Cutaneous undifferentiated pleomorphic sarcoma is a pleomorphic dermal sarcoma

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Abstract

Pleomorphic dermal sarcoma is a cutaneous soft tissue sarcoma that presents as a rapidly enlarging tumor, typically on a sun-exposed location of elderly individuals. The neoplasm shares many similar features — clinical, pathologic, immunohistochemical and genomic — with atypical fibroxanthoma. However, adverse histologic characteristics (deep subcutaneous invasion, tumor necrosis, lymphovascular invasion, and/or perineural invasion) differentiate pleomorphic dermal sarcoma from atypical fibroxanthoma and may account for the more aggressive biologic behavior of pleomorphic dermal sarcoma: local recurrence and metastases. The features of a woman with pleomorphic dermal sarcoma are described. Her sarcoma presented as a rapidly growing ulcerated red nodule on the left side of her face. Imaging studies were performed prior to surgery. The tumor was extirpated with a wide local excision and she received postoperative radiotherapy. There has been no recurrence or metastasis at one-year follow-up. Pleomorphic dermal sarcoma has previously been referred to as a malignant fibrous histiocytoma (until the term became antiquated) and an undifferentiated pleomorphic sarcoma. However, the latter term includes not only neoplasms from the skin but also sarcomas from internal organs, retroperitoneal and osteoid origin. Therefore, when classifying this undifferentiated soft tissue sarcomas of cutaneous origin, the term pleomorphic dermal sarcoma may be preferred.

Keywords: atypical, cutaneous, dermal, fibroxanthoma, pleomorphic, sarcoma, skin, undifferentiated

To the Editor:

I read with interest the excellent report by Cerejeira et al. that described an 83-year-old immunocompetent man who developed a sarcoma that presented as a fast growing ulcerated exophytic pedunculated 5cm tumor on his left scapula [1]. The patient remained free of recurrence or metastases two years after a wide local excision was performed. Although, based on the 2002 World Health Organization classification, the man's tumor was diagnosed as an undifferentiated pleomorphic sarcoma, it may be more appropriate to refer to his tumor as a pleomorphic dermal sarcoma [2-6].

The term pleomorphic dermal sarcoma was proposed by Dr. Christopher D. Fletcher when he worked in the Department of Pathology at the Brigham and Women's Hospital in Boston, MA [2, 4-8]. He considered the pleomorphic dermal sarcoma to be distinctive from the morphologically similar atypical fibroxanthoma; whereas the atypical fibroxanthoma is confined to the dermis and lacks features such as necrosis or vascular invasion, the pleomorphic dermal sarcoma invades into the subcutis and may be associated with metastases [2, 7, 8].

Fletcher's concept of pleomorphic dermal sarcoma is generally accepted [2]. Previously this tumor was referred to as a malignant fibrous histiocytoma [2-4, 6, 9]. Subsequently, this designation has become antiquated and the term undifferentiated pleomorphic sarcoma was introduced [5, 6, 10]. However, undifferentiated pleomorphic sarcoma not only encompasses skin neoplasms but also a heterogeneous group of soft tissue malignant

neoplasms of internal organ, retroperitoneal, and osteoid origins [5, 6]. Therefore, to focus on undifferentiated soft tissue sarcomas of cutaneous origin, the term pleomorphic dermal sarcoma is recommended [2-9, 11, 12].

To emphasize the features of this rare tumor, the characteristics of another patient with a pleomorphic dermal sarcoma shall be described. An 81-year-old immunocompetent woman presented with a new and rapidly enlarging mass on the left side of her face. The tumor had grown from a small papule to a large nodule within less than four weeks. Her past medical history was significant for hypertension, hyperlipidemia, hypothyroidism and a cerebral vascular accident three months earlier; prior surgeries included a cholecystectomy, hysterectomy, total hip arthroplasty, and back surgery for sciatica.

Cutaneous examination showed an ulcerated 3.0×3.5cm red nodule on the left lateral zygomatic arch (**Figure 1**). The tumor was mobile. There was no palpable adenopathy in the neck.

Microscopic examination of a biopsy from the tumor showed a densely cellular dermal tumor, without any connection to the overlying epidermis, characterized by sheets and fascicles of spindle cells with a few bizarre and giant cells. Frequent mitoses, including atypical mitoses, were present. The tumor cells expressed CD10; immunohistochemical stains for CK5/6, pankeratin, p63, SOX-10, Melan-A, S100, desmin, SMA and CD34 were all negative.

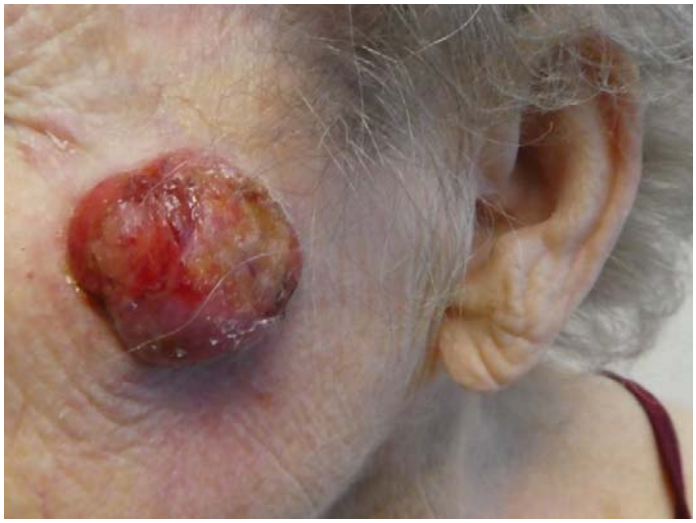

Figure 1. *Left side of the face of an 81-year-old woman shows a pleomorphic dermal sarcoma presenting as a rapidly growing ulcerated red mobile 3.0×3.5cm nodule on the left lateral zygomatic arch.*

Correlation of the clinical history, lesion morphology, and pathologic findings were those of a pleomorphic dermal spindle cell neoplasm. The differential diagnosis included an atypical fibroxanthoma and a pleomorphic dermal sarcoma. Examination of the completely excised specimen would be necessary to distinguish between these diagnostic possibilities.

A computerized tomography scan of the neck did not reveal any enlargement of the lymph nodes on the left side of her neck. A positron emission tomography/computerized tomography scan of the head and neck, thorax, abdomen, and pelvis only demonstrated intense focal uptake of 18F-2-fluoro-2-deoxyglucose within the left zygomatic cutaneous malignant mass. The tumor continued to enlarge to nearly 4×4cm prior to wide local excision.

The excision specimen showed similar pathologic changes to those observed on the biopsy. The spindle cell neoplasm consisted of highly pleomorphic cells that extended into the subcutis. Twenty-one mitotic figures per 10 high power fields, including atypical mitoses, were identified; neither necrosis, lymphovascular invasion, nor perivascular invasion were identified. The tumor cells only expressed CD10.

In summary, the morphologic appearance of the tumor and the immunohistochemical profile were similar to those of an atypical fibroxanthoma. However, the high-risk histologic features — particularly the invasion of the tumor into the subcutaneous tissue — established the diagnosis of a pleomorphic dermal sarcoma, pT2Nx. The excision margins were negative for tumor. The operative wound was closed using a cervicofacial flap.

Two months after surgery, she received 60 Gray of postoperative radiotherapy to the sarcoma site over a period of 6 weeks. There has been no recurrence or metastases after one year of follow-up (**Figure 2**). She continues to be evaluated clinically every 6 months.

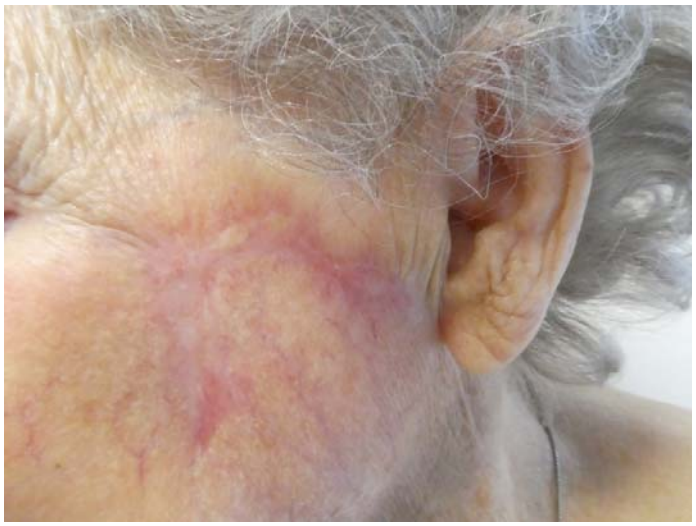

Figure 2. *Left side of the woman's face following surgery and postoperative radiation therapy. There has been no recurrence or metastases of the pleomorphic dermal sarcoma.*

The characteristics of the sarcomas of the woman described and the man reported by Cerejeira et al. are similar [1]. Both patients are octogenarians who presented with new rapidly growing spindle cell dermal tumors with numerous mitoses on potentially sun-exposed areas; the woman's tumor also extended into the subcutis. Wide local excision, with or without postoperative radiotherapy, resulted in no evidence of recurrence or metastases after one to two years of follow-up.

Pleomorphic dermal sarcoma typically occurs in elderly individuals. It presents as a rapidly growing tumor and is usually located in sun-exposed sites. The clinical differential diagnosis includes squamous cell carcinoma [2-6, 8].

Pleomorphic dermal sarcoma is a dermal tumor with no connection to the epidermis. Pathologic features include not only spindle cells with numerous mitoses, some of which are atypical, but also pleomorphic epithelioid cells and giant multinucleated cells. In addition, pleomorphic dermal sarcoma has adverse histologic features: deep subcutaneous invasion, tumor necrosis, lymphovascular invasion, and/or perineural invasion [2-4, 9]. These findings may account for the more aggressive clinical course of this neoplasm — up to 28 percent local recurrence and up to 20 percent metastases [12].

Most researchers do not consider pleomorphic dermal sarcoma to be synonymous with atypical fibroxanthoma. However, the tumors share several similar clinical, pathological and immunohistochemical features [2-6, 8]. In addition, recent genomic studies have discovered that both neoplasms have a similar molecular phenotype [11]. Hence, pleomorphic dermal sarcoma and atypical fibroxanthoma may belong to a common tumor spectrum [11].

Definitive guidelines for evaluation and management of pleomorphic dermal sarcoma remain to be established. The reported woman had preoperative computerized tomography and positron emission tomography/computerized tomography scans performed; however, data is not available regarding the role of imaging and sentinel lymph node biopsy in patients with this sarcoma. Wide local excision is recommended for tumor extirpation. Currently, there are no evidence-based recommendations for utilizing Mohs micrographic surgery for removal of the neoplasm [5, 6, 9].

The reported woman received postoperative radiotherapy. Although there are no established guidelines regarding the role of radiation therapy for patients with pleomorphic dermal sarcoma, adjuvant radiation may be effective in the setting of unresectable, locally recurrent, or regionally metastatic disease. Recent studies show a potential role for immunotherapy for some of the patients with pleomorphic dermal sarcoma [5, 6, 9, 12].

For the purpose of categorizing dermatologic neoplasms, it is reasonable to recommend that undifferentiated pleomorphic sarcoma of the skin be classified as a pleomorphic dermal sarcoma.

Potential conflicts of interest

The author declares no conflicts of interests.

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