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RADIOLOGICAL-PATHOLOGICAL CORRELATION

Dermatofibrosarcoma Protuberans of the Scalp with Fibrosarcomatous Degeneration and Pulmonary Metastasis

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ABSTRACT

Dermatofibrosarcoma protuberans is a rare locally aggressive cutaneous tumor of intermediate malignancy. It is a slow-growing neoplasm with a marked propensity to recur after resection. Head and neck involvement is unusual and distant metastases are quite rare but tend to be more frequent in tumors that undergo fibrosarcomatous degeneration. We present the imaging and corresponding histopathology in a case of dermatofibrosarcoma protuberans of the scalp demonstrating fibrosarcomatous degeneration and lung metastasis.

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare slow-growing soft tissue neoplasm arising in the dermis. It usually occurs in the trunk or extremities but can involve the head and neck particularly the scalp. While often locally aggressive and frequently recurrent, metastases are rarely reported. Relatively little has been reported in the radiology literature regarding the imaging features of DFSP of the head and neck. We describe a case of DFSP of the frontal scalp with fibrosarcomatous degeneration and lung metastasis.

A 27-year-old female initially presented with painful swelling over the left eye which she first noticed 5 years earlier after minor trauma. She described an acute increase in pressure and pain in the lesion associated with headaches. On physical exam, a fluctuant mildly tender 8-cm mass was noted arising from the left forehead. After obtaining contrast-enhanced computed tomography

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(CT), she was referred to plastic surgery, and 1 month later underwent wide local excision to the periosteum with 1 cm surgical margins. Histopathological examination and immunohistochemical analysis were consistent with DFSP demonstrating areas of low-grade fibrosarcomatous transformation; surgical margins were positive. The patient was scheduled for postoperative chemotherapy with imatinib, but failed to return for treatment.

Eight months later, she presented again after multiple episodes of bleeding from a recurrent left forehead mass. MRI was obtained during admission [Figure 1], and she subsequently underwent repeat wide local excision to obtain negative margins and a selective neck dissection. Pathology was positive for recurrent DFSP with no evidence of tumor involvement in resected lymph or glandular tissue. No invasion of deep structures was found though surgical margins were again positive. Concurrent chemoradiation therapy was planned, but the patient was again lost to follow-up.

Two years later, the patient presented with a several week history of left-sided chest discomfort, cough, and shortness of breath. Physical examination revealed a third recurrence of her frontal scalp tumor now measuring 12 cm with extensive superficial ulceration. Repeat MRI of the head [Figure 3] and CT of the thorax [Figure 4] demonstrated new pulmonary metastasis. She again underwent debulking of her scalp lesion and is currently being followed by otolaryngology and oncology with plans for possible repeat chemotherapy.

Radiological features:

First described in 1924, dermatofibrosarcoma protoberans (DFSP) is a rare neoplasm of spindle cells arising in the dermis, usually in the trunk or extremities.[1] Its estimated incidence in the United Sates is 4.5 cases per million per year and it accounts for approximately 6% of soft tissue sarcomas. Although more common in young and middle aged adults, tumors can occur at any age and involve any body part.[2,3] Overall 5-year recurrence-free survival rate for each group has been reported as 81% in standard DFSP and 28%, DFSP with fibrosarcomatous degeneration.[4] Metastases do occur but are rare, occurring in <10% of cases, but most often after local recurrence.[5] Lung involvement is most common with metastasis to the abdomen and local lymph nodes also being reported.[5,6] Scalp DFSPs have also been rarely reported to demonstrate intracranial extension.[5,7]

Imaging of DFSP is usually performed to assess local extent of the lesion as well as to identify any distant metastasis, and may assist in surveillance for recurrence. In the case presented, initial contrast enhanced CT [Figure 2] demonstrated a rounded protuberant heterogeneously enhancing mass arising from the left frontal scalp. There was no evidence of underlying osseous invasion and no other lesion was present. MRI of her recurrent tumor showed a large mass predominantly hypointense on T1 and hyperintense on T2 with heterogeneous enhancement after gadolinium. Areas of central necrosis or hemorrhage were evident with extensive superficial ulceration [Figures 1 and 3]. Contrasted CT of the chest during the patient's third admission, revealed a large, rounded mass filling the majority of the right middle lobe and compressing the right heart representing a lung metastasis [Figure 4].

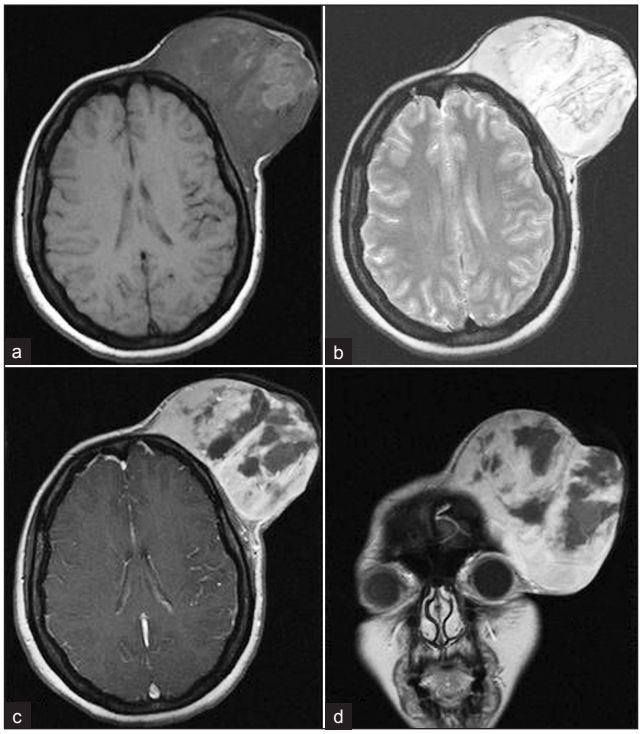

Figure 1: 27-year -ld with DFSP. (a) Axial T1 pre-contrast image demonstrates a large exophytic mass slightly hypointense to gray matter which has recurred within the left frontal scalp. (b) The mass is predominantly T2 hyperintense and demonstrates prominent heterogeneous enhancement after gadolinium on (c) axial and (d) coronal T1 post-contrast images. Areas of central necrosis or hemorrhage are noted.

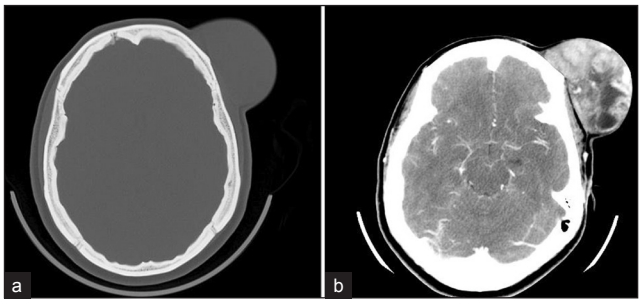

Figure 2: 27-year-old with DFSP. Axial CT head (a) without and (b) with contrast demonstrate a large protuberant soft tissue mass arising from the left frontal scalp. The mass enhances heterogeneously after contrast with areas of central hypovascularity but no invasion of the calvarium.

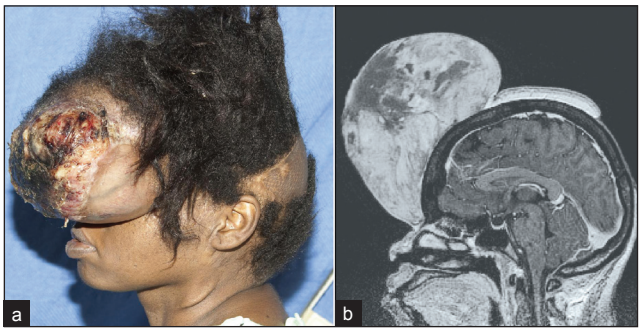

Figure 3: 27-year-old with DFSP. (a) Patient photograph and (b) corresponding saggital T1 post-contrast MRI demonstrating the recurrent DFSP which is larger, and more ulcerated than previously with continued heterogeneous enhancement.

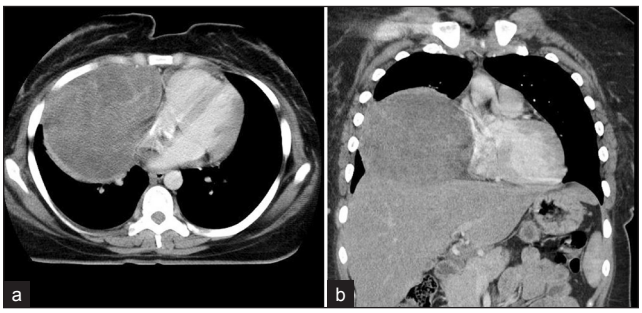

Figure 4: 27-year-old with DFSP. (a) Axial and (b) coronal post-contrast CT images of the chest demonstrate a large round mass with mild enhancement in the right lung centered in the right middle lobe and causing mass effect on the adjacent right atrium.

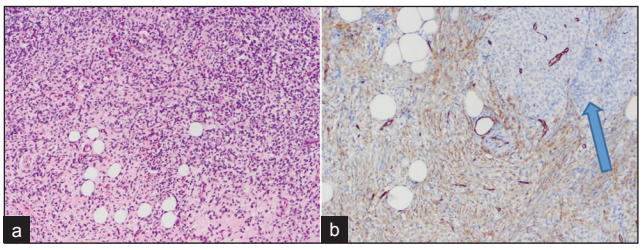

Figure 5: 27-year-old with DFSP. (a) H and E stain demonstrating spindle-type cells with areas of classic "storiform" or "cart wheel" pattern. (b) Immunostain for CD34 shows predominantly strong positivity but areas with loss of CD34 immunopositivity representing fibrosarcomatous transformation are also present (arrow).

The CT and MR characteristics of DFSP were previously described by Kransdorf and Meis-Kindblom in a series of 11 patients of which only one had a scalp lesion. In their study, most tumors were nodular or lobular lesions with attenuation similar to muscle and moderate inhomogeneous enhancement.[2] On MR imaging, T1 low signal and T2 high signal were consistent features with enhancement seen after gadolinium. Fibrosarcomatous areas may tend to be more necrotic and account for some of the heterogeneity seen on contrast-enhanced MR and CT scans.[2] Torreggani et al, in a series of 10 cases of DFSP of the trunk and extremities, also make the case for utilizing STIR (or other T2 fat saturated) sequences to image these lesions. They reported several cases where mass was difficult to visualize on conventional T2 sequences due to the similar signal intensity of adjacent subcutaneous fat.[3]

Older imaging techniques with catheter-based angiography have shown DFSP to be mildly to moderately hypervascular. Bone scintigraphy has also been occasionally used in evaluation of these tumors and in three cases showed increased uptake of the radiopharmaceutical on delayed imaging.[2]

Histopathological features

Clinically, DFSP initially presents as a cutaneous pink to red-bluish painless plaque which grows more nodular with time.[8] Tumors are typically superficial in location and have a nodular exophytic growth pattern.[2] Larger tumors may ulcerate and tend to be superficially mobile but fixed to the deeper structures such as muscle or fascia.[2,8] Slow tumor growth is characteristic and there is often a delay in diagnosis as patients can ignore the lesion for some time due to its indolent nature. Lesions most commonly arise in the trunk with the extremities being the next most common site followed by the head and neck. There is also a slight male predominance and its incidence in African Americans is almost twice that of Caucasians.[3,8]

Histolopathogically, DFSP shows a distinct "storiform" or "cartwheel" arrangement of uniform appearing fibroblasts [Figure 5]. Immunohistochemical staining demonstrates strong positivity for CD34 (sensitivity 84-100%) and vimentin.[5,8,9] DFSP can rarely present as a more aggressive fibrosarcomatous variant which occurs in around 10-15% of cases. Areas of fibrosarcoma are characterized by increased cellularity and loss of CD34 imunopositivity.[8] Patients with this more aggressive variant tend to be more prone both to local recurrence and distant metastasis. Currently only around 150 cases of transformed DFSP have been reported and of these 13% had distant metastasis.[10] Older studies have reported metastatic rates of around 21% in DFSP cases with fibrosarcoma.[2]

Standard treatment of DFSP is wide local excision with margins of at least 2 cm and many now advocate Moh's micrographic surgical technique as achieving better results.[9-11] While the prognosis is excellent with complete resection, there is a marked tendency to recur if adequate surgical margins are not obtained. Radiotherapy also has a role in treatment as the tumor has been shown to be radiosensitive; chemotherapy with selective tyrosine kinase inhibitory such as imatinib mesylate has also shown good results.[8]

In conclusion, DFSP is a rare tumor with a classic clinical presentation. While imaging features are often suggestive but not specific, cross-sectional techniques, particularly MR, can aid in clearly delineating the anatomic structures involved and rule out invasion of deep structures. While

metastases are rare, in patients with fibrosarcomatous degeneration and multiple recurrences, surveillance imaging for metastasis may play a role.

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