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Clear Cell Sarcoma of Soft Tissue: A Case Report

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Abstract **Case Report**

Introduction: Clear cell sarcoma (CCS), is an extremely rare type of sarcoma, frequently diagnosed in young adults, typically arises in the extremities of the limbs and has a predilection for occurring in the tendons and aponeuroses. In the past CCS was called malignant melanoma of the soft parts because the tumor shares clinical, histological and immunohistochemical similarities with amelanotic melanoma, but it is a unique lesion, distinct from melanoma. ***Case Report:*** A 33-year old woman with no significant history was referred with an unpainful bulky ulcerated tumor of the right ankle, appeared 05 years prior to presentation following a minimal trauma. The CT of the ankle revealed a bulky mass of the medial surface locally infiltrative associated to nodules of the surrounding adipose tissues and a unilamellar periosteal reaction without bone lysis or osteocondensation. MRI of the ankle showing a lobulated mass arising in the deep structures in iso-intense signal relative to adjacent muscle on T1 and heterogeneous high-signal intensity on T2 infiltrating surrounding adipose tissues without infiltration of muscles. Histological examination showed dermo-hypodermic malignant tumor proliferation. Round cells were arranged in nests separated by dense collagenous septa, with an abundant eosinophilic to clear cytoplasm. The immunohistochemical supplement were positive for Melan-A and HMB-45. Cytokeratin, Vimentin, Desmin and CD99 were negative. The extension assessment made of a head-chest-abdominal-pelvic CT scan showed multiple homolateral inguinal lymphadenopathies necrotic. No other metastasis was detected. The anatomo-clinical confrontation allowed to retain the diagnosis of CCS. ***Discussion:*** The tumor occurs most often in adolescents and young adults and was slightly more common in female than in male patients, CCS typically arises in the extremities of the limbs, its occurrence in the upper limbs is less common, described as a slowly growing mass painless. A history of trauma may be present. Macroscopically, the tumor usually appears as a firm subcutaneous mass, well limited, partially encapsulated of whitish grey, lobulated or multinodular, cystic and necrotic remaniements are occasionnally observed. Magnetic resonance imaging studies of the lesion reveals iso-intense signal relative to adjacent muscle on T1 and heterogeneous high-signal intensity on T2, and gadolinium uptake. Histologically, CCS is characterised by nests of uniform, rounded polygonal fusiform or ovoid cells with abundant eosinophilic to clear cytoplasm, separated by dense collagenous septa. The clear cell appearance is due to the presence of large amounts of intracellular glycogen. Immunohistochemistry is usually positive for S100 protein and melanocyte-specific markers such as HMB-45 and Melan-A. Keratin, epithelialmembrane antigen (EMA), smooth muscle actin and desmin are negative in CCS. CCS can be distinguished from melanoma by molecular biology. A recurrent chromosomal translocation t(12;22)(q13;q12) or a resultant fusion of the EWSR1 gene on 22q12 and the ATF1 gene on 12q13 is specifically detected in most cases with CCS . The surgical treatment of choice is stated to be wide local excision. Radiotherapy is indicated to improve local control. The benefit of chemotherapy is unclear, although DAV chemotherapy (DTIC, ACNU and VCR) appears to reduce recurrence rate in localized CSS and may be effective in patients with metastases. The prognosis is poor, with 5-year survival rates between 40 and 60%. ***Conclusion:*** Clear cell sarcoma of soft tissues is a rare melanocytic tumor with poor prognosis, that poses diagnostic and therapeutic challenge.

Keywords: Soft tissue sarcoma - Sarcoma - Clear cell sarcoma - Malignant melanoma of the soft tissue - Case report of clear cell sarcoma.

INTRODUCTION

Clear cell sarcoma (CCS), first described by Enzinger [1] in 1965, is an extremely rare and aggressive type of sarcoma, frequently diagnosed in young adults between 20 and 40 years, typically arises in the extremities of the limbs.

In the past CCS was called malignant melanoma of the soft parts because the tumor shares clinical, histological and immunohistochemical similarities with amelanotic melanoma, but it is a unique lesion, distinct from melanoma, which has a predilection for occurring in the tendons and aponeuroses.

CASE REPORT

An apparently healthy 33-year old woman with no significant history, was referred to us because of an unpainful bulky ulcerated tumor of the right ankle, appeared 05 years prior to presentation following a minimal trauma and had showed progressive growth during the last year, evolving in a context of altered general condition (Fig 1).

Physical examination revealed right medial malleolus tumor bulky, ulcerated and lobulated, with an offensive odor, fibrin and pigmented areas were observed on the skin surface of the lesion. The tumor was surrounded by multiple nodular lesions (erythematous, firm, fixed and painless), the largest measured 2*2.5 cm, with the presence of homolateral enlarged inguinal lymph node fixed to underlying structures. There was no associated nervous or vascular disorder.

The X-rays of the ankle and right lower limb showed a soft parts mass with irregular contours measuring approximately 102 * 87 mm and discreet irregular appearance of the cortex of the tibia without bone lysis or osteocondensation.

The computed tomography of the ankle revealed a bulky mass of the medial surface locally infiltrative associated to nodules of the surrounding adipose tissues and a unilamellar periosteal reaction without bone lysis or osteocondensation.

The patient went also under MRI of the ankle showing a lobulated mass arising in the deep structures measuring 82.2*37.1*112mm in iso-intense signal relative to adjacent muscle on T1 and heterogeneous high-signal intensity on T2 infiltrating surrounding adipose tissues without infiltration of muscles of the posterior compartment of the leg associated to 7 nodules with the same signal intensity as the tumor.

Histological examination showed dermo-hypodermic malignant tumor proliferation. Round cells were arranged in nests separated by dense collagenous septa, with an abundant eosinophilic to clear cytoplasm.

The immunohistochemical supplement were positive for Melan-A and HMB-45. Cytokeratin, Vimentin, Desmin and CD99 were negative.

The extension assessment made of a head-chest-abdominal-pelvic CT scan showed multiple homolateral inguinal lymphadenopathies necrotic. No other metastasis was detected.

The anatomo-clinical confrontation allowed to retain the diagnosis of CCS. The therapeutic decision taken in multidisciplinary consultation was an amputation surgery. The patient refused radical surgery and was lost to follow up.

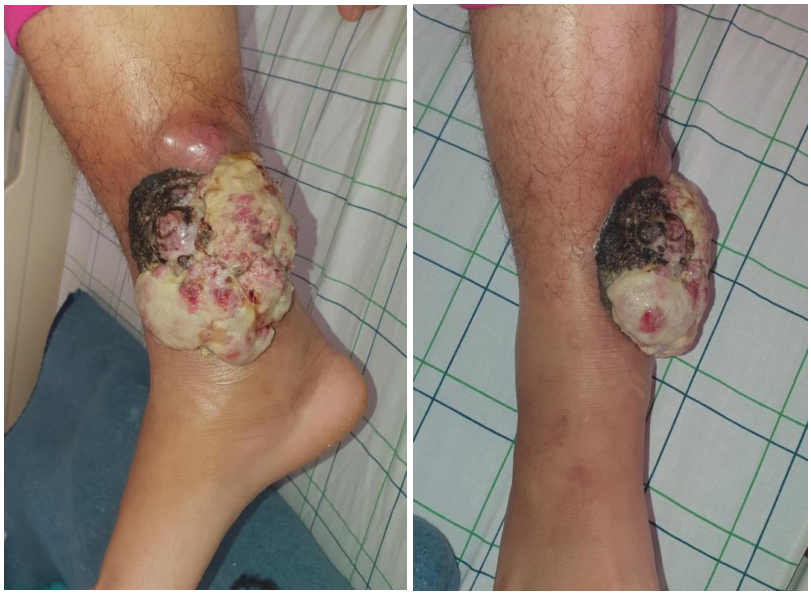

Fig 1: Clear cell sarcoma of the right ankle

Fig 2: Inguinal lymphadenopathy

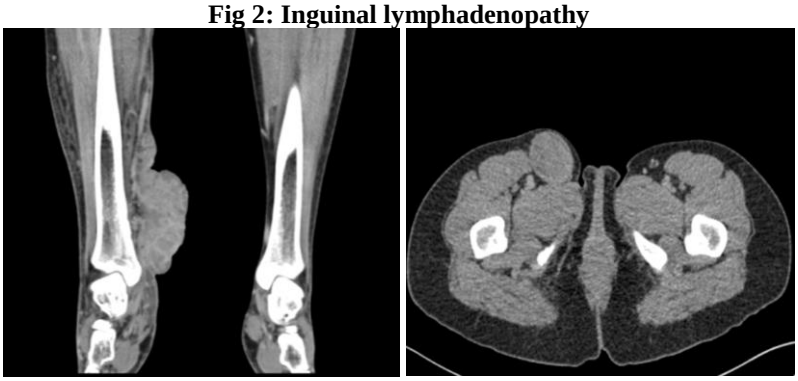

Fig 3: Tumor and Inguinal lymphadenopathy in CT

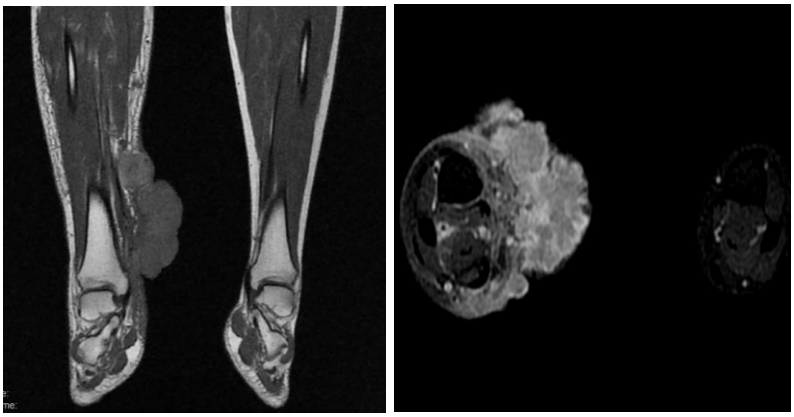

Fig 4: Tumor iso-intense signal relative to adjacent muscle on T1 rising after injection of gadolinium

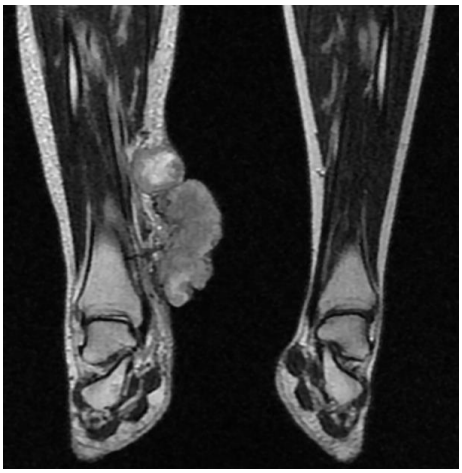

Fig 5: Tumor in heterogeneous high-signal intensity on T2

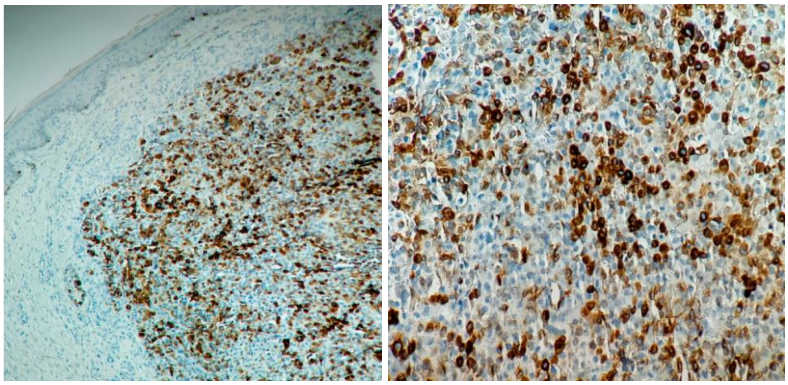

Fig 6: Immunohistochemical staining for Melan A antibody showing positive cytoplasmic staining in the tumor cells (A: magnification X20, B: magnification X40)

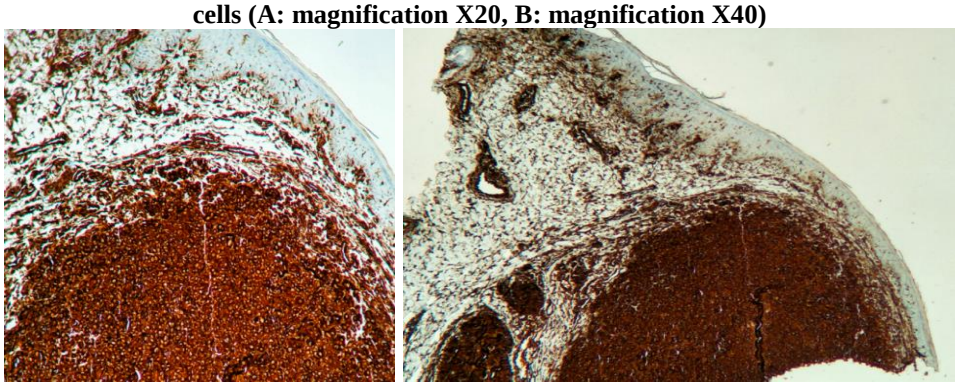

Fig 7: Immunohistochemical staining for Vimentin antibody showing positive cytoplasmic staining in the tumor cells (A: magnification X10, B: magnification X20)

DISCUSSION

Clear cell sarcoma was first described in 1965 by Enzinger [1] from a serie of 21 patients, most often young, who had tumors of the extremities, localised deep in the fascial structures. Then, in 1983, a largest serie was published by Chung and Enzinger [2], including 141 cases from the files of the Institute of Anatomical Pathology of the United States Armed Forces. These authors proposed the name of soft melanoma, given the histological similarities with melanoma. The tumor was slightly more common in female than in male patients, with an F / M sex ratio of 1.13 in Chung and Enzinger's serie.

It occurs most often in adolescents and young adults rarely in children under 10 years old or adults over 50 years old [3].

CCS typically arises in the extremities of the limbs (40% ankle or foot), its occurrence in the upper limbs is less common, described as a slowly growing mass painless that has appeared on average 3 years before diagnosis (with extremes ranging from 1 month to 30 years). The tumor measures on average 4 cm (1 to 20 cm) at diagnosis [4]. A history of trauma may be present as in our case [5].

Macroscopically, the tumor usually appears as a firm subcutaneous mass, well limited, partially encapsulated of whitish grey, lobulated or multinodular, cystic and necrotic remaniements are occasionnally observed [6].

Magnetic resonance imaging studies of the lesion reveals iso-intense signal relative to adjacent muscle on T1 and heterogeneous high-signal intensity on T2, and gadolinium uptake as in our case [7].

Histologically, CCS is characterised by nests of uniform, rounded polygonal fusiform or ovoid cells with abundant eosinophilic to clear cytoplasm, separated by dense collagenous septa. The clear cell appearance is due to the presence of large amounts of intracellular glycogen. Immunohistochemistry is usually positive for S100 protein and melanocyte-specific markers such as HMB-45 and Melan-A. Keratin, epithelial-membrane antigen (EMA), smooth muscle actin and desmin are negative in CCS [8].

CCS can be distinguished from melanoma by molecular biology. CCS has been shown to be genetically different from malignant melanoma. A recurrent chromosomal translocation t(12;22)(q13;q12) or a resultant fusion of the EWSR1 gene on 22q12 and the ATF1 gene on 12q13 is specifically detected in most cases with CCS and angiomatoid fibrous histiocytoma (AFH). Recently, another variant fusion gene EWSR1-CREB1, probably resulting from a yet unidentified chromosomal translocation t(2;22)(q34;q12), has been found in a subset of CCS. These characteristic cytogenetic and molecular genetic

changes are considered to play a pivotal role in the development of CCS and AFH [9].

The surgical treatment of choice is stated to be wide local excision. Radical surgery is performed when the lesion cannot be resected safely, or when resection would lead to vascular or neural impairment of the limb [10]. Radiotherapy is indicated to improve local control. The benefit of chemotherapy in CCS is unclear, although DAV chemotherapy (DTIC, ACNU and VCR) appears to reduce recurrence rate in localized CSS and may be effective in patients with metastases [11].

Most important prognostic factors have been found to be presence of tumor necrosis, tumor size equal to or more than 5 cm, regional lymph node metastasis, local recurrence, and distant metastases [5]. The prognosis is poor, with 5-year survival rates between 40 and 60% [8].

CONCLUSION

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

"We have no Conflict of Interest"

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