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Clear Cell Sarcoma of the soft tissues. A retrospective analysis of 35 cases

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BACKGROUND: Clear cell sarcoma is a rare soft tissue tumor with a poor prognosis. This lesion shows morphologic similarities to malignant melanoma but has a distinct genetic background. Early diagnosis and initial wide excision are essential for a favourable outcome. The objective of this study is to determine the clinical, pathologic and genetic features of this tumor, in order to recognize it at an earlier stage and improve treatment and prognosis.

METHODS: A retrospective analysis was performed on 35 consecutive cases, treated at the Rizzoli Institute between 1979 and 2009.

RESULTS: There were 19 male and 16 female patients with an age ranging from 8 to 75 years (mean 40 years). Most of the tumors (24) were located in the lower extremity, 8 in the upper extremity and 3 in the trunk. Twenty-five patients (71%) had undergone previous treatments elsewhere. Thirty-one patients (89%) had localized disease at presentation, 2 had lymphnode metastases, one had lung metastases, and one had lung and bone metastases. Half of the tumors was more than 5cm in diameter. All but one patient underwent surgical excision of the tumor. Six patients (18%) underwent an amputation, 28 had limb salvage surgery performed. Surgical margins were inadequate (marginal or intralesional) in 4 cases. Mean follow-up was 64 months (range 0-311 months). Eight patients developed local recurrence, 17 patients had metastatic disease. At last follow-up, 19 patients had no evidence of disease, one patient was alive with disease and 15 patients died of disease. The overall survival rate was 58% at 5 years and 50% at 10-years.

CONCLUSIONS: Clear cell sarcomas are often unrecognized at initial presentation, causing diagnostic delay and inadequate treatments. We believe that early referral to a tertiary centre can improve outcome for patients with clear cell sarcoma. Wide surgical excision is the main treatment, radiotherapy is often used as adjuvant treatment for local control. The role of chemotherapy needs further investigation.

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