Clear Cell Sarcoma

The clear cell sarcoma is thought to be a deep, non-cutaneous variant of the pigmented melanoma. It is a very rare tumor affecting more females than males. It is typically seen between the ages of 20 and 40 years. It usually occurs in tendon sheaths and fascial planes, especially around the foot and ankle area, and has a clinical appearance similar to the synovial sarcoma with which it can be confused. The tumor usually begins as a slow-growing lump that has a benign appearance but after a period of several years the tumor will start to grow more rapidly and become painful. It has a high potential to metastasize to local lymph nodes and to the lung. Approximately 50 per cent of patients with this tumor will be dead at five years. The microscopic appearance is similar to that of the epithelioid sarcoma, especially if melanin is not found in the specimen. Treatment usually consists of wide resection, if possible, and radiation therapy, but a high local recurrence rate is common because of its location in extracompartmental structures such as tendon sheathes. If the tumor does recur, amputation is carried out for local control of the disease. Because of the poor prognosis with clear cell sarcoma, adjuvant chemotherapy has been advised but the response is usually not beneficial.