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Undifferentiated Pleomorphic Sarcoma

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Abstract: ***Undifferentiated pleomorphic sarcoma (UPS) is a rare soft tissue sarcoma with high risk of metastasis and local recurrence. Largely affects elderly males, ultraviolet radiation has predominant etiological role, most common sites being extremities. Undifferentiated pleomorphic sarcoma closely shares similar features with atypical fibroxanthoma but deep subcutaneous invasion, lymphovascular invasion, tumor necrosis differentiates UPS from atypical fibroxanthoma. Wide excision is the gold standard treatment, chemoradiotherapy acts as adjunctives. A 63 year old male patient presented with multiple swellings over left thigh and left groin since 6months, on examination multiple well defined nodules, hard in consistency, non mobile, adherent to underlying skin, were seen over left thigh, with left sided inguinal lymphadenopathy which was non tender and hard in consistency. Histopathology and Immunochemistry features were suggestive UPS and PET scan revealed no metastasis. Patient was treated with chemoradiotherapy. Undifferentiated pleomorphic sarcoma is a diagnosis of exclusion hence this case is reported for its rarity.***

Keywords: Undifferentiated pleomorphic sarcoma (UPS), Malignant fibrous histiocytoma, Pleomorphic spindle cells, Wide excision

1. Introduction

Sarcomas are heterogenous group of malignancies arising from mesenchymal cells and accounts for less than 1% of all cutaneous malignancies[1]. Undifferentiated pleomorphic sarcoma (UPS) was previously known as Malignant fibrous histiocytoma, later in 2013 WHO classified UPS as a separate entity[2]. The origin of the tumor is from mesenchymal stem cells not from histiocytes[3]. The tumor can affect skin, soft tissues, bones, retroperitoneum and metastasize to several organs. Ultraviolet radiations acts as an important etiology with telomerase reverse transcription (TERT) mutation in 75% affected individuals[4]. Incidence of UPS was higher beyond 6th decade of life and it is always a diagnosis of exclusion.

2. Case Report

A 63 year old male patient presented with multiple asymptomatic swellings over left thigh and left groin since 6months, on examination multiple well defined skin colored nodules with largest measuring 3x2cm and smallest measuring 1x0.5cm, hard in consistency, non mobile, smooth surface, adherent to underlying skin, were seen over left thigh, with left sided inguinal lymphadenopathy which were non tender, hard in consistency without any discharge (Fig1). Differential diagnosis of lobomycosis, tuberous xanthoma, dermatofibroma, pleomorphic sarcoma was considered.5mm Punch biopsy was sent for histopathology and immunohistochemistry which showed thinned out epidermis, dermis with dense infiltration with spindle shaped cells with moderate pleomorphism having round to oval vesicular nucleoli. Mitosis of 5 - 7/10 hpf was noted, tumor was involving adjacent dermal appendages. Tumor was Vimentin positive and was suggestive of high grade pleomorphic sarcoma (Fig2). PET scan was done it revealed no metastasis. Patient was referred to oncologist and was treated with chemoradiotherapy.

3. Discussion

Undifferentiated pleomorphic sarcoma (UPS) is spindle cell neoplasm arising from skin or soft tissues with different names in the literature such as atypical fibroxanthoma, malignant fibrous histiocytoma, pleomorphic dermal sarcoma. The term UPS was given by Fletcher[5]. The tumor arises in elderly males usually after 6th decade of life[5]. The exact pathogenesis remain obscure, ultraviolet radiation plays an important role with telomerase reverse transcription (TERT) mutation in 75% affected individuals. Upregulation of Hedgehog and Notch signaling, mutation in tumor protein 53 (TP53), deletion mutation of phosphate and tensin homolog (PTEN) also play a role[6, 7]. Clinically presents as rapidly growing nodular or exophytic maculopapular lesions of more than 2 cm, with extremities being most common site (55%) followed by trunk (35%), retroperitoneum (9%) [8]. Diagnosis is made by histopathology which shows atypical pleomorphic spindle cells, extending in to deep dermis, hypodermis, facia, muscles[9]. The tumor shows LN2, CD10, Vimentin, Ki63 positivity on immunohistochemistry[10]. MRI is done for local staging, PET scan to look for distant metastasis. Wide excision with 2cm margin is the gold standard treatment, for distant metastasis, unresectable cases chemoradiation is given. Epirubicin, ifosfamide, gemcetabine, pembrolizumab are some drugs used in chemotherapy. Atypical fibroxanthoma, liposarcoma, dermatofibrosarcoma, angiosarcoma, melanoma are some differential diagnosis. The 5 and 10 year survival rates were 60% and 48% respectively.

4. Conclusion

Undifferentiated pleomorphic sarcoma continue to represent a diagnostic and clinical management challenge. Uniform adoption and application of soft tissue sarcoma classifications and minimum reporting datasets will promote homogeneity in clinicopathological analysis. Aggressive surgical management to prevent local recurrence, clinical surveillance, and multidisciplinary management following diagnosis remains the mainstay of treatment.

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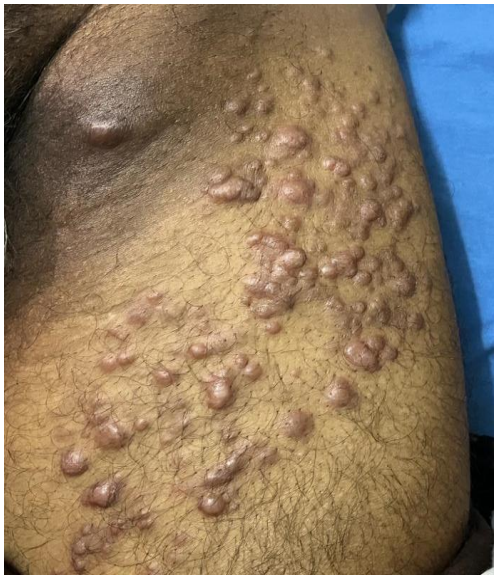

Figure 1: Multiple well defined skin colored nodules over left thigh

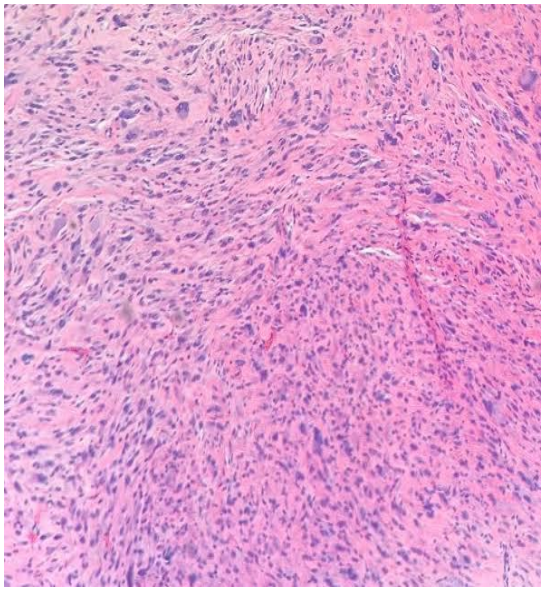

Figure 2: HPE 40X - Showing pleomorphic spindle cells in the dermis

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