

CLEAR-CELL SARCOMA. RARE TYPE OF SOFT TISSUE SARCOMA

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Abstract

INTRODUCTION

Clear cell sarcoma is a rare but highly malignant tumour of soft tissues. Often appearing as a small tender mass in the deep tissues of the distal extremities with a different cytogenetic and natural history presenting high rates of recurrences and metastases.

In the literature, few studies discuss these tumours because it is very rare pathology.

MATERIAL AND METHODS

This is an observational study that included patients diagnosed in our Centre with clear cell sarcoma from 1999 to 2005, estimating the prevalence of these tumours in our hospital. Demographic data (sex and age) were collected and analysed the evolution of these cases and the rate of local recurrence and survival rate.

RESULTS

In our series, 6 patients were diagnosed with clear cell sarcoma. In our Centre, this type of tumour, presented a prevalence of 0.52%, in respect of all soft tissue sarcomas diagnosed.

A woman and 5 men, with a mean age of 50.5 years (19-86). Regarding the location of the tumour in 4 cases were in upper extremity (one wrist, 2 forearm, one elbow) and in 2 cases in lower limb (1 popliteal area, one thigh). Mean follow-up was 69 months (27-146).

In all cases, surgical resection was performed in the first two months after the pathological diagnosis of the previous biopsy. In 3 cases, adjuvant radiotherapy was performed and adjuvant chemotherapy in two cases. We noticed a 66.67% rate of local recurrence requiring additional surgery (2 amputations and 2 new resections). Also, we note lung metastases and systemic spread and survival rates of 50 to 33.34%.

CONCLUSION

In our series, we observed that clear cell sarcoma show considerable aggression with a high rate of disease recurrence and a low survival rate especially in older patients.

It is a condition to be considered for early diagnosis and treatment.