

Clear Cell Sarcoma of Soft Tissue: A Rare Malignancy Entity

Kenza Kandri Rody¹, Soumia Faras¹, Najat Elaattoul¹, Imane Boujguenna², Hanane Rais², Ouafa Hocar¹, Said Amal¹

¹Dermatology-Venerology department; Mohammed VI university hospital center; Marrakesh Morocco

²Pathological anatomy department; Mohammed VI university hospital center; Marrakesh Morocco

DOI: [10.36347/sjmcr.2021.v09i03.027](https://doi.org/10.36347/sjmcr.2021.v09i03.027)

| Received: 02.02.2021 | Accepted: 08.03.2021 | Published: 28.03.2021

*Corresponding author: Kenza Kandri Rody

Abstract

Case Report

Clear cell sarcoma (CCS) of soft tissue or soft tissue melanoma is a rare malignant tumor in young adults, preferentially affecting the lower limbs. We report a new observation of a 17-year-old patient, without a significant history, consulted for a nodular lesion of the left ankle, painless, appeared 06 months before, evolving in a context of general impairment. The clinical examination found a purplish nodule, about 2/2 cm, hard, fixed, associated with multiple nodular lesions along the homolateral lower limb, with the presence of homolateral inguinal lymphadenopathy. There was no associated motor or vascular disorder. The nodule ultrasound showed a mass of soft tissue, hypoechogenic, heterogeneous. The computed tomography of the ankle revealed a lesional process of the medial face infiltrating the tendon and ligament structures and encompassing the posterior tibial artery. Histological examination showed round cell malignant tumor proliferation. The immuno-histochemical supplement showed an expression of the S100 protein and monoclonal antibody HMB-45 by tumor cells. The biopsy of an inguinal node affirmed its metastatic nature. The anatomo-clinical confrontation allowed to retain the diagnosis of clear cell sarcoma. The extension assessment made of a chest-abdominal-pelvic computed tomography showed metastatic lymph nodes intra and retroperitoneal. The patient was given chemotherapy (ifosfamide-adriamycin). Sarcoma with clear soft tissue cells or melanoma of the soft parts is a rare malignant tumor. It occurs most often in young adults at the lower limbs. It is a firm subcutaneous mass, slowly progressive, well limited, usually deep attached to tendon and aponeurotic structures. The positive diagnosis is based on histology coupled with immunohistochemistry. It can be helped by molecular biology in case of doubt diagnosis with melanoma. Its poor prognosis requires early and adequate management.

Keywords: Clear cell sarcoma of soft tissue, Tumors, soft tissue melanoma.

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INTRODUCTION

Clear cell sarcoma (CCS) or soft tissue melanoma is a rare entity. It is a malignant tumor of young adults, preferentially affecting the deep soft tissues of the lower limbs. We report a new metastatic CSC observation.

CASE REPORT

A 17-year-old patient, with no significant history, consulted for a nodular lesion of the left ankle, painless, appeared 06 months earlier, following a minimal trauma, evolving in a context of altered general condition (Figure 1). The clinical examination found a purplish nodule, about 2/2 cm, hard, fixed, associated with multiple nodular lesions (erythematous, hard, fixed and painless) without local inflammatory signs of the antero external of the homolateral lower limb of which

the largest measured 4x3 cm, along the face with the presence of homolateral inguinal lymphadenopathy. There was no associated motor or vascular disorder. The radiographs of the ankle and right lower limb were free of abnormalities. The nodule ultrasound showed a mass of soft, hypoechogenic, heterogeneous tissues. The computed tomography of the ankle revealed a lesional process of the medial face infiltrating the tendon and ligament structures and encompassing the posterior tibial artery. Histological examination showed round cell malignant tumor proliferation. The immuno-histochemical supplement showed expression of the S100 protein and Ag HMB 45 by tumor cells in favor of either a clear cell sarcoma or a melanoma. The biopsy of an inguinal node affirmed its metastatic nature. The anatomo-clinical confrontation allowed to retain the diagnosis of CCS. The extension assessment made of a chest-abdominal-pelvic computed tomography showed

metastatic lymph nodes intra-and retroperitoneal. The therapeutic decision taken in multidisciplinary consultation was to put the patient on chemotherapy (ifosfamide-Adriamycin protocol). The evolution was marked by tumor progression. The patient had refused second-line chemotherapy.



Fig-1: Nodular lesion of the left ankle, hard and fixed



Fig-2: Multiple nodular lesions of the antero external of the homolateral lower limb

DISCUSSION

Sarcoma in soft tissue clear cells or soft tissue melanoma is a rare malignant tumor that accounts for only 1% of all soft tissue sarcomas [1]. It was described for the first time by Enzinger in 1965 and then in 1983, the term soft tissue melanoma was proposed after the finding of melanin in tumor cells [2–10]. Slightly female predominant [2–5], It occurs most often in adolescents and young adults rarely in children under 10 years old or the adult over 50 years old [7–10]. It is primarily located at the extremities adjacent to the ankle and foot (40% of cases) [2–10]. A history of trauma may be present [5, 8] as in our case.

The typical clinical presentation is the appearance of a firm subcutaneous mass, well limited, partially encapsulated, of whitish grey, lobed or multinodular color and usually deep attached to the tendino-aponevrotic structures [2, 4, 5, 8]. Tumor size ranges from 1 to 15 cm with a median size of 2 to 6 cm [2–5]. Cystic and necrotic lesions are occasionally observed [2–7]. Pain is reported in 50% of cases [4–8]. At first slow and then fast growing, metastases are usual and the prognosis remains unfavorable [2–11]. Radiologically, standard radiographs don't show bone abnormalities, but lytic lesions and bone destruction have rarely been described [5]. This is the case with our observation. MRI is of great contribution in early diagnosis by showing a tumor mass in low signal on T1, rising after injection of gadolinium and variable signal in T2 [2, 6]. Histologically, the tumor is made up of nests, bundles or lobules of cells delimited by fibrous septa which are continued by collagen-oriented structures corresponding to tendinous or aponevrotic structures. The cells are monomorphic, polygonal or fusiform. They have a low eosinophilic or clarified, finely granular cytoplasm. The nucleus is round or ovoid, densified at the periphery and centered by a prominent basophilic nucleolus. Mitotic activity is low. Multinucleated giant cells can be seen [2-5, 7-9, 11]. Detection of intracellular melanin by Fontana staining is observed in approximately 50% of cases [2-4, 7, 9, 11]. Periodic acid schiff stain shows the presence of intra-cytoplasmic glycogen as in our case [4, 7, 8]. The immuno-histochemical study shows in the majority of cases an intense and diffuse cytoplasmic marking of tumor cells by the anti-HMB45 antibody and a less intense and more focal nuclear and cytoplasmic marking by the anti-PS100 antibody. Sarcomatous cells are also positive for vimentin, Melan A, CD99 and specific enolase neuron. Cytokeratin, membrane epithelial antigen, desmin and actin are usually negative [2-5, 7-9]. Cells also have abundant cytoplasm containing numerous mitochondria and glycogen aggregations [2, 4, 8].

The positive diagnosis is based on histology coupled with immunohistochemistry. Cytogenetic data is a great contribution to make the diagnosis for

difficult cases. Soft Tissue Clear Cell Sarcoma pose a differential diagnosis problem with malignant melanoma due to the similarity of the clinical picture (high rate of lymph node metastases) and histological and immunohistochemical presentation. However, it is distinguished by the absence of pigmented nodules in the skin and the presence in cytogenetic study of a chromosomal translocation (t[12; 22] [q13; q12]) [3,4]. Differential diagnosis can also occur with other types of sarcoma: synovial sarcoma, fibrosarcoma and malignant schwannoma [3, 4, 7, 8]. Its management is complex and multidisciplinary. The treatment is surgical, consisting of a broad excision with possibly lymph node curage, supplemented by radiotherapy and chemotherapy [3, 6-9, 12]. Poor prognosis factors are tumor size greater than 5 cm, necrosis, metastasis and local recurrence [3, 7, 8]. Prognosis is unfavorable (54% survival) [3, 4].

CONCLUSION

Clear cell soft tissue sarcoma is a rare melanocytic tumor with a poor prognosis that poses a diagnostic and therapeutic challenge.

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