

October 2016 Case of the Month

Mesenchymal chondrosarcoma

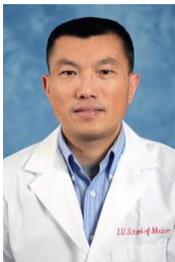
A 22-year-old male with right ocular proptosis and progressive right vision loss.

Contributed by:



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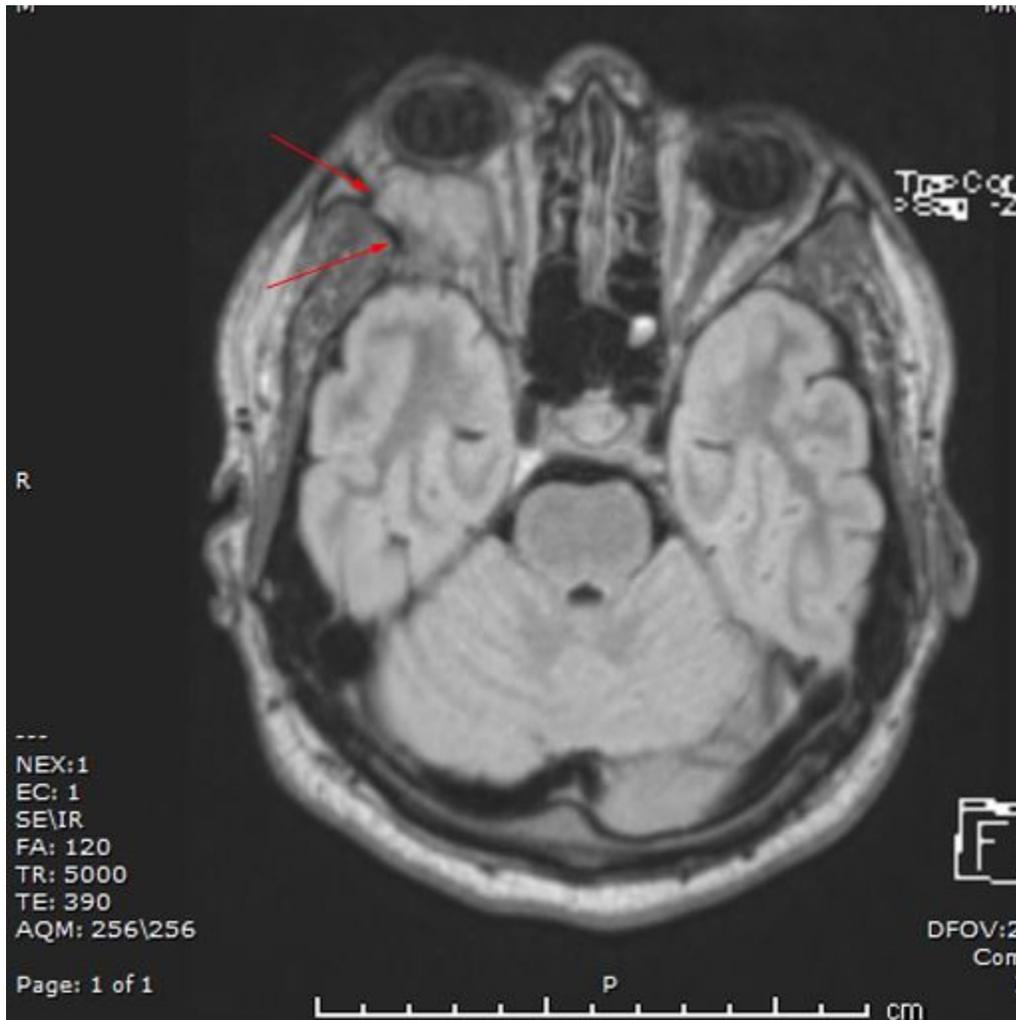


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Clinical History:

The patient is a 22-year-old male who presented with right ocular proptosis and progressive right vision loss over a few weeks. An orbital Magnetic Resonance Imaging (MRI) revealed a 3 x 2.8 x 3.3 cm heterogeneously enhancing mass centered in the right lateral orbital wall (greater wing of sphenoid) with extensive involvement of the extraconal, conal, and intraconal spaces. Additionally, mass effect was seen on the right lateral and inferior rectus muscles with a mild effect on the right optic nerve, and an extension into the right temporalis muscle and right middle cranial fossa was observed. The radiology differential diagnosis included hemangiopericytoma versus osteosarcoma, or angiosarcoma (Fig. 1). A biopsy was performed.

Figure 1: Orbital Magnetic Resonance Imaging (MRI) showing a mass (red arrows) centered in the right lateral orbit.



Gross Examination:

Six irregular, gray-tan to red-brown soft tissue and bone fragments were identified.

Microscopic Examination:

Permanent hematoxylin and eosin (H&E) staining microscopic sections demonstrated a biphasic tumor (Fig. 2).

There were sheets of small round and spindle cells (Fig. 3) associated with nodules of hyaline cartilage that were both basophilic and eosinophilic in appearance (Fig. 4). Highly pleomorphic chondrocytes were identified (Fig. 5).

The tumor permeated fragments of trabecular bone with reactive woven bone formation (Fig. 6). The neoplastic matrix was cartilaginous.

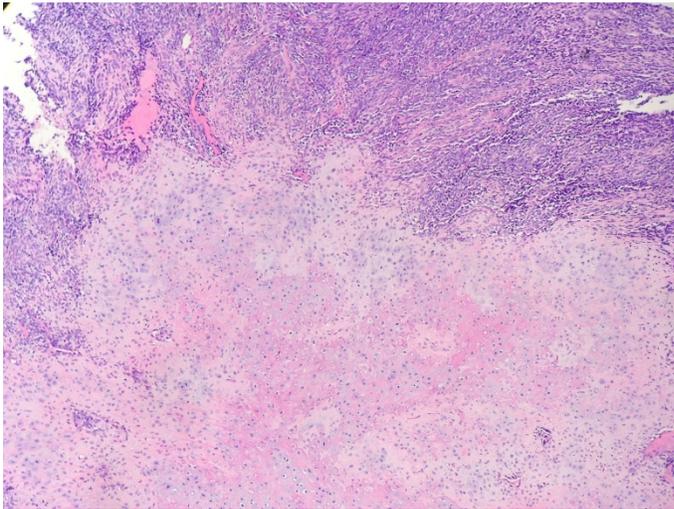


Figure 2: The tumor has two distinctive components. Sheets of primitive mesenchymal cells and hyaline cartilage. 100x

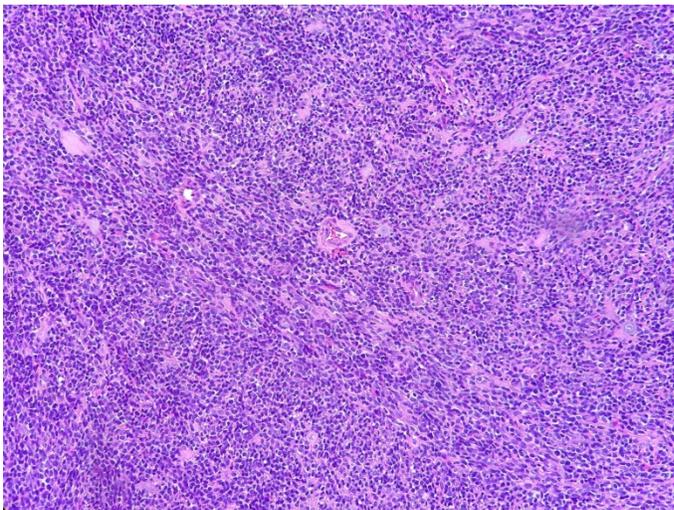


Figure 3: The mesenchymal component, sheets of small round and spindle cells. 200x

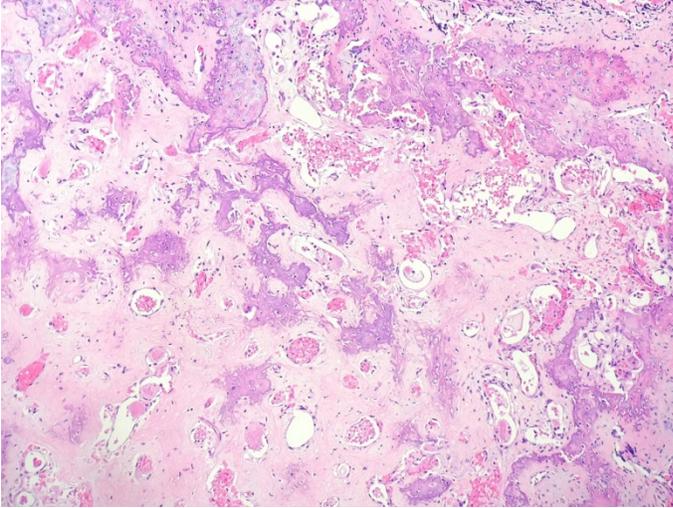


Figure 4: The cartilage component with basophilic and eosinophilic appearance. 100x

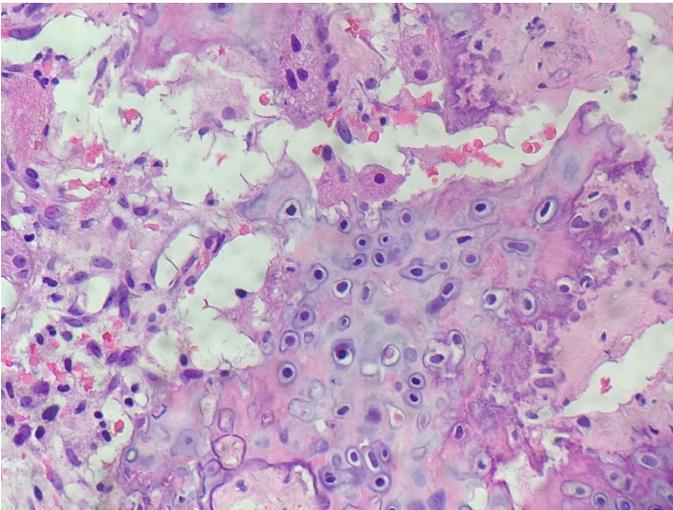


Figure 5: The cartilage component with highly pleomorphic chondrocytes. 400x

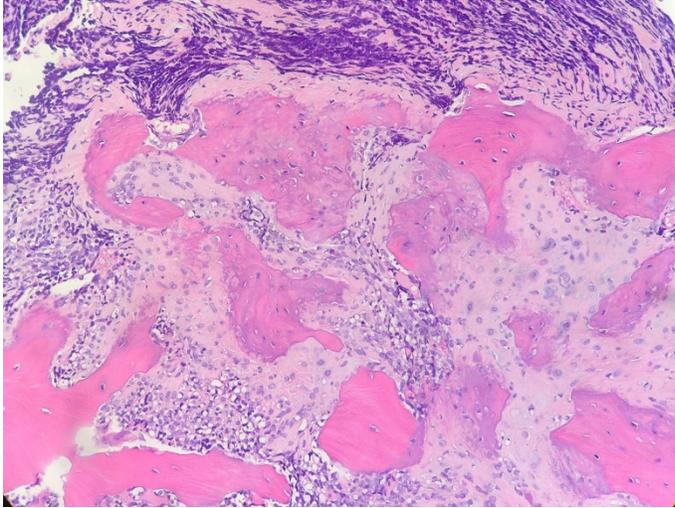


Figure 6: The tumor permeated fragments of trabecular bone with reactive woven bone formation. 200x

Discussion:

Mesenchymal chondrosarcoma (MCS) is a malignant cartilaginous tumor composed of two distinctive components: sheets of primitive mesenchymal cells and interspersed islands of well-differentiated hyaline cartilage. The primitive mesenchymal component consists of undifferentiated round, oval, or spindle-shaped cells with hyperchromatic nuclei and scanty, poorly outlined cytoplasm. Usually, there is an abrupt transition of the mesenchymal component with well-defined nodules of well-differentiated, relatively low-grade appearing hyaline cartilage, frequently with central calcification and ossification. The amount of cartilage is highly variable. Osteoclast-like multinucleated giant cells may occasionally be seen.

Mesenchymal chondrosarcoma accounts for 3–10% of all chondrosarcoma diagnoses. It is preponderant in young adults 15 to 35 years of age with a slightly more frequent occurrence in females than in males. The tumor may also occur in young children [1]. The axial skeleton and femur are the most common primary sites for skeletal MCS while soft tissue MCS may affect meninges, and visceral organ involvement has also been reported in the literature [2].

Cells in the chondroid areas are positive for S100, while small cells of mesenchymal chondrosarcoma are positive for CD99. If the biopsy only shows the small cell component, differential diagnosis include Ewing sarcoma, mesenchymal chondrosarcoma, etc. Molecular study of t(11;22) should be helpful. This translocation has not been reported in mesenchymal chondrosarcoma. The HEY1-NCOA2 fusion has been described in MCS and can be used as a diagnostic tool in difficult cases [3].

The orbital lesions tend to produce exophthalmos, orbital pain, blurring of vision, and headaches. In the extremities, this tumor usually manifests as a painless, slowly enlarging mass situated in the musculature. Radiography frequently reveals a well-defined soft tissue mass,

often with irregular radiopaque stippling, arcs, or streaks, as result of focal calcification or bone formation in cartilaginous areas [4].

The recommended treatment is surgery with clear margins and (neo-) adjuvant chemotherapy in patients with localized disease. Surgery and chemotherapy has been associated with significant reduction in the risk of recurrence and death in these patients [1].

The prognosis appears to be poor, with the 10-year survival rate in the ranges of 27-67% [1].

References:

1. Frezza, A.M., et al., Mesenchymal chondrosarcoma: prognostic factors and outcome in 113 patients. A European Musculoskeletal Oncology Society study. *Eur J Cancer*, 2015. **51**(3): p. 374-81.
2. Doyle, L.A., Sarcoma classification: an update based on the 2013 World Health Organization Classification of Tumors of Soft Tissue and Bone. *Cancer*, 2014. **120**(12): p. 1763-74.
3. Wang, L., et al., Identification of a novel, recurrent HEY1-NCOA2 fusion in mesenchymal chondrosarcoma based on a genome-wide screen of exon-level expression data. *Genes Chromosomes Cancer*, 2012. **51**(2): p. 127-39.
4. Goldblum, J.R., et al., *Enzinger and Weiss's soft tissue tumors*. 6th ed. 2014, Philadelphia, PA: Saunders/Elsevier. xiv, 1155 p.