Available online at https://www.mmpubl.com/

International Journal of Histopathological Interpretation

Case Report

CLEAR CELL LEIOMYOSARCOMA OF THE ANTERIOR ABDOMEN -A RARE CASE PRESENTATION

Syeda Iqra Usman[1] , *Mazhar Fahim*[2] , Md. Sakib Anwer[3], Kafil Akhtar[4*]

1,2,3 Junior Resident , 4 Professor
1,2,3,4 Department of Pathology, Jawaharlal Nehru Medical College (JNMC), Aligarh Muslim University, Aligarh-UP-202002*

How to cite: Kafil Akhtar et al., Clear Cell Leiomyosarcoma of the Anterior Abdomen – A Rare Case Presentation. Int J Histopathol Interpret 2023; 12(1), 1-5 DOI: https://doi.org/10.56501/intjhistopatholinterpret.v12i1.741

Received :24/12/2022 *Accepted: 17/01/2023* *Web Published: 29/01/2023*

Abstract

Leiomyosarcoma is a rare type of malignant smooth muscle tumor. It accounts for around 10-20% of soft tissue sarcomas. We report a case of Leiomyosarcoma with clear cell change in a 62 years old male, who had complaints of anterior abdominal swelling for the last one year. On local abdominal examination, the swelling was diffuse, 10x3cm in size, firm and non-tender and appeared to arise from the anterior abdominal wall. On ultrasonography a heterogenous mass of 10x4 cm was found. Computed tomography scan showed a mass of size 10x4x2 cm in the anterior abdominal wall infiltrating into the fat spaces. He was diagnosed as a case of leiomyosarcoma with clear cell change on the basis of histopathological examination of the excisional biopsy.

Keywords: Sarcomas, Clear Cell, Leiomyosarcoma, Immunohistochemistry

Address for Correspondence:
Dr. Kafil Akhtar
Professor, Department of Pathology, Jawaharlal Nehru Medical College (JNMC),Aligarh Muslim University, Aligarh-UP-202002
Email: drkafilakhtar@gmail.com

INTRODUCTION

Leiomyosarcoma is malignant soft tissue sarcoma originating from smooth muscle cells. Incidence is common in adulthood usually present between the fifth to sixth decades of life. Women has higher incidence of disease as compared to men [1]. Retroperitoneum is commonest site of involvement making 12-69.0% of the cases [2]. Uterus and skin are the only sites where clear cell variant of leiomyosarcoma has been reported [3].

Clear cell tumors of soft tissue include heterogenous neoplasms of both benign and malignant potential with different morphological patterns, clinical behavior and prognosis. Malignant neoplasms include clear cell sarcoma, liposarcoma, malignant perivascular epithelioid cell tumor, leiomyosarcoma and rhabdomyosarcoma [4,5].

CASE REPORT

A 62-year-old male presented to the Surgery Out-Patient Department with complaints of swelling in the abdomen for the past one year. The swelling was gradually increasing in size, with rapid increase in the last three months. He complained of mild pain in the swelling with no other associated symptoms. There was no family history of similar mass. There was no significant treatment history.

On local examination, the swelling was diffuse and appeared to arise from the anterior abdominal wall, approximately 10x3cm in size, non-tender and firm in consistency. No lymph nodes were palpable. All the hematological investigations of the patient were within normal limits.

Ultrasonography was performed which showed a heterogenous abdominal mass of 10x4cm in size, but the nature of the lesion could not be ascertained. Computed tomography scan showed an abdominal mass of 10x4x2cm, in the anterior abdominal wall with minimal ascites with increased vascularity and infiltration into the upper abdomen fat, suggesting a malignant sarcoma.

Fine needle aspiration cytology showed a moderate cellularity smear with scattered atypical plump to spindle shaped cells with mild anisonucleosis, hyperchromatic nucleus with blunt ends and indistinct cytoplasm, suggestive of Spindle Cell Neoplasm favoring a malignancy with the differentials of leiomyosarcoma, fibrosarcoma, rhabdomyosarcoma, liposarcoma and malignant peripheral nerve sheath tumor.

Excisional biopsy was performed which on gross examination showed a well circumscribed mass, 10x3x2 cm in size, greyish in colour and firm in consistency with focal areas of necrosis and hemorrhage. Microscopic examination on Hematoxylin and Eosin-stained sections showed highly atypical spindle shaped cells arranged in fascicular pattern intersecting each other at wide angles. Some tumor cell showed cigar shaped nuclei with increased nuclear to cytoplasmic ratio with moderate anisonucleosis with 10/10hpf mitotic figures with foci of necrosis and hemorrhage. Several tumor cells showed presence of perinuclear cytoplasmic clearing (Figure 1 and 2). On immunohistochemistry S-100 and desmin showed focal cytoplasmic positivity (Figure 3) and HMB-45 was negative. Based on the histomorphology, a final diagnosis of Leiomyosarcoma with clear cell change was given. Adjuvant chemotherapy with 4 cycles of Docetaxel (50mg/m2) and Gemcitabine (100mg/m2) was administered to our patient. On follow up after 6 months, there was no recurrence or any evidence of metastatic disease in our patient

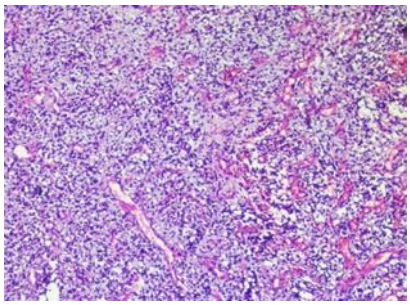

Figure 1 Microscopic examination showed highly atypical spindle shaped cells arranged in fascicular pattern intersecting each other at wide angles. Some tumor cell showed cigar shaped nuclei with increased nuclear to cytoplasmic ratio with moderate anisonucleosis. Several tumor cells showed presence of perinuclear cytoplasmic clearing. Hematoxylin and Eosin x 10 X.

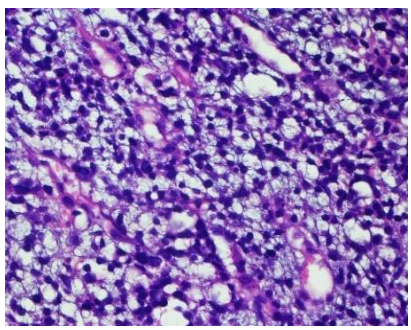

Figure 2 Shows the microscopic picture of perinuclear cytoplasmic clearing – High power (40x)

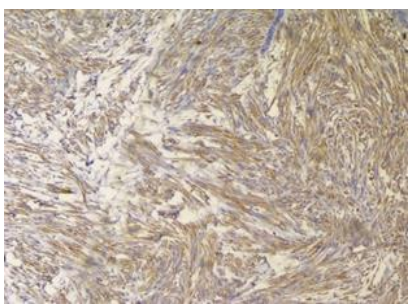

Figure 3 On immunohistochemistry S-100 showed focal cytoplasmic positivity in the tumor cells; IHC, S-100, 40X

DISCUSSION

diagnosed cases of soft tissue sarcomas are leiomyosarcomas [1]. It occurs commonly in abdomen, retroperitoneum, large blood vessels and the uterus [2]. Incidence of occurrence in extremities is less as compared to other soft tissue sarcomas [3]. Our patient presented with an anterior abdominal mass for the last one year.

There is no solid evidence that leiomyoma transforms into leiomyosarcoma and it is currently believed that they arise de novo [6,7]. Predisposing factors for leiomyosarcoma includes previous irradiation and immunosuppression mainly because of human immunodeficiency virus. It has also been reported to be associated with Epstein-Barr virus infection and number of syndromes like Gardner's syndrome, Recklinghausen's neurofibromatosis and retinoblastoma [8].

Leiomyosarcomas typically have fascicles of spindle cells which are sharply marginated and intersect each other. Tumor cells have elongated hyperchromatic nuclei and abundant eosinophilic cytoplasm. Usually, these tumors show positivity for alpha smooth muscle actin, desmin and h-caldesmon but none of these markers are specific for smooth muscle differentiation [9,10]. HMB-45 positivity is seen in clear cells of uterine

epithelioid leiomyosarcoma but it's expression has not been investigated in conventional leiomyosarcomas [11]. Fine needle aspiration in our case was suggestive of spindle cell neoplasm favouring malignancy and biopsy findings of the excised mass showed typical features of leiomyosarcoma with fascicles of spindle cells intersecting each other. Some tumor cells showed cigar shaped nuclei with increased nucleus to cytoplasmic ratio and moderate anisonucleosis, with presence of perinuclear cytoplasmic clearing in several tumor cells. Periodic Acid Schiff stain was negative. Immunohistochemical studies showed positivity for S-100, desmin and negativity for HMB-45 in our case.

Common histological types of malignant abdominal soft tissue sarcomas include leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma, liposarcoma and malignant peripheral nerve sheath tumor [4,5]. Fibrosarcoma shows a characteristic herring bone pattern, with moderate degree of pleomorphism, is vimentin positive and occasionally positive for smooth muscle actin [4]. Liposarcoma shows multivacuolated lipoblast with scalloped nucleus [6]. Malignant peripheral nerve sheath tumor has fusiform nuclei with streaming and intersecting fascicle arrangement. It is mostly hypercellular and often associated with precursors lesions such as neurofibromatosis [7]. Rhabdomyosarcoma shows tumor cells with tadpole like rhabdomyoblasts that have eccentric nucleus and sparkling eosinophilic cytoplasm, which expresses desmin [5]. On the basis clinical, cytological and histomorphological findings, a diagnosis of leiomyosarcoma with clear cell change was given in our case. Clear cell changes in leiomyosarcoma, to the best of our knowledge has been described only in cutaneous and uterine lesions [3].

Our patient was administered adjuvant chemotherapy, with a one year follow up. There are no specific trials of adjuvant chemotherapy in leiomyosarcoma that originates outside the uterus [8,12]. There is institutional variation in the use of adjuvant chemotherapy [12].

Conclusion

Leiomyosarcoma is one among the rare smooth muscle tumor. With advancements in diagnosis & treatment, this can be identified earlier & might help in the improvement of the prognosis of the patient.

Financial support and sponsorship:

Nil

Conflicts of interest

There are no conflicts of interest

References

1. George S, Serrano C, Hensley ML, Ray-Coquard I. Soft Tissue and Uterine Leiomyosarcoma. J Clin Oncol 2018; 36(2):144-50.

2. Shiu MH, Weinstein L, Hajdu SI, Brennan MF. Malignant soft-tissue tumors of the anterior abdominal wall. Am J Surg 1989;158(5):446-51.

3. Atzori L, Pilloni L, Zanniello R, Ferreli C, Rongioletti F. Clear-cell variant of superficial cutaneous leiomyosarcoma associated with RB1 mutation: Clinical, dermoscopic, and histopathological characteristics. J Cutan Pathol 2020; 47(6):571-5.

4. Wilkinson R, Sonarkar R, Nazar ZK, Sonawane S, Dhole R. Fibrosarcoma of anterior abdominal wall: A rare case report. Int J Surg Case Rep 2020; 77:210-13.

5. Hartmann S, Lessner G, Mentzel T, Kübler AC, Müller-Richter UD. An adult spindle cell rhabdomyosarcoma in the head and neck region with long-term survival: a case report. J Med Case Rep 2014; 19: 208-210.

6. Sun P, Ma R, Liu G, Wang L, Chang H, Li Y. Pathological prognostic factors of retroperitoneal liposarcoma: comprehensive clinicopathological analysis of 124 cases. Ann Transl Med 2021; 9(7):57-61.

7. Lucas CG, Vasudevan HN, Chen WC, Magill ST, Braunstein SE, Jacques L et al. Histopathologic findings in malignant peripheral nerve sheath tumor predict response to radiotherapy and overall survival. Neuro Oncol Adv 2020; 2(1):131-35.

8. Jones BF, Srinivasan V, Gumparthy K, Hughes D. Leiomyosarcoma of the tonsil. J Laryngol Otol 2011;125: 869-72.

9. Marko J and Wolfman DJ. Retroperitoneal Leiomyosarcoma from the Radiologic Pathology Archives. Radiographics 2018; 38(5):1403-20.

10. Serrano C and George S. Leiomyosarcoma. Hematol Oncol Clin North Am 2013;12: 957-74.

11. Simpson KW and Albores-Saavedra J. HMB-45 reactivity in conventional uterine leiomyosarcomas. Am J Surg Pathol 2007;13: 95-8.

12. Auerbach A and Cassarino DS. Clear Cell Tumors of Soft Tissue. Surg Pathol Clin 2011;13: 783-98.

Published by MM Publishers
https://www.mmpubl.com/ijhi