Dedifferentiated Chondrosarcoma

Of all the chondrosarcoma variants, by far the most malignant and potentially fatal is the dedifferentiated chondrosarcoma that accounts for approximately 5-10 per cent of all chondrosarcomas. It arises most likely as a result of a second mutation within a pre-existing low to intermediate grade chondrosarcoma, resulting in the formation of a malignant fibrous histiocytoma or osteosarcoma immediately adjacent to the lower grade chondrosarcoma. Histologically, the low and high-grade portions of this tumor are geographically separated by sharp margins. The tumor usually occurs in patients between the ages of 50 and 70 years in the same areas where primary central chondrosarcomas are found – the pelvis, femur and proximal humerus. The characteristic radiographic appearance is that of a flocculated calcific lytic lesion arising from the central area of bone with an adjacent area with more extensive infiltration into the surrounding cortical bone. There is no evidence of calcification in the high-grade portion of the lesion and it typically breaks out through the cortex into the subperiosteal space.

The prognosis for this variant of chondrosarcoma is extremely poor, most patients dying from metastatic disease within one or two years after the diagnosis is established. Adjuvant chemotherapy and radiation therapy are not very effective, mainly because of the older age group in which the tumor occurs. The primary treatment modality is wide surgical resection.