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Case Report

Undifferentiated Pleomorphic Sarcoma: A Case Study of an Elusive Malignancy – A rare case report

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1. Introduction

Undifferentiated pleomorphic sarcoma (UPS), previously called malignant fibrous histiocytoma, is a rare and highly aggressive soft tissue sarcoma that originates from mesenchymal stem cells [1]. (UPS) accounts for approximately 5-10% of all soft tissue sarcomas. UPS represents less than 1% of cancers in adults, UPS is characterized by its rapid growth and often presents as a large, painless nodule either on or under the skin [2]. The tumor is marked by a variety of atypical, spindle shaped or oval cells, a high number of mitotic figures, and occasional giant cells.

UPS typically affects individuals between the ages of 50 and 70. Due to the nonspecific histology of this disordered tumors, UPS is a diagnosis of elimination. We present a case involving a 57-year-old female diagnosed with undifferentiated pleomorphic sarcoma (UPS) to highlight the distinct aspects of our case in comparison to others. We aim to explore the available diagnostic and therapeutic options for managing such cases.

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2. Case Details

A 57-year-old female presented to the surgical oncology department with a four month history of a painful, nodular swelling located over the anterior aspect of her right knee. The lesion exhibited an ulcero-proliferative surface but did not involve the underlying bone or joint. She denied any associated trauma, restriction of joint movement, or discharge from the lesion. Her past medical history was unremarkable, with no known comorbidities such as diabetes, hypertension, tuberculosis, or asthma. General physical and systemic examinations revealed no abnormalities. On local examination, the lesion appeared as a firm to hard, 8 × 6 cm mass with overlying skin discoloration and a centrally located ulcer measuring 3 × 3 cm.

Ultrasound imaging of the right inguinal region demonstrated enlarged lymph nodes, the largest measuring 2.6 × 1 cm, with preserved fatty hila. MRI of the right knee revealed multiple subcutaneous lesions suggestive of a neoplastic process, most likely of mesenchymal or nerve sheath origin. Arterial Doppler of the right lower limb showed mildly increased blood flow in the posterior tibial artery and identified two well-defined lesions—suprapatellar and anterior patellar - with both peripheral and central vascularity, indicating a probable neoplastic etiology.

A CT scan of the chest, performed with and without contrast, revealed fibrobronchiectatic changes in the right middle lobe. The patient underwent a wide local excision of the lesion, followed by a sural flap procedure and split-thickness skin grafting was done and the resected specimen was sent for histopathological examination which was a single skin covered fibrofatty tissue mass measuring 10 × 9 × 2.5 cm with attached skin measuring 10 × 9 cm Figure 1.

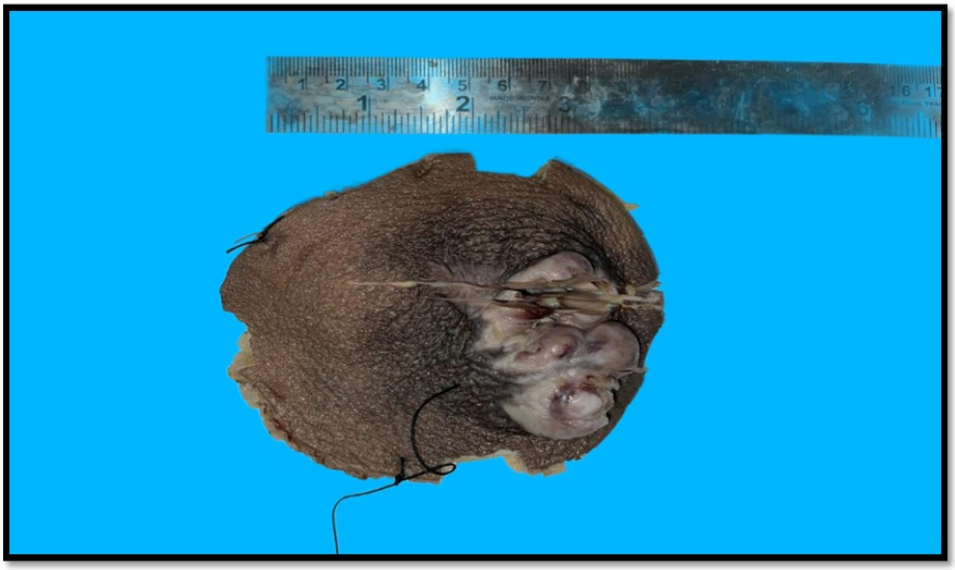

Figure 1: Gross image showing external appearance of the tumor

External surface showed ulcerative nodules on skin measuring 5×4×2.3 cm. On serial sectioning, identified a grey white lobulated lesion measuring 6.5×6×2.6 cm firm to hard in consistency with areas of necrosis Figure 2.

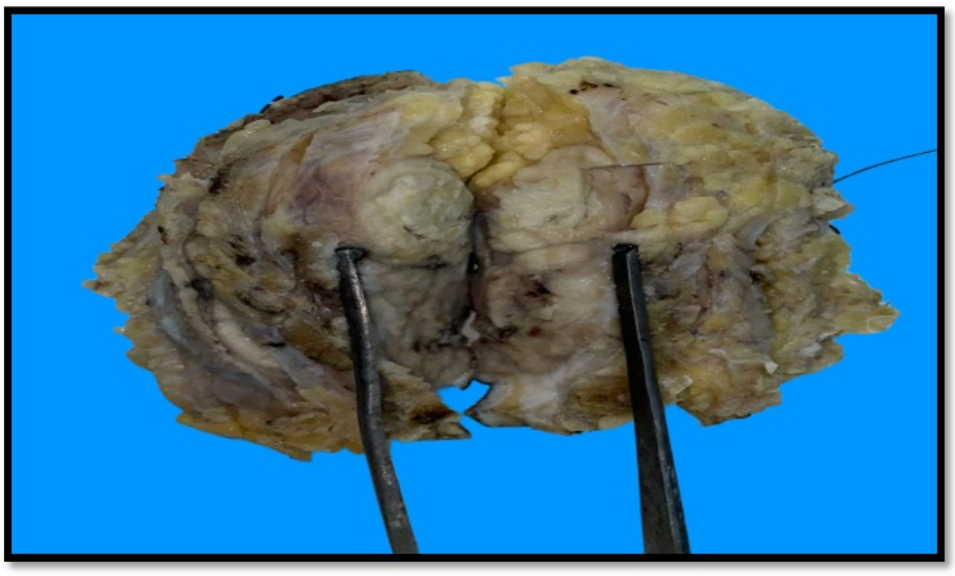

Figure 2: Gross image showing cut section of tumor

On microscopy examined tissue sections reveal a malignant tumor consisting of oval to spindle cells arranged in various patterns, including bundles, whorls, and sheets. These tumor cells are showing marked pleomorphism having enlarged nuclei with coarse chromatin and having moderate amount of cytoplasm, and multinucleated giant cells are also present with areas of hemorrhage and necrosis are observed, and with tumor-infiltrating lymphocytes and plasma cells. With all These features final diagnosis of high grade soft tissue sarcoma - Possibility of undifferentiated pleomorphic Sarcoma can be considered Figure 3 and Figure 4.

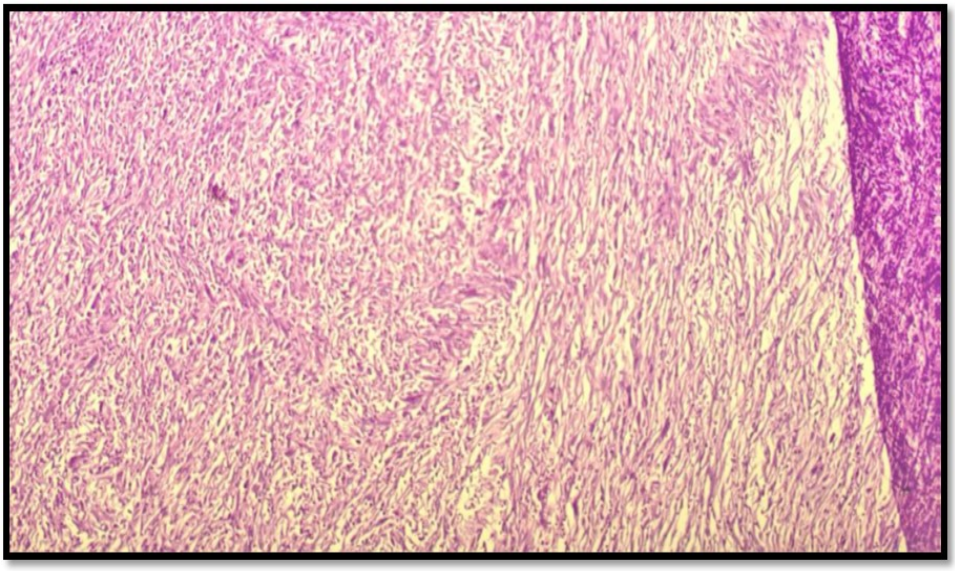

Figure 3: Microscopy H&E (10X)

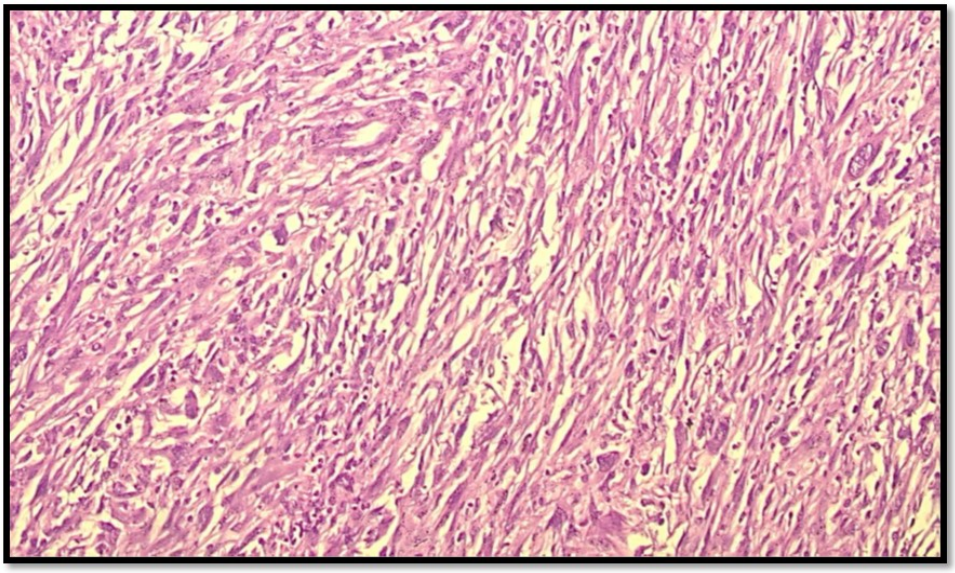

Figure 4: Microscopy H&E (40X)

Immunohistochemical staining showed that the tumor cells of histiocytes are positive for S100, CD68 and CD34 and SMA highlights vessels and negative for Desmin, PanCK, ALK and CK and the impression after IHC is Spindle cell sarcoma showing epithelioid / histiocytic morphology of Right knee Figure 5.

3. Discussion

Undifferentiated pleomorphic sarcomas (UPS) are aggressive soft tissue tumors characterized by the absence of specific markers that would classify them into a distinct cell lineage [3]. These tumors most commonly arise in the thigh and are typically seen in individuals over the age of 60 [4]. A definitive diagnosis relies on tissue biopsy, which helps determine the tumor grade and histological subtype—essential factors for assessing prognosis [5]. Imaging modalities such as MRI and CT scans play a crucial role in evaluating tumor size, location, and its relationship with adjacent structures. Immunohistochemistry is integral to diagnosing soft tissue sarcomas, with a diagnosis of UPS often being one of exclusion, made by ruling out other malignancies through a panel of immunohistochemical markers [6]. In a study by Austin H. Allen et al., a large UPS located in the posterior thigh showed strong positivity for vimentin during histopathological and immunohistochemical analysis, which supported the diagnosis of UPS [7]. Similarly, Miettinen et al. reported case involving UPS on the

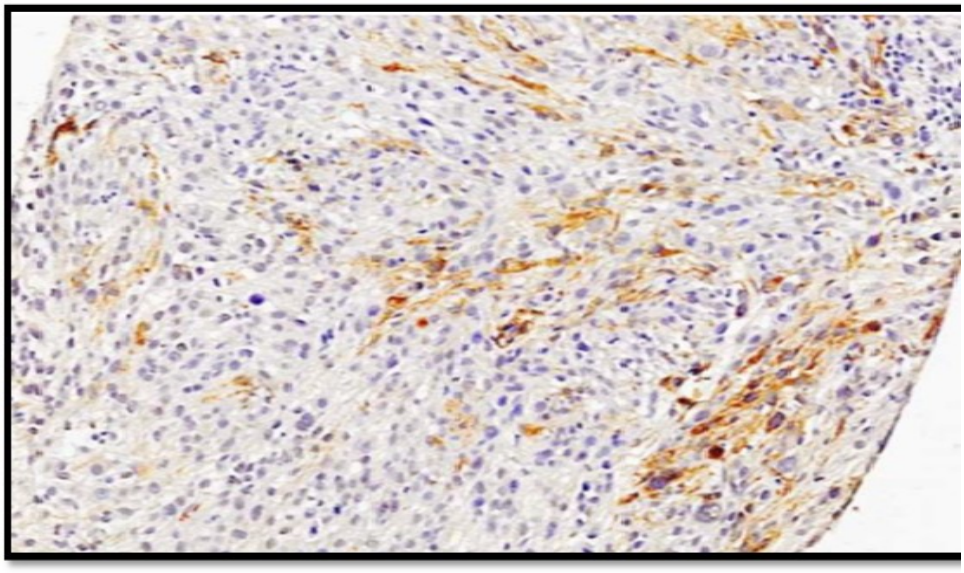

Figure 5: Immunostain showing positivity for SMA

dorsal aspect of the fifth digit, where tumor cells exhibited positive staining for CD10. Notably, CD10 is expressed in nearly half of all soft tissue sarcomas, with stronger expression commonly observed in high-grade variants. Additionally, soft tissue sarcomas often show intense staining for epithelial markers AE1 and AE3 [8].

In the present case, immunohistochemical evaluation revealed tumor cells positive for S100, CD68, and CD34, with SMA highlighting vascular structures. The tumor was negative for Desmin, PanCK, ALK, and CK. These findings contributed to confirming the diagnosis. Detection and timely intervention are critical, asUPS can be rapidly progressive and frequently diagnosed at a later stage [9]. The mainstay of treatment is wide surgical excision, aiming to remove the tumor along with a margin of healthy tissue. A positive surgical margin significantly increases the risk of local recurrence.

4. Conclusion

Undifferentiated pleomorphic sarcomas (UPS) should be considered in the differential diagnosis when a patient presents with a growing mass, especially in the extremities. Immunohistochemical staining remains a vital aspect in diagnosing soft-tissue sarcomas, and a definitive diagnosis of UPS is made by excluding other malignancies with a panel of immunohistochemical markers.

Statement of Declaration

We declare that the manuscript has been read and approved by all the authors, that the requirements for authorship have been met, and that each author believes that the manuscript represents honest work.

All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by Dr. Ranjith Kumar R and Dr. T.N. Suresh. The first draft of the manuscript was written by Dr. Ranjith Kumar R and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Article Information

Disclaimer (Artificial Intelligence): The author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.), and text-to-image generators have been used during writing or editing of manuscripts.

Competing Interests: Authors have declared that no competing interests exist.

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