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Pleural, Pancreatic and Muscular Metastasis of Dermato Fibro Sarcoma: A Report Case and Review of Literature.

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a relatively rare skin tumor that is considered to have intermediate malignancy; it demonstrates frequent local recurrence, but systemic metastasis is rare [1]. A 38-year-old male presented with multiple cutaneous nodular lesions of DFSP since 6 months, who underwent for CT scan that revealed distant metastasis to: the pleura, pancreas and psoas muscle.

Introduction

Dermato FibroSarcoma Protuberans (DFSP), first described by Darier and Ferrand in 1924, is a relatively rare skin tumor. DFSP is considered to have intermediate malignancy; it frequently exhibits local recurrence and, rarely, systemic metastasis [2].

Case Report

We report a 38-year-old woman, with no medical history who presented for appearance of multiple discrete nodules on neck, scalp and upper torso. The nodules gradually increased in size and enlarged to form ulcerative masses.

Biopsy was taken from different sites and its histopathological examination showed a tumor in the dermis and subcutis and was composed of interwoven bundles of spindle cells with plump nuclei arranged in a storiform pattern. Biopsy was diagnostic of DFSP.

The patient underwent a thoraco-abdomino-pelvic CT scan for pre-operative evaluation, which revealed three secondary locations: an extensive pleural mass with compressive atelectasis of the underlying lung and with invasion of the intercostal spaces (Figure 1), a pancreatic mass with heterogeneous enhancement (Figure 2), and a retroperitoneal mass at the expense of the psoas muscle (Figure 3).

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a soft tissue neoplasm of intermediate malignancy and is considered a low-grade sarcoma. Most DFSPs occur during middle age (20–50 years)[3].

The cause of dermatofibrosarcoma protuberans is not clearly understood, though studies have implicated a chromosomal translocation that results in a fusion protein that promotes tumor growth through the overproduction of platelet-derived growth factor (PDGF). Diagnosis is made via skin biopsy[4].

Although DFSP lesions have low metastatic potential, they have a pronounced tendency to recur, and there are occasional reports of metastases and death[3].

The classic form of DFSP is a low-grade tumor constituting approximately 85-90% of cases with very low risk of metastasis (approximately 5%) and local recurrence rate of about 26%. The remaining patients present with a fibrosarcomatous variant, which is associated with increased risk for the development of local recurrence (approximately 58%) as well as metastasis to vital organs(approximately 15%) [5].

Diagnosis of dermatofibrosarcoma protuberans is made with a skin biopsy, preferably an incisional or excisional biopsy. A full history and physical exam, including lymph node examination, should be completed. Some sources suggest obtaining chest imaging to evaluate for any metastatic disease before treatment, though this is not currently a general recommendation. A preoperative MRI, though not necessary, is sometimes performed to help define tumor extension before surgery [6].

The treatment of dermatofibrosarcoma protuberans is surgical removal, ideally with Mohs micrographic surgery (MMS), a surgical technique that ensures complete histopathologic margin control at the time of surgery. MMS is preferred over wide local excision, as dermatofibrosarcoma protuberans tends to have an unpredictable subclinical extension. In select situations or when Mohs micrographic surgery is not available, wide local excision may be performed with 2- to 4 cm margins [7][8][9].

The chemotherapy agent imatinibmesylate, an oral tyrosine kinase inhibitor, can be used for recurrent,

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unresectable, and metastatic dermatofibrosarcoma protuberans in adults. Imatinibmesylate competitively inhibits ATP binding to the PDGF-beta receptor, a tyrosine kinase. This slows down kinase activity, limiting the growth of the tumor, and promoting apoptosis. Patients with the t(17;22) translocation show a greater response to imatinibmesylate, and thus screening for this translocation should be performed before initiating therapy. Testing for the translocation can be performed using fluorescent in situ hybridization (FISH) or reverse transcription-polymerase chain reaction (RT-PCR). Side effects of imatinibmesylate include gastrointestinal upset, edema, fatigue, anemia, and rash. The majority of patients with dermatofibrosarcoma protuberans with the translocation respond favorably to imatinibmesylate therapy, with studies suggesting a response rate of approximately 65%. The duration of therapy varies. Some sources recommend 6 months of therapy, but this may be extended if needed. Alternatively, radiation therapy may also be used for unresectable or recurrent tumors, and adjuvant radiation may decrease the risk of local recurrence [7][8][9].

As local recurrence is common, patients require close clinical follow-up after completing treatment. The risk of recurrence is highest in the first 3 years after treatment, and thus patients should be evaluated every 3 to 6 months during this time and at least annually thereafter. Some sources advocate baseline and serial chest CT scans, especially in the case of fibrosarcomatous dermatofibrosarcoma protuberans, which has a higher risk of local recurrence and metastasis. Otherwise, routine imaging is not required unless there are symptoms to suggest metastasis [6].

Conclusion

Dermatofibrosarcoma protuberans (DFSP) is a rare superficial tumor characterized by high rates of local recurrence and low risk of metastasis. A radiological follow-up on long term is necessary to detect the local recurrence and the metastasis.

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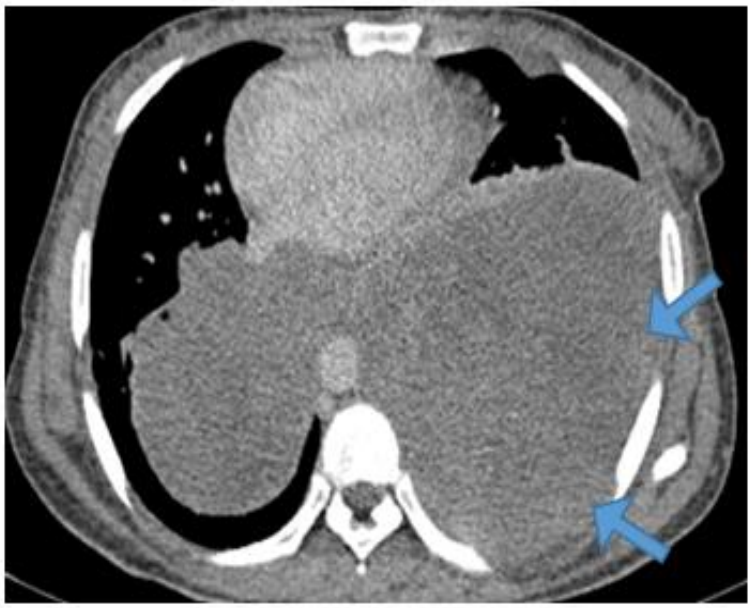

Figure 1. CT Scan showing an extensive pleural mass with compressive atelectasis of the underlying lung and with invasion of the intercostal spaces.

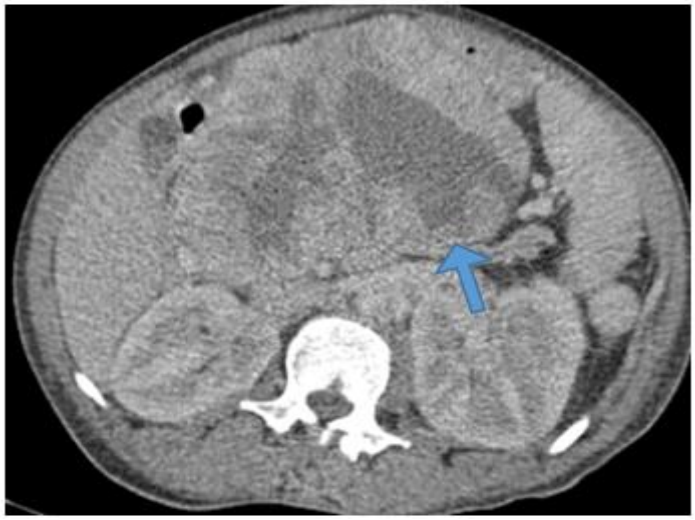

Figure 2. CT Scan showing pancreatic mass with heterogeneous enhancement.

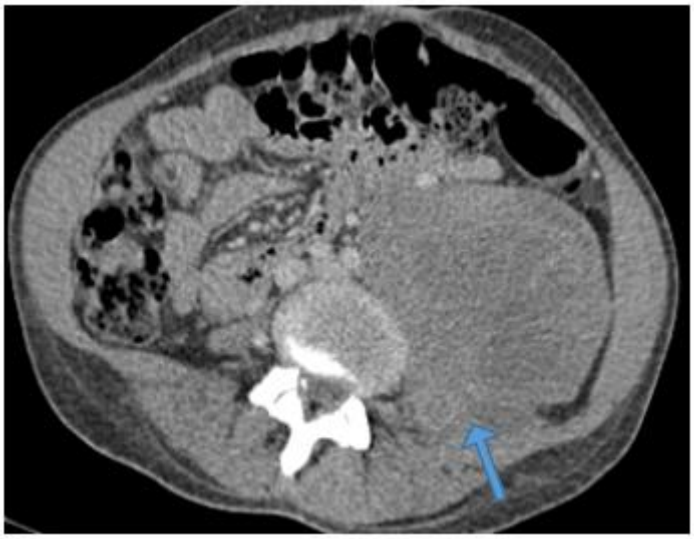

Figure 2. CT scan showing a retroperitoneal mass at the expense of the psoas muscle.