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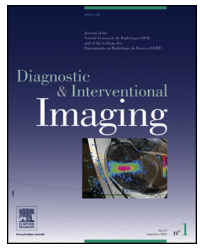

LETTER /*Muskuloskeletal imaging*

Imaging features of extraskeletal mesenchymal chondrosarcoma of the hand

Keywords Chondrosarcoma; Soft tissue; Hand

Dear Editor,

Mesenchymal chondrosarcoma of the soft tissues represents a very rare tumor, even rarer than its skeleton counterpart. It grows rapidly and if untreated may reach a large size. Histological diagnosis may be difficult as it often mimics other tumors such as Ewing sarcoma, hemangiopericytoma, small cell osteosarcoma, synovial sarcoma, etc. Visualization of areas of cartilaginous differentiation may help with diagnosis. For its severe prognosis, surgical treatment must be aggressive whether associated or not to radiation or chemotherapy. We here describe the case of an 18-year-old boy with extraskeletal mesenchymal chondrosarcoma (EMC) of the hand. To our knowledge, this is the first case of EMC in this site described in literature.

Case report

An 18-year-old boy with swelling of the right hand presented at our institution in April 2013. The mass appeared about 3—4 months before with no significant changes since then. An incisional biopsy was performed elsewhere with a histological diagnosis of EMC (a review of the slides confirmed this diagnosis). At physical examination, a painless mass, non-mobile, measuring 4 × 3 cm, was observed at the medial border of the thenar eminence. The previous surgical incision had already healed. Radiographs of the hand showed a calcified mass between the third and fourth metacarpal bone with no involvement of the bone itself (Fig. 1A). MRI confirmed a soft tissue mass underneath and in contact with the flexor tendon sheath with dense calcification (mixed high and low signal in T2, black pepper sign) and non-involvement of the bone (Fig. 1B—C).

The total body CT scan previously performed elsewhere was otherwise normal.

In May 2013 the patient underwent surgery. Due to the complex anatomy of the hand and the close location of the tumor to the tendon sheath of third and fourth metacarpal bones, a marginal excision was performed preserving the tendons. The histological examination of the specimen confirmed EMC. Translocation t (8; 8) was found (Fig. 2A—D). Postoperatively no vascular deficiencies were present but only slight hypoesthesia of the 4th finger. The patient refused postoperative chemotherapy or radiation.

At one year follow-up the patient is free of disease.

Discussion

Mesenchymal chondrosarcoma was first described more than 50 years ago by Lichtenstein and Bernstein [1]. It represents a very rare subtype of chondrosarcoma (about 1% of all chondrosarcomas) with poor survival and a strong tendency to local recurrence and distant metastases especially in lungs and bones. The central nervous system represents the most frequent extraosseous site, especially in the meninges. About 30—50% of EMC occur in the soft tissues [2]. It has a slight predominance for the female gender [1,2]. As reported by Louvet et al. in their review of literature the most frequent localization in the soft tissues is the thigh with only one case localized in the hand [2].

Due to the rarity of the disease and its aggressiveness, a timely and correct diagnosis must be made. Radiographs and CT can reveal ''ring and arc calcifications'' and if present, bone involvement [3]. The typical MRI finding on EMC is the ''black pepper'' sign which represents areas of high intensity signals (non-calcified component) surrounding areas of low intensity signal (mineralized component). CT and MRI with contrast medium show a strong uptake. The association of this strong uptake with cartilaginous calcifications may be suggestive of the diagnosis.

Various benign and malignant tumors may show similar imaging characteristics, especially highly myxoid tumors (myxoid liposarcoma, myxoma), which present with a homogeneous or heterogeneous high intensity signal on T2-weighted MRI images [3]. However, in these cases, the myxoid component does not enhance strongly and quickly after injection with contrast medium. Other cartilaginous tumors (soft tissue chondroma or chondrosarcoma, extraskeletal myxoid chondrosarcoma, synovial chondrosarcoma) may contain cartilaginous calcifications, but lack the strong and quick enhancement. Their clinical and histological patterns are different and in most cases a differential diagnosis can easily be made.

Histologically EMC shows a biphasic pattern composed of undifferentiated small, round cells and islands of well differentiated hyaline/cartilage. In areas of undifferentiated tissue, cells may mimic a Ewing sarcoma, synovial sarcoma or hemangiopericytoma. This could render diagnosis difficult especially when the specimen analyzed is small and the areas of hyaline cartilage are not visualized. Differential

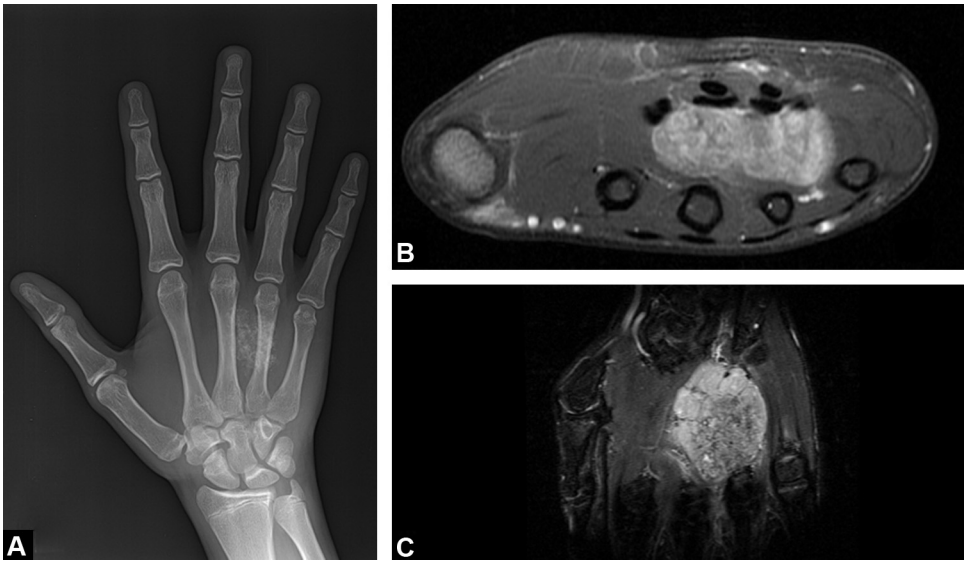

Figure 1. Radiographs (A) and fat-suppressed T1-weighted MR images in the axial (B) and coronal (C) planes after contrast medium injection of the hand show soft tissue calcifications, a strong uptake of contrast and the congruity of the lesion with the tendon sheath and metacarpal bone.

diagnosis may in some cases be facilitated by searching for specific translocations (like Ewing sarcoma or synovial sarcoma). Markers like S-100 protein, CD999 and Vimentin may be positive in EMC but are not specific. Muller et al. demonstrated that type II collagen is a selective marker for chondrocyte cell differentiation and therefore can be used to identify mesenchymal chondrosarcoma that lack overt cartilage formation [4]. Wehrli et al. reported that anti/Sox9 was positive in mesenchymal chondrosarcoma and negative in other small cell tumors [5]. Nakayama et al. demonstrated that fusion gene HEY1-NCOA2 is a reliable method for diagnosis of mesenchymal chondrosarcoma [6].

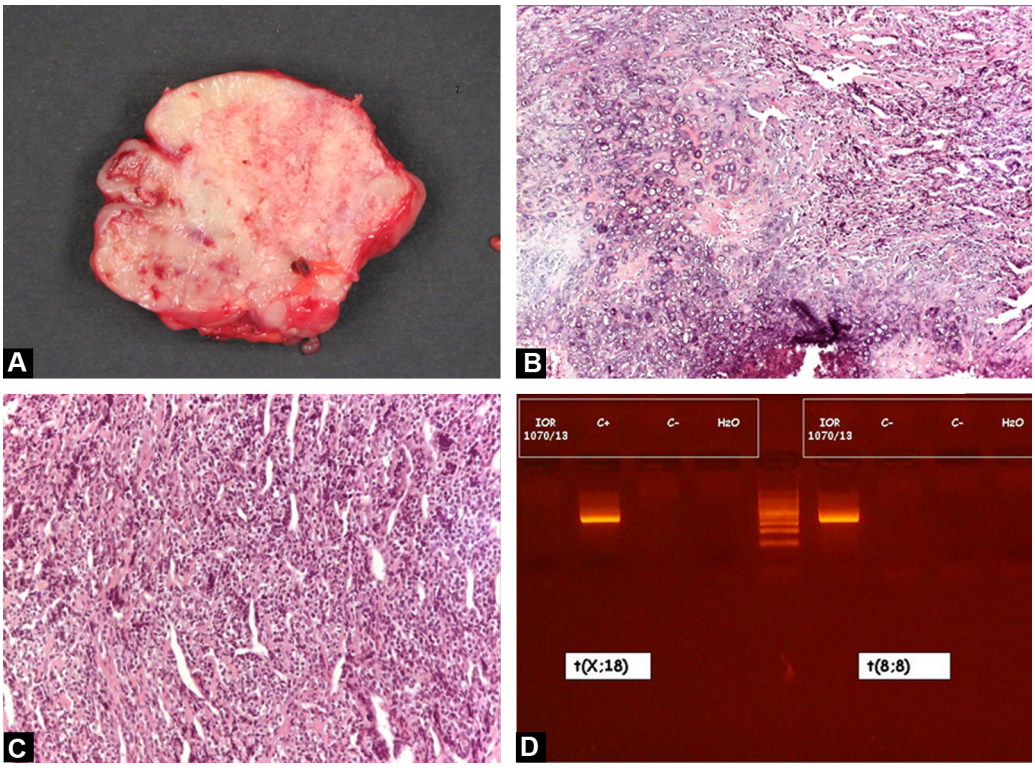

Figure 2. On gross analysis, the lesion appears soft and lobulated with well demarcated margins (A). On hematoxylin and eosin staining, mesenchymal chondrosarcoma is constituted of lobules of cartilage associated with coarse calcifications surrounded by malignant small cells (B, × 100 magnification). Morphologically, vessels are surrounded by small malignant cells which compress and deform them, producing the typical staghorn appearance (C, × 200 magnification). The diagnostic t (8; 8) translocation, that identifies the HEY1-NCOA2 fusion gene, was found using RT-PCR (D).

Chondrosarcomas represent a group of tumors usually resistant to chemo- and radiation therapy, probably for their slow growth, poor vascularity and large amount of extracellular matrix. However, MC is an exception. Many authors sustain the role of a doxorubicin-based combination therapy used in addition to surgery. Cesari et al. [7] observed a higher overall survival rate at 10 years in patients who received chemotherapy compared to those who did not (31% vs 19%). Kawaguchi et al. [8] in their study demonstrated that adjuvant radiation therapy improved local recurrence-free survival.

Conclusion

EMC represents a rare malignant chondrogenic neoplasm. On imaging, the association of cartilaginous calcifications together with a strong uptake after contrast medium injection on CT or MRI may be suggestive of the diagnosis [9]. Histologically EMC presents with a biphasic pattern: lobules of well differentiated cartilage and proliferation of small undifferentiated round cells that may mimic other malignant tumors. Although chemotherapy and radiotherapy are important in obtaining a better disease-free survival and local control, surgery remains the treatment of choice.

Disclosure of interest

The authors declare that they have no conflict of interest concerning this article.

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