Dermatofibrosarcoma Protuberans Presenting as a Subcutaneous Mass in the Frontal Muscle of the Forehead

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a cutaneous fibrohistiocytic tumor of intermediate malignancy that typically arises in the dermis and subsequently infiltrates the subcutaneous tissue. Here, we present an extremely rare case of DFSP confined to an intramuscular lesion on the forehead. A 61-year-old female patient presented with a tender bicuspid subcutaneous mass on her forehead that had been gradually increasing in size for 10 years. The patient underwent surgical resection and pathological examination. Microscopically, cells with spindle-shaped nuclei and scant eosinophilic cytoplasm were arranged in a haphazard or monotonous pattern. However, no relationship was observed between the tumor and skin appendages. At the superficial margin, tumor cells infiltrated the surrounding frontalis muscle tissue, demonstrating typical DFSP morphology. In immunohistochemical studies, neoplastic cells were identified as anti-CD34 positive, which was a decisive diagnostic factor.

Key words: CD34, dermatofibrosarcoma protuberans (DFSP), intramuscular mass

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a cutaneous fibrohistiocytic tumor of intermediate malignancy that typically arises in the dermis and subsequently infiltrates the subcutaneous tissue. Typically, it presents as a plaque or nodule on the trunk or proximal portion of extremities in young or middle-aged adults. The tumor shows locally aggressive behavior and a significant capacity for local recurrence but rarely metastasizes. In this report, we present an extremely rare case of DFSP confined to a subcutaneous lesion on the forehead.

Case report

A 61-year-old woman presented with an ellipsoidal, indurated plaque 63 × 22 mm in size. The plaque developed into a tender, double-headed subcutaneous mass on the forehead (Fig. 1). The tumor had been growing slowly for approximately 10 years. Magnetic resonance imaging (MRI) showed a relatively well-defined mass within the frontalis muscle (Fig. 2). A lipoma, epidermal cyst, or tumor related to the supraorbital nerve was suspected as the differential diagnosis.

An excisional biopsy was performed under general anesthesia. The tumor was excised via two transverse skin incisions in the middle of the forehead and along the anterior hairline. The tumor was located within the layer of the frontal muscle of the periosteum. Tumor margins were clear and completely resected visually, leaving the subcutaneous tissue (Fig. 3).

Microscopically, the neoplasm contained cells with spindle-shaped nuclei and scant eosinophilic cytoplasm, arranged haphazardly or in a monotonous storiform pattern. However, no relationship was observed between the tumor and the cutaneous adnexal structures (Fig. 4a). Neoplastic cells infiltrated the surrounding frontalis muscle tissue at the superficial margin, consistent with typical DFSP (Fig. 4b). Immunohistochemical studies revealed striking cytoplasmic positivity in neoplastic cells for anti-CD34 (Fig. 4c) and vimentin but negative for epithelial membrane antigen (EMA), S100, smooth muscle actin (SMA), and cytokeratin (AE1/3).

Based on these findings, we diagnosed the tumor as intramuscular DFSP. Two years after tumor resection, no recurrence or metastasis was observed, and the course was...
good without complications such as ptosis. In addition, no further treatment was performed (Fig. 5).

**Discussion**

Since DFSP was first described by Darier and Ferrand in 1924\(^1\), the cutaneous origin of DFSP has been confirmed. The tumor appears to invariably arise from the dermis and infiltrate the subcutaneous tissue. However, the existence of purely subcutaneous DFSP was reported in the first and fourth editions of Enzinger and Weiss’s soft tissue tumors\(^2\). In the first and second editions of this textbook, the authors indicate that occasional cases of DFSP may be confined to the subcutaneous tissue and lack dermal involvement. This variant was termed deep DFSP by Cascajo\(^3\) owing to the lack of dermal involvement and muscle involvement. Its histopathological and immunohistochemical features are indistinguishable from those of a typical DFSP. Likewise, the present case was histopathologically indistinguishable from a typical DFSP. There have been a few previous reports on deep DFSP\(^4-7\).
However, we were unable to identify any report on DFSP confined to the frontal muscles.

Immunohistochemically, the deep DFSP exhibited strong vimentin and CD34 staining. As CD34 has been identified as a sensitive marker for periadnexal (hair follicles and eccrine glands and ducts), perineurial, and perivascular fibroblasts in normal skin, these cells are considered as possible progenitors of neoplastic cells of DFSP.

The exclusive involvement of a subcutaneous lesion in deep DFSP cases may complicate the histopathological diagnosis. The differential diagnosis of deep DFSP includes malignant fibrous histiocytoma, malignant peripheral nerve sheath tumor, storiform perineurial fibroma (perineurioma), and benign fibrous histiocytoma of the subcutaneous tissue. Malignant fibrous histiocytoma differs from DFSP owing to the presence of pleomorphic cells with prominent nuclei that often exhibit atypical mitotic figures. Malignant peripheral nerve sheath tumors contain interlacing spindle cell bundles with hyperchromatic nuclei and sometimes pleomorphic nuclei. Perivascular whirling by tumor cells is a typical feature.

Immunohistochemically, only 50% of these tumors were S100-positive, suggesting different degrees of cellular differentiation. Unlike DFSP, malignant peripheral nerve sheath tumors exhibit dense fascicles of atypical hyperchromatic cells interspersed with hypocellular areas. Perineuriomas are well-circumscribed, non-encapsulated tumors, often located in subcutaneous tissues. Neoplastic cells are arranged in whorls or short fascicles. The focal storiform growth pattern in our case may suggest DFSP. Like other benign fibrohistiocytic lesions, perineurioma immunohistochemically differs from DFSP due to the absence of CD34 immunostaining.

It is recommended that a typical DFSP is resected with a margin of 2–3 cm. However, considering the location of the lesion in the present case, the functional and esthetic loss due to extensive resection was deemed to be significant. The treatment in our case may have been inadequate, as wide

Fig. 4. Pathological findings.
(a) The neoplasm contains cells with spindle-shaped nuclei and scant eosinophilic cytoplasm arrange haphazardly or in a monotonous storiform pattern. No relationship between the tumor and cutaneous adnexal structures can be observed (H&E, ×40).
(b) At the superficial margin, neoplastic cells infiltrate the surrounding frontal muscle tissue, consistent with typical dermatofibrosarcoma protuberans (H&E, ×100).
(c) Immunohistochemistry is positive for CD34 (×100).

Fig. 5. Postoperative photograph.
(a), (b): Two years after surgery, no recurrence, metastases, or complications were detected.
excision was not performed; however, we completely excised the tumor with sharply demarcated borders. The clinical course was uneventful, and no recurrence was observed. Follow-up of these patients is essential to prevent recurrence.

In conclusion, we report an extremely rare case of DFSP that clinically presented as an intramuscular mass on the forehead with no involvement of the dermis.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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