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Undifferentiated pleomorphic sarcoma presenting as an exophytic pedunculated tumor on the left scapula

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Abstract

Undifferentiated pleomorphic sarcoma is a common soft tissue sarcoma. Unfortunately, any attempt to describe the line of differentiation fails. It represents a final common pathway in tumors that undergoes progression towards dedifferentiation. We report a man with an undifferentiated pleomorphic sarcoma presenting as an exophytic pedunculated tumor of the left scapula. Histopathology analysis revealed spindle-shaped cells with great pleomorphism and numerous mitoses. Immunohistochemistry showed diffuse expression of vimentin. Wide local excision was performed after an oncology consultation. After two-years of follow-up, the patient has shown no evidence of recurrence or metastases.

Keywords: undifferentiated pleomorphic sarcoma, malignant fibrous histiocytoma, soft tissue sarcoma.

Introduction

Undifferentiated pleomorphic sarcoma (UPS), formerly known as malignant fibrous histiocytoma (MFH), is a common soft tissue sarcoma; attempts to describe the line of differentiation have failed [1]. The tumor arises both in soft tissue and bones, but no true cell of origin has been identified. In 2002, the World Health Organization (WHO) declassified MFH as a formal diagnostic entity and renamed it as UPS, not otherwise specified [2]. UPS represents a final common pathway in tumors that undergooes progression towards dedifferentiation [3]. We report a man with UPS presenting as an exophytic pedunculated tumor of the left scapula.

Case Synopsis

An 83-year-old immunocompetent man was referred for evaluation of a three-month history of a fast-growing, ulcerated, pedunculated, 5cm wide tumor on the left scapula (**Figure 1**). Histopathologic analysis revealed spindle-shaped cells with great pleomorphism and numerous mitoses (18 mitotic figures per 10 high power fields), (**Figure 2**). Immunohistochemistry showed diffuse expression of vimentin in the neoplastic cells, with focal expression of CD31 and S100 protein (not shown). It was negative for HMB-45, MElanA, MiTF, CD34, actin, desmin, myogenin, P40, 34BE12, and pan-CK AE1/AE3. The patient was diagnosed with UPS based on the WHO classification of soft tissue tumors. Wide local excision was performed and after two-years of follow-up, the patient has shown no evidence of recurrence or metastases.

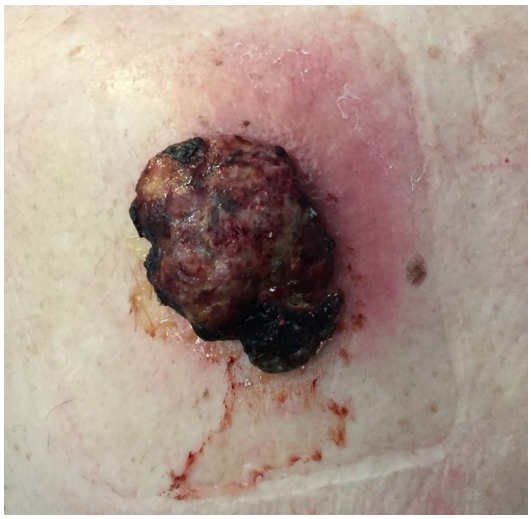

Figure 1. *Ulcerated, pedunculated, 5cm-wide tumor on the left scapula.*

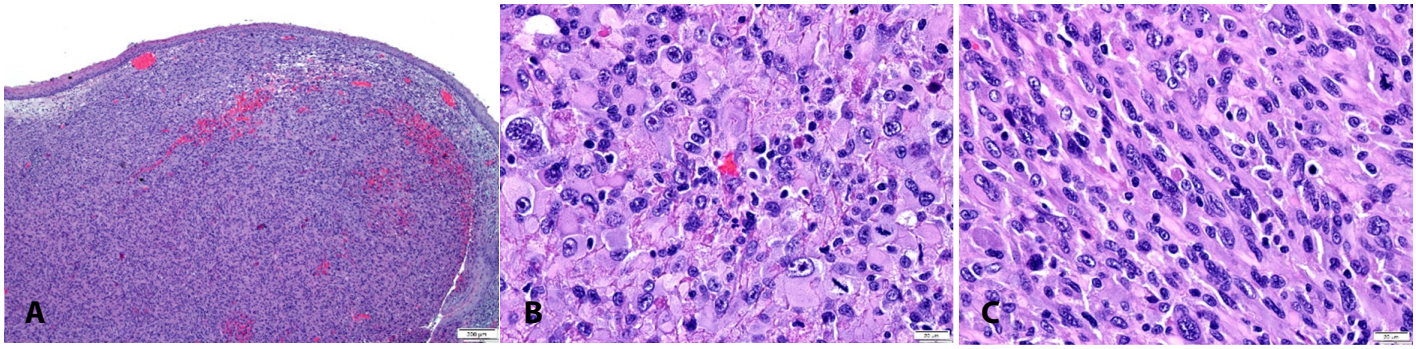

Figure 2. ***A)*** *Solid tumor with high cellularity invading the dermis, 10×.* ***B)*** *Highly pleomorphic tumor cells and frequent mitotic activity, 40×.* ***C)*** *In other areas, spindle cells predominate, 40×. H&E.*

Case Discussion

Undifferentiated pleomorphic sarcoma is a common soft tissue sarcoma that occurs most frequently in the deep soft tissues of the extremities or retroperitoneum, with most cases affecting patients between 50 and 70 years of age [4]. The most common clinical presentation is an enlarging, painless, soft-tissue mass in the thigh, typically 5-10cm in diameter. The use of immunohistochemistry is essential in the diagnostic workup of UPS, which typically demonstrates vimentin immunoreactivity [5]. The local recurrence rate is 19-31% and the metastasis rate is 31-35% [1]. The treatment of choice is surgery (Mohs micrographic surgery or wide excision with 2cm margins, when possible) [6]. Adjuvant radiotherapy can be helpful and adjuvant chemotherapy may be performed when metastases are present.

Conclusion

Undifferentiated pleomorphic sarcoma is a rare condition and most clinicians do not have experience in recognizing this tumor. Undifferentiated pleomorphic sarcoma should be part of the differential diagnostics of rapid growing tumors.

Potential conflicts of interest

The authors declare no conflicts of interest.

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