Case Report

A rare case of primary clear cell sarcoma of the leg in an elderly

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Abstract

Clear cell sarcoma (CCS) is an aggressive, rare soft-tissue tumor representing approximately 1 per cent of all soft tissue sarcomas. Also called malignant melanoma of soft parts because it shares a melanocytic phenotype that include presence of melanin, ultra structural evidence of melanosomes, and immunohisto chemical staining for S-100 protein and melanoma associated antigen HMB-45. Diagnostic difficulties arise with metastatic malignant melanoma to soft tissue. DNA ploidy analysis reveals that CCS has a diploid or lesser degree of an euploidy when compared with malignant melanoma. This paper reports on a clear cell sarcoma occurring at a very old age in upper part of leg. The tumor consisted of round to spindle cells with clear cytoplasm and multinucleated giant cells. Both S100 and HMB45 were positive. DNA analysis by Flow Cytometry showed a diploid pattern of growth. Patient underwent wide excision of the tumor. The case supports the contention that distinctive histology, IHC and DNA ploidy analysis were helpful to delineate CCS. Wide excision with negative margins remains the mainstay of treatment.

Keywords: Clear cell sarcoma, case report, DNA ploidy, Histology, Immunohistochemistry

Abbreviations: Clear cell sarcoma: CCS; Immunohistochemistry: IHC; Translocation: t

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INTRODUCTION

Franz M. Enzinger first described clear cell sarcoma in 1965. It is an aggressive, rare soft-tissue tumor representing approximately 1 per cent of all soft tissue sarcomas. Chung and Enzinger in 1983 proposed the name malignant melanoma of soft parts because it shares a melanocytic phenotype that includes presence of melanin, ultra structural evidence of melanosomes, and immunohistochemical staining for S-100 protein and melanoma associated antigen HMB-45. WHO classified it under "Tumor of uncertain differentiation." However CCS is genetically distinct harboring characteristic t 12; 22 translocation involving EWSR1 gene not found in melanoma. CCS is mainly found in children and young adults, but in rare instances it can affect anyone above the age of 40. Few studies have found a male predominance.[4,6] The most common sites of this sarcoma are the extremities, favouring the foot and ankle region, followed by knee, thigh and hand[5]. This paper reports on a clear cell sarcoma occurring at a very old age in upper part of leg.

MATERIAL AND METHODS

The present study on a case report of Clear cell sarcoma (CCS) was undertaken in Cachar Cancer Hospital and Research Centre, Meherpur, Silchar, India.

CASE REPORT

A 82 year old man presented with painless, gradually increasing swelling of 2 years duration over the medial

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aspect of upper part of right leg. It was a firm mass 14x9 cm in size, about 6 cm from knee joint occupying anterior compartment of leg. Overlying skin was erythematous and shining. Patient didn't have enlarged popliteal or inguinal nodes. Metastatic workup was negative. Patient was Hbs Ag positive. Other lab reports were within normal limits. Conventional x-ray and MRI demonstrate the soft tissue tumor without any changes in the underlying bone. FNAC from the soft tissue mass was inconclusive. Patient underwent wide excision of the tumor with periosteum and reconstruction with gastrocnemius muscle flap and split skin grafting. During the post operative period the skin graft sloughed off allowing the wound to heal by secondary intention. At the time of discharge, patient was able to walk with support. Adjuvant radiation therapy wasn't considered in view of delayed wound healing, advanced age and possibility of lymphaedema following radiation therapy, which would have affected quality of life. At one year follow-up patient is alive, disease free and is able to walk without support.

HISTOLGOY REPORT

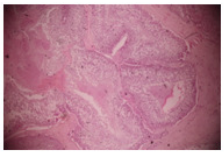

Figure 1

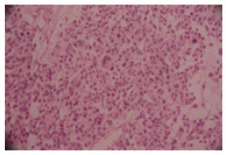

Figure 2

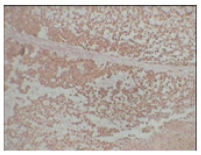

Figure 3

During surgery margins were sent for frozen section and all were free from tumor. Grossly it was a skin covered soft tissue mass measuring 11.5x9x7cm. Cut surface shows a multicystic growth measuring 6 cm in greatest dimension with hemorrhage and necrosis. Histological studies with hematoxylin and eosin shows abundant hemorrhage, necrosis and multinodular pattern of growth. Tumor cells are round to spindle with prominent neucleoli and clear cytoplasm (Fig. 1). Multinucleated giant cells and prominent mitosis noted (Fig. 2). Immunohistochemical studies demonstrated strong positive cytoplasmic staining with S-100 and focal positivity with HMB-45 (Fig. 3). With the tumor displaying these morphological and IHC features, the neoplasm was diagnosed as Clear cell sarcoma of tendon sheath/ malignant melanoma of soft tissue. Flow Cytometry was performed and it showed a diploid pattern of growth (Fig. 4).

DISCUSSION

This case highlights the known clinical feature of clear cell sarcoma originally reported by Enzinger: presenting as a slowly enlarging mass with a predilection for the soft tissue of the lower extremity[1]. Although CCS mostly afflicts young adults, in rare instances it may occur in the extreme of ages. Hocar, *et al.*[4] reviewed 52 cases of clear cell sarcoma and the age range was 6-81 years. This case represents the occurrence in extreme of age (82 years). Distinctive histology and immunoreactive markers plays an important role to delineate clear cell sarcoma. The gross appearance of clear cell sarcomas is usually that of a lobular and well-bordered or encapsulated lesion.[7] The cut surface may be marred by focal hemorrhage, necrosis, or cystic change. The microscopic features include solid nests and short fascicles of fusiform to polygonal cells enveloped by delicate to coarse fibrous septa, cells with clear to granular cytoplasm, vescicular nuclei, and one or two prominent nucleoli. A highly characteristic feature is the multinucleated tumor giant cells with 10–15 peripherally placed nuclei and intracellular melanin.[2, 5] although melanin is present in over 50% of clear cell sarcoma but is usually not abundant enough to be seen on hematoxylin-eosin stain[5]. Histologic variation includes presence of marked pleomorphism, abundant tumor necrosis and high mitotic rate.[3, 4] Imunohistochemically, the tumors are consistently positive for S-100, variably or focally for HMB45 and other melanoma markers[3]. All these histological and immunohistochemical features were present in our case. Ultrastructurally there are melanosomes[8]. The immunohistochemical and electron microscopic findings indicate that clear-cell sarcoma is a homogenous entity among soft tissue sarcomas, of probable neural crest derivation [9]. Melanocyte progenitors normally migrate from the neural crest to the basal layer of the epidermis. In CCS, the progenitor cell may not reach its final destination during embryogenesis and remains within the deeper soft tissues. Diagnostic difficulties may arise with metastatic melanoma in the absence of a known primary cutaneous tumor. El-Naggar *et al.*[10] specifies that when compared with conventional melanoma of skin metastatic to soft tissue by DNA ploidy analysis, CCS is more likely to be diploid or to show a lesser degree of aneuploidy. Also molecular analysis reveals a consistent balanced translocation t (12; 22) (q13; q12) not found in melanoma which results in fusion of the EWS gene on 22q with the ATF1 gene on 12q. The chimeric protein EWS/ATF1thus formed is responsible for the melanocytic differentiation[11]. Complete excision of the primary tumor appears to be the optimal approach to treatment [4, 12, 13]. The role of regional lymph node dissection in CCS has not been established to date. Some authors recommend prophylactic elective lymph node dissection, whereas others suggest lymphadenectomy only in patients with clinically enlarged nodes[4]. Adjuvant radiation therapy has shown to improve local control without any impact on overall survival[16]. Role of adjuvant

chemotherapy is controversial based on heterogeneity of available data[17]. In this particular case only wide excision was done followed by split thickness skin grafting. From several studies reviewed [1, 2, 4], it appears that the overall prognosis of clear cell sarcoma is poor especially once regional lymph node metastasis and haematogenous dissemination occurs. Moreover, local recurrence or metastasis (or both) are quite common after many years of freedom from disease. Tumor size and the presence of necrosis are statistically significant predictors of prognosis [14]. Jacobs *et al.*[12] described that tumors greater than 5 cm have a poorer prognosis and a higher frequency of local regrowth. Deewan M *et al.*[15] stated that a lesion on the proximal part of an extremity progresses slower than one that is more distal. Also the prognosis is better if the tumor is superficial and DNA content is diploid. The survival rate for CCS at 5, 10 and 20 years was 67, 33, and 10% respectively[14]. In this case poor prognostic factors were size (more than 5 cm), necrosis and deeper location while good prognostic factors were old age, absence of regional lymphadenopathy and diploid pattern of growth.

CONCLUSION

We are reporting this case because of its very rare incidence. Distinctive histology, IHC and DNA ploidy analysis were helpful to delineate CCS. Wide excision with negative margins remains the mainstay of treatment.

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