Soft tissue tumors: Malignant melanoma of soft parts

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Identity
Alias: Clear cell sarcoma of soft parts.

Classification
This tumour, initially described by Enzinger as "clear cell sarcoma of tendons and aponeuroses", is of uncertain origin, but its immunohistochemical profile shows its melanocytic nature; however it has no genetic relationship with the cutaneous malignant melanoma.

Clinics and pathology
Embryonic origin
Being of melanocytic origin, this tumour should be classified as a neuroectodermal tumour.

Etiology
Unknown.

Epidemiology
It is a very rare tumour representing a minority of all soft tissue sarcomas.

Clinics
The malignant melanoma of soft parts (MMSP) preferentially occurs in young adults, between ages of 20 and 40 years; the tumour develops mainly in the extremities, especially the legs (foot, knee, heel, ankle); it is usually deeply seated, and often bound to tendons and aponeuroses.

Pathology
The tumours show compact nests and strands of round or fusiform cells with a clear cytoplasm, separated by fibrocollagenous tissue often connected to adjacent tendons or aponeuroses; mitotic index is generally low; the cells of nearly all cases express S-100 protein and the melanoma-associated antigen HMB45.

Treatment
The treatment protocols vary greatly according to the institutions; however, the melanoma of soft parts is a highly malignant tumour which requires surgical excision combined with radiotherapy and/or chemotherapy.

Evolution
Many patients develop recurrences and regional and distant metastases, in lymph nodes, lung, and bones; in the series of Enzinger, the average time between diagnosis and recurrence was 2.6 years, between diagnosis and metastasis, 3.5 years.

Prognosis
The prognosis is poor; in the series of 115 patients studied by Enzinger, 46% had died; of the 62 living patients, 21 experienced one or more recurrences, and 7 had a metastatic disease.

Cytogenetics
Cytogenetics Morphological
This tumour is characterised by the presence of a chromosome translocation t(12;22)(q13;q12), which involves genes ATF-1, on chromosome 12, and EWS, on chromosome 22.

Genes involved and proteins
EWSR1
Location
22q12
Protein
RNA binding protein.

**ATF-1**

**Location**
12q13

**Protein**
Transcription factor.

**Result of the chromosomal anomaly**

**Fusion Protein**

**Description**
The chimaeric protein is composed of the N-terminal domain of EWS linked to the bZIP domain of ATF-1.

**Oncogenesis**
Binds to ATF sites present in cAMP-responsive promoters via the ATF1 bZIP domain and activates transcription constitutively, dependent on the activation domain (EAD) present in EWSR1.

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**References**


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This article should be referenced as such: