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Rhabdomyosarcoma – Pleomorphic Type at Medial Aspect of Left Thigh With Unusual Presentation As Ulcerative Growth – A Case report

¹Dr Sukhdev V Adhikari, JR-3 General Surgery, MGM Medical College and Hospital, Navi Mumbai

²Dr Lisha Suraj, Professor, Department of General Surgery, MGM Medical College and Hospital, Navi Mumbai

³Dr Sameer Kadam, Head of Department, Department of General Surgery, MGM Medical College and Hospital, Navi Mumbai -410209.

Corresponding Author: Dr Sukhdev V Adhikari, JR-3 General Surgery, MGM Medical College and Hospital, Navi Mumbai

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Abstract

Rhabdomyosarcoma is highly malignant type of soft tissue tumor. It is divided into 3 subtypes that is Embryonal, alveolar and pleomorphic. Primary pleomorphic rhabdomyosarcoma is a rare type of soft tissue tumor accounting for <2% of all adult sarcomas. Here we present a unusual case of 44-year-old male presenting with ulcerative growth over medial aspect of left thigh. With keeping in mind for differential diagnosis of squamous cell carcinoma patient was worked up wherein edge biopsy of the lesion revealed findings consistent with pleomorphic rhabdomyosarcoma. This case report demonstrated that pleomorphic rhabdomyosarcoma can be considered as rare etiology for differential diagnosis of ulcerative growth at lower limb.

Keywords: Rhabdomyosarcoma, Malignant, Pleomorphic

Introduction

Rhabdomyosarcoma is highly malignant type of soft tissue tumor. It is divided into 3 subtypes that is Embryonal, alveolar and pleomorphic. The most common subtypes are embryonal and alveolar subtypes. Primary pleomorphic rhabdomyosarcoma is a rare type of soft tissue tumor accounting for <2% of all adult sarcomas. The present study describes a case of 44-year-old male with primary pleomorphic rhabdomyosarcoma at medial aspect of left thigh with unusual presentation as ulcerative growth.

Case Study

44-year-old male presented with a chief complaint of ulcerative growth at inner aspect of left thigh since 2 months increased in size since 1 month associated with bloody discharge since a week. History of previous scar present. No history of pain, fever, loss of weight and appetite. On examination vitals are stable, on local examination 10x8x5 cm in size irregular ulcerative growth present on medial aspect of left thigh, everted

edge, it was hard and non-tender on palpation, induration present, no lymph nodes felt clinically, all blood reports were normal, X ray local part no bony involvement, MRI of knee suggestive of large irregular lobulated mass in medial aspect of left knee abutting and extending into muscular plane. Edge Biopsy was done because we had suspected Marjolin ulcer and histological findings were consistent with pleomorphic rhabdomyosarcoma, on IHC, tumor cells were positive for Desmin, myo-D1 and SMA, while these were negative for EMA, S100, CD 34, myogenin and caldesmon. NCCT thorax suggestive of lobulated right lower lobe mass, appears suspicious of metastasis, 2D ECHO was normal and patient is undergoing chemotherapy at present.

Differential Diagnosis -

Squamous cell carcinoma, Marjolins ulcer

Discussion

Pleomorphic Rhabdomyosarcoma is defined as high grade sarcoma composed of undifferentiated round and spindle cells that display skeletal muscle differentiation without embryonal or alveolar component (1). Pleomorphic rhabdomyosarcoma was first described by Stout and colleagues in 1946(2). It is least common subtype and reported to commonly affect older than 45 years. (3) Lymphadenopathy is least common in pleomorphic subtype. It is aggressive lesion arising from deeper soft tissue of the extremities with high chance of metastasis. Treatment of patients with rhabdomyosarcoma is usually primarily surgery if excision is attainable followed by radiotherapy and chemotherapy. (4) Histological subtype pleomorphic type has least favorable outcome. Pulmonary metastasis has been reported as most frequent distant metastasis. Presence of metastasis at presentation along with tumor

size, tumor resectibility and patient age are prognostic factors. (5)

Conclusion

This case report demonstrates an exceptionally rare malignancy in adult patient with unusual presentation as ulcerative growth at medial aspect of left thigh which turned to be pleomorphic type of rhabdomyosarcoma. So this diagnosis may be considered as rare etiology for differential diagnosis of ulcerative growth at lower limb.

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Legend Figures



Figure 1: Ulcerative growth at medial aspect of left thigh

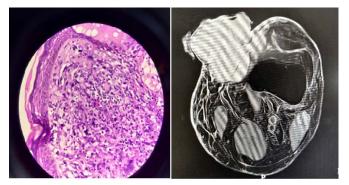


Figure 2: a) Hematoxylin and eosin staining shows spindled, pleomorphic cells arranged in fascicular pattern with prominent nucleoli and abundant eosinophilic cytoplasm b) MRI Left knee joint shows large, irregular, lobulated mass in medial aspect of left knee measuring 8.2x.9x8.5 cm abutting and extending into muscular plane.