

CASE REPORT

CLEAR CELL SARCOMA OF GLUTEAL REGION MALIGNANT MELANOMA OF SOFT PARTS

Haren V. Oza¹, Jignasa N Bhalodia², Kinara A. Patel³, Palak J. Modi⁴

Authors' Affiliation: ¹Professor & Head; ²Associate Professor; ³Assistant Professor; ⁴Tutor, Department of Pathology, G.M.E.R.S. Medical College, Sola, Ahmedabad

Correspondence: Dr. Haren V. Oza, Email: drharenoza@yahoo.co.in

ABSTRACT

Clear cell sarcoma (CCS) is described as variant of sarcoma characterized by prominent clear cells showing features similar to malignant melanoma of soft parts. This neoplasm was first described by Dr. Franz m. Enzinger. Primary CCS usually arises in deeper soft tissues, in association with fascia, tendons, or aponeuroses. Clear cell sarcoma (CCS) is a rare malignant tumor with a propensity for slow progressive invasion. It is a tumor derived from Melanoblast like cell. They occur most commonly in the extremities, with a predilection for young females. Clear cell sarcoma of tendons and aponeuroses (malignant melanoma of soft parts) and conventional malignant melanoma may demonstrate significant morphologic overlap at the light microscopic and ultra structural level. The tumor is very rare and can pose clinical challenges in early diagnosis. This case report demonstrates an unusual site of occurrence for clear cell sarcoma.

Key words: Clear cell sarcoma (C C S), Malignant melanoma (M M) of soft part, gluteal region.

INTRODUCTION

Described in 1968 by Enzinger, clear cell sarcoma has become a well-accepted clinicopathologic entity. Clear cell sarcoma (CCS) is a rare neoplasm with a difficult clinical and histological differential diagnosis. The entity of CCS is also known as malignant melanoma of soft parts and it represents about 1% of soft tissue tumors. Because of the presence of melanin, pre-melanosomes, S-100 protein and the tendency for regional nodal metastasis, it has been suggested that this entity may be considered as malignant melanoma rather than soft tissue sarcoma. Although it produces melanin, it differs from the conventional melanoma in several important aspects. It is a deeply situated tumor that is nearly always intimately associated with tendons and aponeuroses.

It lacks junctional changes and rarely involves epidermis. Cytogenetic analysis showing characteristic translocation t(12;22) (q13;q12) has been considered pathognomonic for CCS. This translocation has been observed in neither cutaneous Malignant Melanoma. This genetic translocation demonstrates that CCS resembles MM but has a different pathogenesis. Although the term malignant melanoma of soft parts is used as synonym for this tumor, it is important that this lesion not loosely be considered malignant melanoma but rather a unique lesion.¹

Clinically, most cases present as a slowly progressive, painless mass on the lower limbs. The tumor increases in size followed by metastatic dissemination to lymph

nodes and lungs. Malignant melanoma (MM) is the most important differential diagnosis to exclude.

CASE REPORT

A Twenty three years old woman presented with fever and discharge from wound over right buttock near perianal region since one month. Physical examination revealed right perianal lesion with discharge was present giving clinical impression of perianal abscess or fistula. There was no change in bowel, bladder habits. On systemic examination RS, CVS, AS, CNS were normal. No organomegaly or lymphadenopathy found. Routine investigations show Hb. 9.0 gm%, Total count 7800/cumm, platelet count 3,90,000/cumm, Renal function test and Liver function test were normal. She was treated with wide local excision & tissue was submitted for histopathological examination. The gross specimen measured 6 cm. in aggregate with skin covered fibro fatty soft tissue. On cut section it shows lobulated mass with dark brown patchy areas seen. Microscopic examination on H & E stain revealed there are round to polyhedral cells arranged in groups, nests, and trabecular formation. The tumor cells are separated by fibrous septa. The cells are having round nucleus with nucleoli and minimal cytological atypia with clear cytoplasm. (FIG 1).

There are brownish pigment laden tumor cells and macrophages present in the deeper tissue. (FIG2).

No junctional activity seen. At places tumor cells also

showed eosinophilic cytoplasm with PAS positivity. On immunohistochemistry tumor cells showed positive for HMB45 (FIG3).

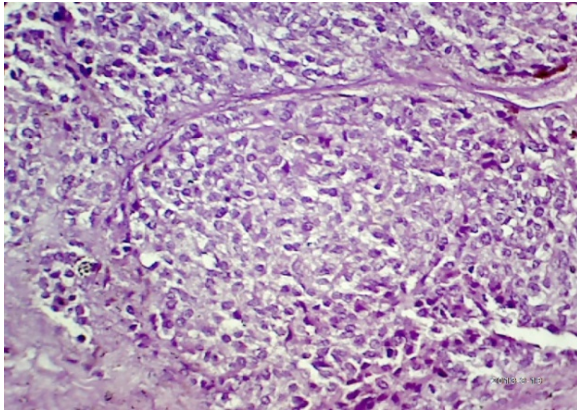


FIG.1: H & E Stain: Tumor cells arranged in nests separated by fibrous septa.(10X)

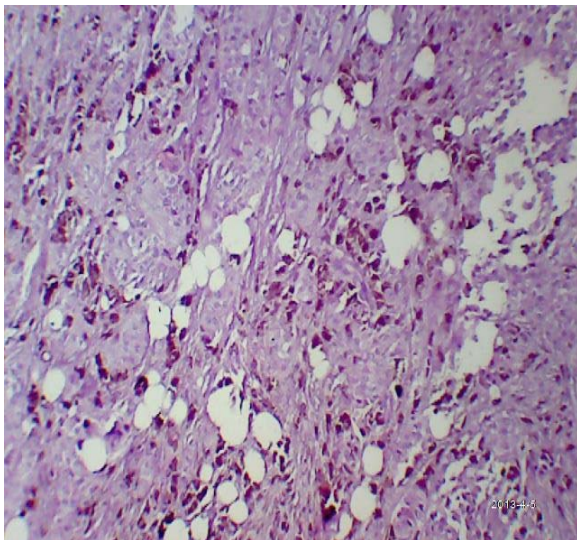


FIG 2: Brownish pigment laden tumor cells deep in to soft tissue. (40 X)

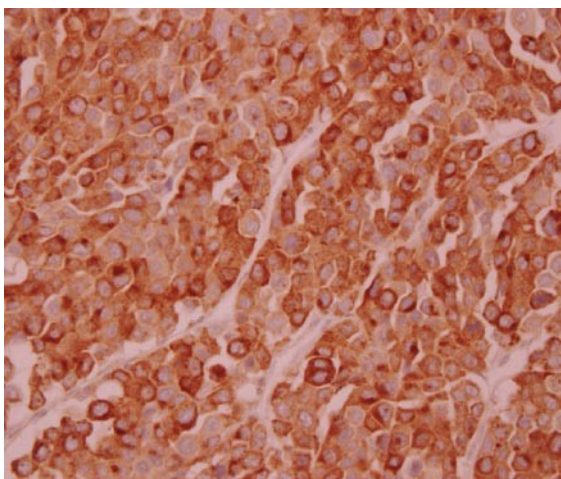


FIG 3: IHC marker HMB45 positive tumor cells with cytoplasmic immunoreactivity (40X)

Focal positive for S100 protein and Negative for CD99. Diagnosis of Clear Cell Sarcoma - Malignant melanoma of soft parts was given.

DISCUSSION

CCS accounts for less than 1% of all soft tissue sarcomas and predominantly affects young adults.^{2,3} There is a slight female predominance.^{3,4} CCS is a recently described variant of sarcoma characterized by prominent clear cell features similar to clear cell melanoma. Primary CCS usually arises in deeper soft tissues, in association with fascia, tendon, or aponeurosis. CCS is a separate entity includes the presence of spindle and clear cells, absence of nuclear atypia, and small and inconspicuous nucleoli. CCS also known as malignant melanoma of soft part, is a tumor with propensity for lymphatic spread.⁵ The gross appearance of clear cell sarcomas is usually that of a lobular and well-bordered or encapsulated lesion. Microscopically, the tumor in this case consisted of well-defined nests separated by fibrous septa.^{3,5} The tumor cells were polygonal or spindle-shaped and contained clear or eosinophilic cytoplasm. Some tumor cells may have contained melanin pigment.^{3,6} Recent cytogenetic studies have shown that clear cell sarcoma has a t(12;22) (q13;q12) translocation, a feature not encountered in malignant melanoma.³ Less common morphologic variations include spindle-cell arrangement, marked pleomorphism, solid-cell aspect, microcystic aspect, and presence of myxoid stroma.⁷ Clear cell sarcoma (CCS) and malignant melanoma share overlapping immunohistochemistry with regard to the melanocytic markers HMB45, S100^{3,5,6} and Melan-A. However, the translocation t(12; 22)(q13; q12) is specific to CCS.^{3,4,5,6,8,9} It has been suggested that CCS may be conclusively diagnosed using cytology, immunohistochemistry (HMB- 45+ and S-100+ stains), cytogenetic analysis (demonstrating the specific translocation), and electron microscopy. Therefore, although these neoplasms are closely related, they are now considered to be distinct entity.⁷ Characteristic translocation t (12; 22) (q13; q12), has been considered pathognomonic for CCS.^{4,5} This translocation has been identified in 70-90% of CCS cases using cytogenetic studies and reverse-transcriptase polymerase chain reaction. Nevertheless, this cytogenetic rearrangement is characteristic but not entirely unique for CCS, because similar fusion genes can also be found in angiomatoid fibrous histiocytoma.⁴ In general, CCS has a high propensity of lymph node metastasis and the prognosis is dismal once metastasis occurs. It does appear from multiple literature reviews, that tumors less than 2 cm have a generally better long term prognosis.¹⁰ However, CCS is currently a distinct entity classified by World Health Organization as soft tissue and bone tumors.⁹ In contrast to MM, and CCS is deeply situated and

generally located in non-pigmented areas. For CCS, prognostic factors are the tumor size and necrosis. Patients are generally subjected to local control through surgery followed by adjuvant radiotherapy. Early detection and adequate local control of the lesion are the most important aspects in the management of CCS. Histological diagnosis requires a high level of suspicion since this tumor may mimic other soft tissue sarcomas.

CONCLUSION

Here interesting clinical case of CCS in young adult female, arising in the gluteal region has been reported. In addition, most important differential diagnosis has been reviewed. In our patient, definitive diagnosis was only possible based on the strength of the immunohistochemistry. Differentiation of CCS from other clear or spindle cell neoplasms, and unusual MM subtypes is an essential component in patient management. Pathologists and clinicians need to be aware of the aforementioned entities, so that an early diagnosis and treatment may improve the prognosis.

REFERENCES

1. Sharon W. Weiss, John R. Goldblum. *Enzinger and Weiss's soft tissue tumors*, 4th ed. St. Louis, Missouri: Mosby; 2001. page 1241.
2. Andrew J. Creager, Martha B. Pitman and Kim R. Geisinger. Cytologic Features of Clear Cell Sarcoma (Malignant Melanoma) of Soft Parts. A Study of Fine -Needle Aspirates and Exfoliative Specimens. *Am J Clin Pathol.* 2002;117:217-224.
3. Xudong Liu, Huizhen Zhang and Yang Dong. Primary clear cell sarcoma of humerus: case report. *World Journal of Surgical Oncology.* 2011; 9:163.
4. M Rodriguez-Martin, M Saez-Rodriguez, B Esquivel, R Sanchez Gonzalez, A Noda Cabrera, A Martin Herrera. Clear Cell Sarcoma :A Case Mimicking Primary Cutaneous Malignant Melanoma. *Indian J Dermatol.* 2009;54(2):168-172.
5. Xu G G, Chong YL, Cheong M O. Clear cell sarcoma of the rectus sheath. *Singapore Med J.* 2007;48(7):e203-205.
6. O. Hocar, A. Le Cesne, S. Berissi, P. Tarrrier, S. Bonvalot, D. Vanel et al. Clinical Study Clear Cell Sarcoma (Malignant Melanoma) of Soft Parts: A Clinicopathologic Study of 52 Cases. *Dermatology Research and Practice.* 2012; DOI:10.1155/2012/984096.
7. Shoko Nakayama, Taiji Yokote, Kazuki Iwaki, Toshikazu Akioka, Yuki Masuda, Motomu Tsuji et al. A rare case of primary clear cell sarcoma of the pubic bone resembling small round cell tumor: an unusual morphologic variant. *BMC Cancer.* 2012; 12:538.
8. Langezaal SM, Graadt van Roggen JF, Cleton-Jasen AM, Baelde JJ, Hosendoorn PC. Malignant melanoma is genetically distinct from clear cell sarcoma of tendons and aponeurosis (malignant melanoma of soft parts). *Br J Cancer.* 2001; 84:535-538.
9. ZA Ibrahim, KL Pan, PS Shanmugam. Clear Cell Sarcoma of the Knee. A Report of Two Rare Cases. *Malaysian Orthopaedic Journal.* 2011; 5(1):63-66.
10. A I Kline. Clear Cell Sarcoma of the Foot: A Case Report of Malignant Melanoma of Soft Parts. *Podiatry Internet Journal.* 2007; 2(6):2.