Clear-cell sarcoma of the soft tissue - a rare diagnosis with a fatal outcome.

Abstract:

Clear-cell sarcomas account for less than 1% of all soft tissue tumours. They most often occur in middle-aged adults as a deeply located lesion with predilection to the tendons and aponeuroses. The aim of the present study was to show possible influencing factors on the outcome after surgical treatment in a detailed case series. We reviewed the medical records of 11 patients with the diagnosis of a clear-cell sarcoma of the soft tissue. These cases were analysed with regard to age, gender, localisation, tumour size, recurrence free survival and overall survival. A minimum follow up of 12 months was achieved. The mean age at the point of diagnosis was 47.9 years. Metastases occurred after a mean of 19.2 months. In the cases with a tumour diameter>5 cm, metastases occurred earlier. When treated in a specialist centre, metastases occurred later. Patients died a mean of 18.4 months after developing metastatic disease. Patients with tumour size>5 cm at the point of primary diagnosis died earlier than patients with a tumour size<5 cm. It is important to detect clear-cell sarcomas as soon as possible and the final surgical treatment should be performed in a centre familiar with the treatment of soft tissue tumours not only to prolong overall survival, but also to treat the patient in a multiprofessional team.