

Extra-skeletal Myxoid Chondrosarcoma (Chordoid Sarcoma)

The extra-skeletal myxoid chondrosarcoma is a very rare soft tissue tumor in the deep muscle belly, occurring most often in the extremities in adults over 40 years of age. Males are affected twice as often as females. The tumor is slow growing and may cause local pain and tenderness. Common locations are the thigh, popliteal fossa, and shoulder girdle. It presents with the clinical appearance of a myxoid liposarcoma. Pathologically, the tumors are gray to tan, depending on the amount of hemorrhage into the tumor. Because hemorrhage often occurs, this tumor can be mistaken for a hematoma. Histologically, the tumor has a myxoid appearance with chords and nests of anastomosing cells that have a chondroblastic appearance. The histology is very similar to that of chordoma of the sacrum. The tumor is considered low grade in most instances; it grows slowly but has the potential for local recurrence and pulmonary metastases in about one-third of cases. Treatment consists of aggressive wide resection or amputation, if need be, followed by local radiation therapy. Chemotherapy is not indicated.