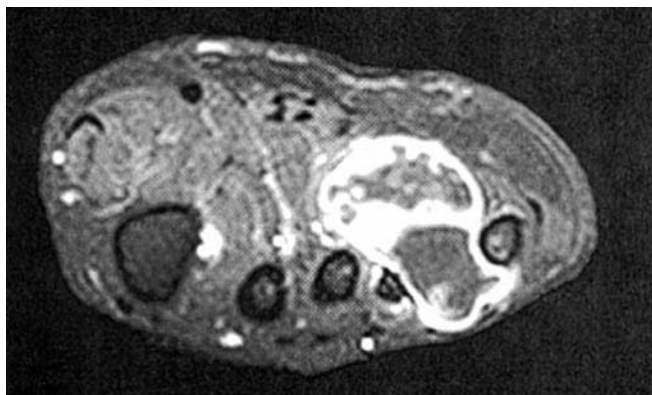


Figure 1. Coronal magnetic resonance image that shows both solid and cystic formations.

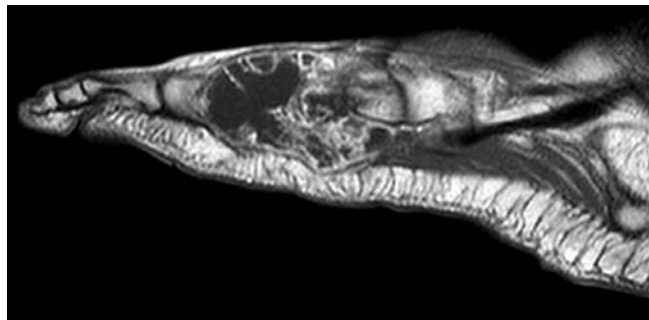


Figure 2. A pseudocapsule is visible in the sagittal view.



Figure 3. The transverse plane shows the displacement of the soft tissues.

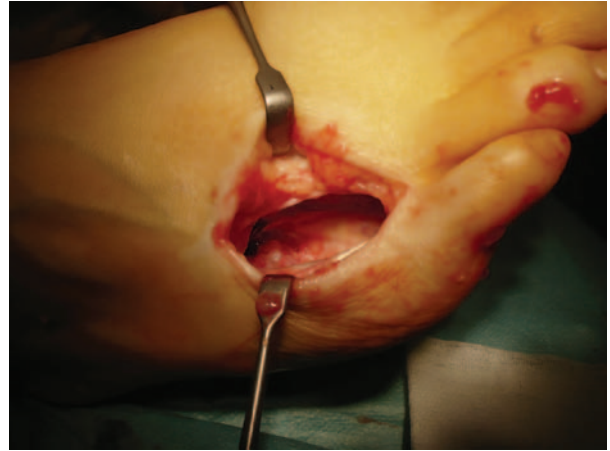


Figure 4. En bloc resection of the tumor from the fourth interspace.

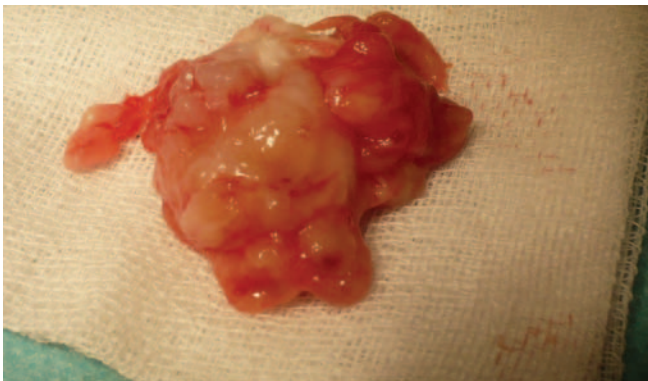


Figure 5. The resected mass.

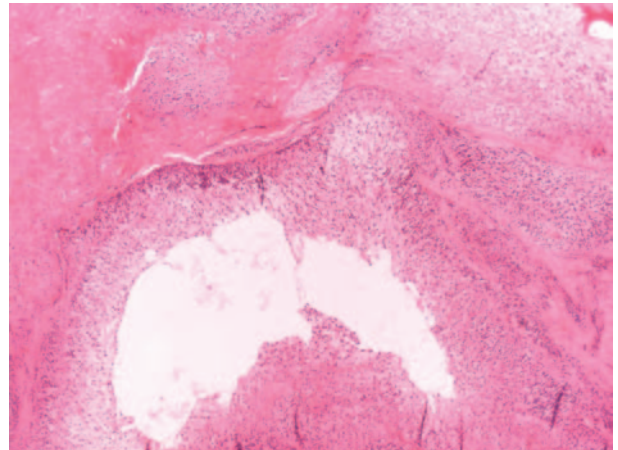


Figure 6. Histology of the tumor with nodular structures with myxoid areas and a pseudo cystic lesion (HE stain, magnification x25).

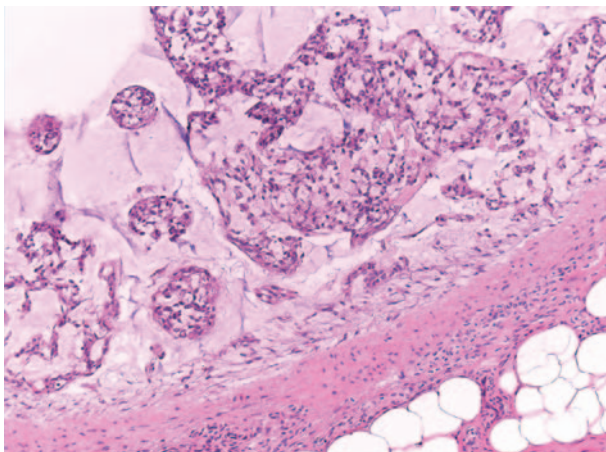


Figure 7. Cells of the tumor embedded in myxoid fluid (HE stain, magnification x100).

After surgery the patient showed primary wound healing and was referred to a center for oncology for further staging and surgical treatment. Later, the University Hospital of Lübeck decided to resect the fourth and fifth rays completely. The postoperative course was uneventful.

DISCUSSION

Several soft tissue formations of the foot have been described. Enzinger and Weiss (11) described a variety of 82 different soft tissue tumors distal to the knee joint. Some of the tumors, like ganglions or epidermal cysts, may resemble malignant neoplasms. The lesions located in the foot are relatively rare and the clinician might have difficulty gaining

experience with these tumor-like lesions. Each lesion should be classified “malignant” unless proven otherwise because the initial therapy is critical for the result of the treatment.

A retrospective analysis by Kirby et al (12) showed an analysis of 83 patients with tumors of the foot. Predominantly these lesions were benign (87%). Out of the benign tumors 29% were classified as ganglions. In the series were 2 extraskeletal myxoid chondrosarcomas with a low- or mid-grade malignancy. The histopathological evaluation is still a challenge. A needle puncture reveals the correct diagnosis in 67% of the cases. If a metastasis or a recurrent tumor is present, the accuracy is 86% (13). In a study by Rinas et al (14) puncture cytology could not identify any malignant cells.

It is imperative to avoid an incisional biopsy; an en bloc resection should always be preferred. The tumors should always be excised with the surrounding connective tissue capsule (10), which might be connected to the surrounding tissue with different intensity. The surgical approach should be planned in a way that allows the incision to be extended in case there is a need for revisional surgery, according to the guidelines of tumor surgery of the foot.

In Kirby’s study, the age distribution of malignant soft tissue tumors showed an accumulation between the ages of 10 and 40 years, and older than 60 years of age. Synovial sarcomas show a peak between the fifth and sixth decades of life (15), and sarcomas in general seem to occur more frequently after the age of 55 years (16). Extraskeletal chondrosarcomas do not have an age-specific incidence. Goldenberg et al reported the youngest patient to be 2 years old and the oldest patient to be 73 years of age (17).

Extraskeletal chondrosarcomas seem to occur twice as often in the male population. Wu et al (10) found 66% of the population in their study to be men. Regarding the incidence of soft tissue sarcomas, other studies showed a preference toward men as well (18).

Regarding the location of the sarcomas, there is a preference for the ankle joint. This is not surprising, as 45% of the reported malignant tumors were of synovial origin (12).

The differential diagnosis of any space-occupying process of the foot could be the extra-articular synovial chondromatosis, among others. This is a very rare tumor, which is characterized by masses of cartilage originating from the altered synovia. This tumor is therefore mostly located intra-articularly; however, it can also occur as an extra-articular process especially at the foot (19). Radiographic imaging may show very aggressive erosive lesions of the adjacent bones, which may resemble a chondrosarcoma (20). Radiographic evaluation mostly reveals a localized soft

tissue mass (10). Both benign and malignant tumors may demonstrate unusual calcifications, which may lead to secondary erosions of the adjacent bones. Subchondral cystoid formation should remind the clinician of villonodular synovitis. There is not a characteristic radiographic finding of the extraskeletal myxoid chondrosarcoma (17). The MRI in the reported case showed cystoid, septed structures that seemed to be typical for a ganglion. A giant-cell tumor could be excluded and the MRI did not show any infiltration of the soft tissue and the bone. Neither the gender of the patient nor the symptoms nor the size of the lesion could be used as criteria for the diagnosis of the tumor. Surprisingly, some patients report a long history of the tumor prior to referral for surgical treatment (10, 12, 17). Therefore chronicity does not exclude malignancy.

The symptoms of the reported patient did not show any criteria for malignancy, which is in line with other studies that show a slow increase of a pain-free soft tissue mass over a long period of time. Only if the tumor infiltrates adjacent nerve structures, will symptoms occur earlier.

The en bloc resection is the surgical treatment of first choice. Amputation should be performed in the case of a large tumor with intra-articular proliferation or with recurrent tumors. The extra-skeletal chondrosarcomas show a 50% rate of recurrence, which can occur even 15 years after the first excision (17), or on average after 2.6 years (10). The lungs were the most frequent location of the metastasis approximately 3.2 years after the first treatment (10), followed by intraabdominal locations, and other skeletal locations. The 1-year survival rate is reported to be 82%, and the 5-year survival rate 81%. The tumors are not responsive to radiation or chemotherapy (21).

SUMMARY

The reported case shows that even a supposedly harmless tumor can be a rare malignant process. The extraskeletal myxoid chondrosarcoma seems to affect the male population of middle-aged patients rather than female patients. The predilection site is the lower extremity. The symptoms are not obvious, with a slow progression of pain free swelling, and no specific radiographic findings.

The intraoperative findings include a septed tumor, which is embedded in a capsule with clear separation from the surrounding soft-tissue and bone. The rare tumor with high rates of recurrence needs to be treated stringently and should be treated in cooperation with an oncology center. Any neoplasm of the foot should be excised en bloc, as the imaging modalities are not always specific. The histopathology should be evaluated in an oncology center.

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