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Solid Tumour Section

Short Communication

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.

References

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