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Case Report

Soft Tissue Back Mass, Case Report

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DOI: 10.31080/eccmc.2022.05.00657

Received: September 26, 2022; **Published:** October 17, 2022

Abstract

Sarcomatoid melanoma is an extremely rare subtype of melanoma and only few case reports have been reported. Sarcomatoid melanoma is a specific entity of melanoma which is characterized by weak or negative expression of melanocyte markers in the sarcomatous like component in addition to another component which is consistent with melanoma *in situ* or melanoma by morphology or immunohistochemistry. Herein, we discuss an 86-year-old male presenting with back mass. He underwent incisional biopsy of the mass which is confirmed as sarcomatoid melanoma.

Keywords: *Sarcomatoid Melanoma; Malignant Melanoma with Sarcomatoid Features*

Introduction

Malignant melanomas show a wide variety of cytological and architectural changes and hence may mimic carcinomas, lymphomas, and sarcomas [1]. Spindle cell melanomas commonly simulate spindle cell carcinomas, peripheral nerve sheath tumors and smooth muscle neoplasms [2]. Here, we report an unusual case of sarcomatoid melanoma in which part of the tumor exhibited pleomorphic spindle and epithelioid tumor comprised by sheets of cells with pleomorphism ranging from cells with small round nuclei, to large cells with prominent nucleoli and vesicular chromatin.

Malignant melanoma with sarcomatous features, or sarcomatoid melanoma, is a rare entity of melanoma. The fact that this lesion can stain negative for some melanocytic markers create a diagnostic dilemma. A broad differential diagnosis and panel of melanocytic markers should be in mind while diagnosing this lesion since melanoma is a great mimicker in diagnostic medicine and can deceive a lot of pathologists by its variable morphological patterns.

Case Presentation

A 86-year-old male was referred to our hospital for a large mass on back measuring 10.5 × 9.5 x 6.2 cm. The lesion had been present for the last 2 months but the patient did not seek for medical advice. Physical examination revealed a fungating mass, with areas of ulceration and pigmentation. he had no neurological deficits. No Pathological enlargement of any lymph nodes. Ultrasound examination showed 10 cm heterogenous mass related to soft tissues of the upper back with nonspecific increased Doppler flow.

Incisional biopsy of the lesion was done. Morphologically, it is a pleomorphic spindle and epithelioid tumor comprised by sheets of cells with marked nuclear pleomorphism ranging from cells with small round nuclei, to large epithelioid cells with prominent nucleoli and vesicular chromatin. Mitotic figures were identified, and tumor necrosis were observed. *In situ* melanoma component is not identified. Immunohistochemical stains was performed to determine the phenotype of the tumor and shows the tumor cells to be negative for MART-1, HMB-45, CD117, Desmin, Pankeratin, SMA, PAX8, CD34 and P40, while they are positive for S100 protein (favors melanoma over MPNST). H3K27me3 is retained (favors diagnosis of melanoma over MPNST). Ki 67 shows high proliferative index (40%). The case is diagnosed as Melanoma with sarcomatoid features and is confirmed by Tertiary Academic Institution.

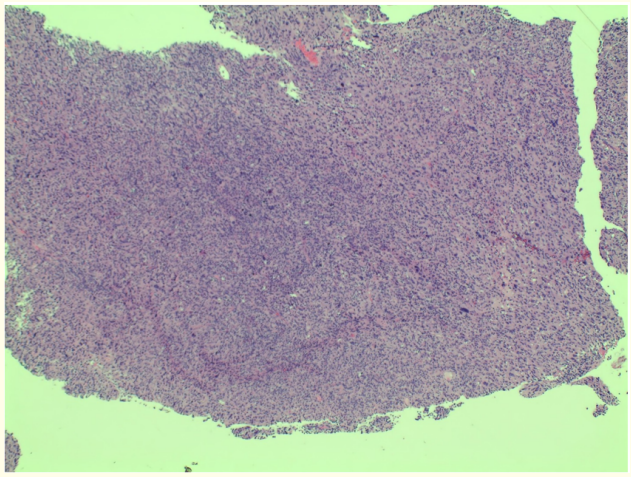

Figure A: *HE stain 10x power.*

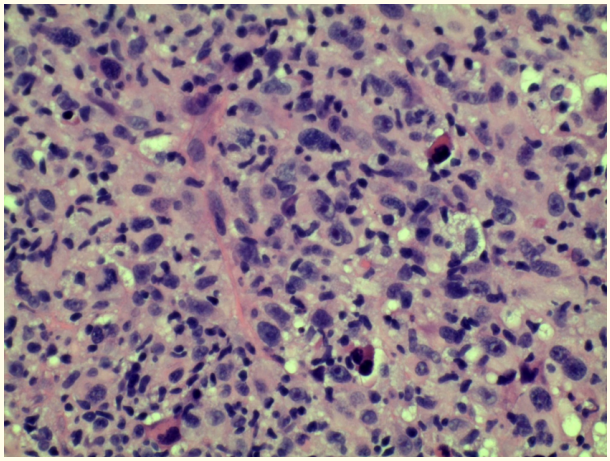

Figure B: *HE stain 40x power.*

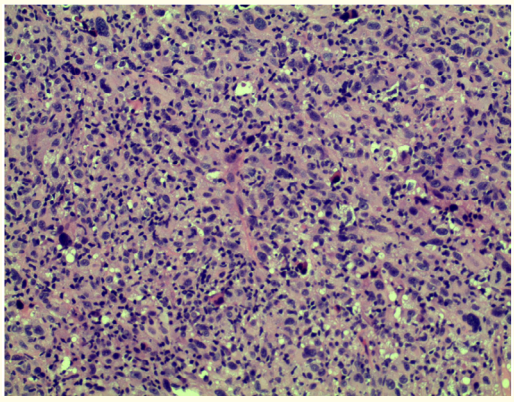

Figure C: *HE stain 20x power.*

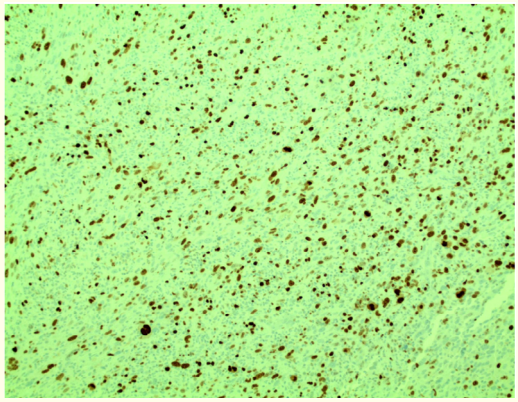

Figure D: *Ki67 index*

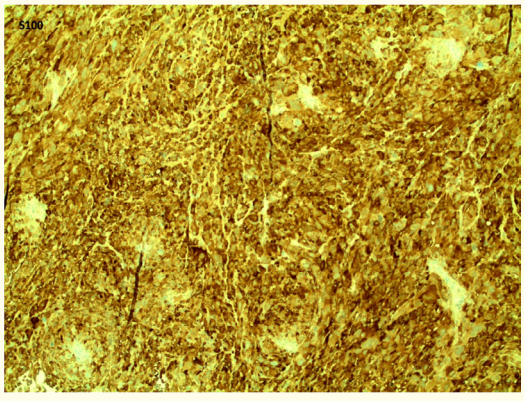

Figure E: *S100 positivity*

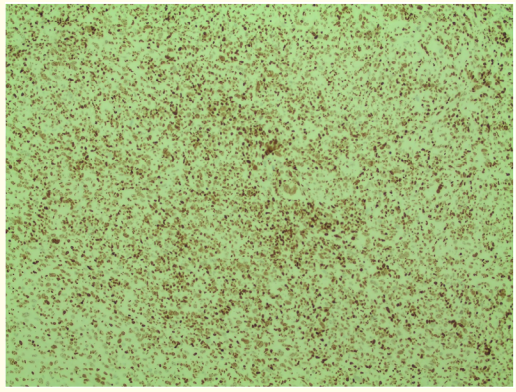

Figure F: *H3K27ME3 : RETAINED.*

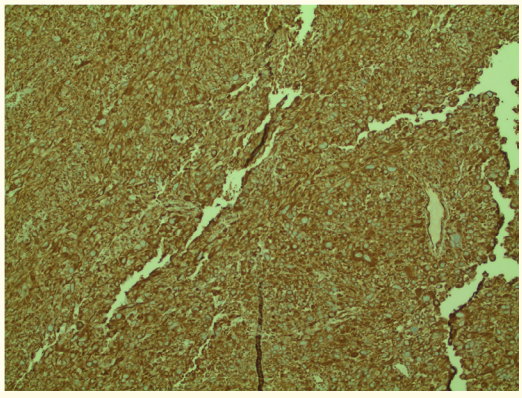

Figure G: *Vimentin Positivity*

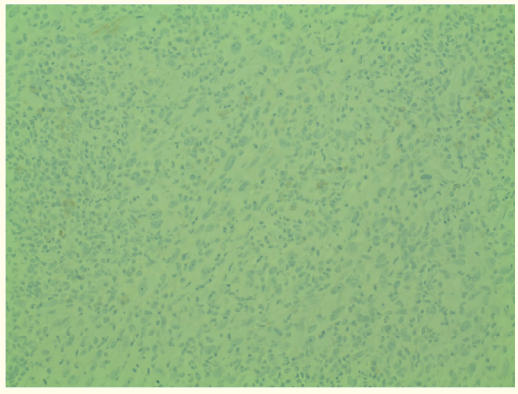

Figure H: *MELAN A negative*

Discussion

Sarcomatoid melanoma has a wide scope differential diagnosis of primary cutaneous pleomorphic lesions, which includes malignant peripheral nerve sheath tumor, sarcomatoid squamous cell carcinoma, pleomorphic dermal sarcoma and atypical fibroxanthoma [7]. These lesions can look similar morphologically. Most of times, there are no clinical clues to consider a sarcomatoid melanoma [1]. The recognition of this lesion requires a broad-spectrum approach and detailed examination of the presented morphology under the microscope [8]. Complete absence of any melanocytic markers due to poor differentiation of the lesion, can further complicate diagnosing these lesions [9].

Our case is also considered a giant melanoma. Giant cutaneous melanomas is defined as melanoma larger than 10 cm, it is rare entity usually attributed to delay of seeking medical advice from the patient side [5].

To differentiate sarcomatoid melanoma from sarcomatoid squamous cell carcinoma, actinic keratosis, malignant squamous component may be present in or nearby the lesion. Sarcomatoid squamous cell carcinomas typically show at least focal staining for cytokeratin and/or p63 or p40 [10]. Malignant peripheral nerve sheath tumors (MPNST) are histologically high-grade fascicles and broad whorls of relatively uniform, spindled cells of variable cellularity [11]. Alternating zones of cellularity and perivascular tumor cell accentuation are common with hyperchromatic, elongated to pleomorphic nuclei. Mitoses are often numerous, and geographic necrosis is common [11]. It may show origin from benign nerve sheath tumor. Focal or negative S100 protein and/or SOX10(+) in up to 95% of MPNST cases. Loss of nuclear H3K27me3 expression by IHC may present [11]. Atypical fibroxanthoma is highly atypical and pleomorphic dermal-based proliferation of spindled to epithelioid-appearing cells with scattered large, bizarre-appearing, multinucleated cells are often seen. Numerous mitoses, including highly atypical forms [12]. IHC is essential to exclude other, more specific diagnoses. The tumor is negative for cytokeratin (especially high-molecular-weight cytokeratins), p63 and melanocytic, myogenic, and vascular markers [12]. Pleomorphic dermal sarcoma is poorly marginated, deeply invasive, rapid growing lesion. Frequent mitosis, tumor necrosis and pleomorphism should present [13]. Lymph-vascular invasion exists in some cases Keratins, S100, Desmin, CD34, are negative [13]. Sarcomatoid melanoma has a sarcomatous component which further make it fair easily identified from spindle cell melanoma and desmoplastic melanoma.

H3K27me3 which is trimethylation at lysine 27 of histone H3, is affected by the polycomb repressive complex 2 (PRC2. PRC2 contains proteins essential for regulating gene expression [7]. It modifies gene expression and play many roles in embryogenesis. H3K37me3 can be lost in malignant peripheral nerve sheath tumors, meningioma and melanoma. H3K27me3 can be also lost in about 37% of melanomas [6]. Complete loss of H3K27me3 occurs in about 72% of malignant peripheral nerve sheath tumors (91% radiation associated, up to 71% neurofibromatosis type 1 associated, around 90% sporadic cases) [7]. As a result its use to rule potential mimickers is questionable.

Upregulation of the histone methyltransferase enzyme EZH2 and its histone modification H3K27me3 has been linked to melanoma progression, metastasis, and resistance to immune checkpoint blockade [15]. Histone posttranslational modifications (PTMs) have been shown to be dysregulated in multiple cancers including melanoma which includes two modifications, H3 lysine 27 trimethylation (histone H3K27me3) and H4 lysine 20 monomethylation (histone H4K20me), that are differentially expressed in the more aggressive compared to the less aggressive cell line [6].

Conclusion

Due to rarity of this lesion, little is known about the prognostic significance of further classify this lesion as sarcomatoid melanoma. Precisive diagnosis is a key in making sure to rule in cases that fulfill the diagnostic criteria of sarcomatoid melanoma. For that we recommend extensive sampling of pleomorphic spindle cell lesions in the skin to identify *in situ* melanoma component or invasive melanoma component which can make a major contribution to solve this diagnostic dilemma.

Conflicts of Interest

Authors has no conflict of interest.

Disclaimer

This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

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Volume 5 Issue 11 November 2022