Dermatofibrosarcoma protuberans (DFSP) represents a spectrum of mesenchymal spindle cell tumors that typically involve both dermis and subcutis. Presented herein are six cases of DFSP, four of which were initially diagnosed by FNAB. The cytologic features useful in the identification of this lesion on Papanicolaou- and Diff-Quik-stained smears are discussed. Chief among these are the storiform stromal fragments, presence of entrapped adipose tissue and the recognition of fibrohistiocytic spindle cells. The potential pitfalls and the differential diagnostic possibilities of spindle-cell lesions, particularly those of fibrohistiocytic origin are discussed. Diagn Cytopathol 1993;9:145-150. © 1993 Wiley-Liss, Inc.

Key Words: Fine-needle aspiration; Soft-tissue tumors; Dermatofibrosarcoma

Dermatofibrosarcoma protuberans (DFSP) is an uncommon mesenchymal spindle cell neoplasm that typically involves dermis and subcutis. While the histopathology of this tumor and its variants has been well described,1-9 the aspiration cytology of these neoplasms has not been fully characterized. Although Perry et al. in 198610 described the fine-needle aspiration biopsy (FNAB) cytology of a DFSP that was metastatic to the lung, there has been no formal attempt to characterize the spectrum of FNAB findings observed in a series of lesions of DFSP. Thus, we present a series of six cases of DFSP in which FNAB was used as a diagnostic procedure. The cytologic criteria that aid in the classification of this spindle cell neoplasm are presented. The potential pitfalls associated with this diagnosis are also discussed.

Materials and Methods

A search of our files of FNA biopsies from The University of Texas Health Science Center at San Antonio (UTHSCSA) and the Medical College of Virginia (MCV) yielded aspirates with the diagnosis consistent with or suggestive of DFSP. Three aspirates from UTHSCSA and three from MCV were retrieved. All aspirates were performed by a pathologist and had confirmation by surgical biopsy and/or excision. Because of the superficial location of these nodular masses a standard FNAB technique without anesthesia was used.11 Smears and in one case (1) a cell block were made from the aspirated material. Smears were either air-dried or fixed in 95% alcohol and stained with a modified Wright-Giemsa stain (Diff-Quik; American Scientific Products, Columbia, MD) or Papanicolaou stain, respectively. In addition, in Case 1, minute fragments obtained from a 10% formalin rinse of 22-gauge needles were routinely processed and embedded in paraffin for routine histology. Hematoxylin and eosin stains were performed on the slides obtained from this cell block. The aspiration biopsies of six cases of DFSP were reviewed and correlated with their previous or subsequent histology.

Results

This series was comprised of four women and two men who ranged in age from 22 to 59 yr. Table I lists the clinical presentation of these patients as well as the diagnostic role played by FNAB. Four of six patients presented with no prior diagnosis of any soft-tissue neoplasms, the remaining patients (cases 5 and 6) presented with a prior diagnosis and surgical treatment of DFSP. All patients presented with similar complaints of a nodular mass usually slowly enlarging over a long period of time. Examination revealed these masses to be located in the superficial dermis and with the exception of an occipital scalp lesion, all masses were located in the upper trunk. FNAB was the initial diagnostic procedure in 4 of 6 cases.
and confirmed recurrences in an area of previous excision in the remaining two cases.

The cytologic features of DFSP are listed in Table II. Aspirated material was stained using both Diff-Quik and Papanicolaou stains. Although all the criteria listed in Table II can be seen using the Diff-Quik stain, the Papanicolaou stain enhances nuclear detail and the occasional mitotic figure. All aspirates were of moderate to high cellularity with both scattered individual and storiform arrays of spindle cells (Fig. 1A, B), as well as variably sized, irregular stromal fragments. Aspirates were frequently bloody and often contained adipose tissue entrapped within the fragments. The storiform pattern and infiltration of adipose tissue was easily identified at low power. The stromal fragments, detailed best with the Diff-Quik stain, were metachromatic with a finely fibrillar matrix. Embedded within the stroma were variable numbers of bland spindle cells with abundant cytoplasm but ill-defined cytoplasmic borders (Fig. 2). Inconspicuous, translucent capillaries often traversed the larger fragments.

The fine granular cytoplasm was pale to slate blue (Diff-Quik, Fig. 3) or light green (Papanicolaou, Fig. 4) and was seldom vacuolated. Often bipolar cytoplasmic processes were present on discohesive or loosely arranged spindle cells. The spindle-cell nuclei were variably sized with smooth, round nuclear membranes and fine homogeneous chromatin. Small distinct nucleoli or multiple small chromocenters were usually present and enhanced with Papanicolaou stain. There was very little cellular or nuclear pleomorphism in most of the aspirates. Mitotic figures were scarce and never atypical. However, in one case of recurrent DFSP (case 6), numerous, discohesive pleomorphic spindle cells with prominent small nucleoli were admixed with the usual bland spindle cells and stroma (Fig. 5).

**Discussion**

FNAB is routinely used to distinguish among carcinoma, lymphoma, and sarcoma. However, FNAB interpretation becomes more difficult when an attempt is made to subclassify soft tissue neoplasms beyond the benign and malignant categories. Several series have examined the role of FNAB in the cytodagnosis of soft-tissue tumors (STT). In their 1986 study, Miralles et al. favored

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**Table I. Clinical Presentation and Role of Fine-Needle Aspiration Biopsy**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/sex</th>
<th>Clinical data</th>
<th>FNAB site</th>
<th>Role of FNAB</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>22/M</td>
<td>Mass × 1 year</td>
<td>Occipital scalp</td>
<td>Initial diagnosis</td>
</tr>
<tr>
<td>2</td>
<td>59/F</td>
<td>Nodular mass × 2 yr slowly enlarging</td>
<td>Back</td>
<td>Initial diagnosis</td>
</tr>
<tr>
<td>3</td>
<td>26/M</td>
<td>Nodular mass × 4 yr recently enlarging</td>
<td>Left scapula</td>
<td>Initial diagnosis</td>
</tr>
<tr>
<td>4</td>
<td>47/F</td>
<td>Nodular mass × 10 yr recently enlarging</td>
<td>Right upper arm</td>
<td>Initial diagnosis</td>
</tr>
<tr>
<td>5</td>
<td>39/F</td>
<td>Prior Dx DFSP mass × 1 year area of prior excision</td>
<td>Right chest wall</td>
<td>Confirm recurrence</td>
</tr>
<tr>
<td>6</td>
<td>25/F</td>
<td>Prior Dx DFSP new onset mass area of prior excision</td>
<td>Shoulder</td>
<td>Confirm recurrence</td>
</tr>
</tbody>
</table>

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**Table II. Cytologic Features of Dermatofibrosarcoma Protubersans**

<table>
<thead>
<tr>
<th>Pattern</th>
<th>Cytologic Features of Dermatofibrosarcoma Protubersans</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blood</td>
<td>Adipose tissue (entrapped)</td>
</tr>
<tr>
<td>Moderate cellularity</td>
<td>Moderate cellularity</td>
</tr>
<tr>
<td>Stromal fragments</td>
<td>Metachromatic</td>
</tr>
<tr>
<td>Fibrillar matrix</td>
<td>Variable cellular</td>
</tr>
<tr>
<td>Occasionally cellular</td>
<td>Storiform pattern</td>
</tr>
<tr>
<td>Inconspicuous vasculature</td>
<td>Discohesive cells</td>
</tr>
<tr>
<td>Plump, oval, and spindle</td>
<td>Usually bland</td>
</tr>
<tr>
<td>Cytoplasm</td>
<td>Moderate in amount</td>
</tr>
<tr>
<td>Pale</td>
<td>Finely granular</td>
</tr>
<tr>
<td>Bipolar processes (discohesive cells)</td>
<td>Ill-defined borders (cells embedded in matrix)</td>
</tr>
<tr>
<td>Nucleus</td>
<td>Smooth, round membranes</td>
</tr>
<tr>
<td>Variously sized</td>
<td>Homogeneous chromatin</td>
</tr>
<tr>
<td>Smooth, round membranes</td>
<td>Nucleoli/chromocenters</td>
</tr>
<tr>
<td>Infrequent mitoses</td>
<td></td>
</tr>
</tbody>
</table>

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the classification of soft tissue sarcomas into five categories: I, low grade; II, myxoid; III, monomorphic; IV, round cell; and V, pleomorphic. The classification of an aspirate into the low grade category would imply a population of spindle cells with minimal cytologic atypia, a low mitotic index, and absence of necrosis. The diagnostic possibilities include not only DFSP and low-grade fibrosarcoma but benign entities such as nodular fasciitis, fibrous histiocytoma, and fibromatosis. Nguyen16 has suggested dividing STT into only three categories: I, pleomorphic cell tumors; II, spindle cell tumors; and III, small round cell tumors. Once classified into one of these broad categories, additional cytologic criteria and immunocytochemistry can be applied to help further classify these tumors. This technique is especially useful if enough material is aspirated to produce a cell block. Based on this classification schema, DFSP would be assigned to the second category, spindle cell tumors.

As in other areas of aspiration biopsy, the importance of clinicopathologic correlation can not be overemphasized. DFSP is usually a firm, nodular (protuberant) cutaneous mass that has been slowly enlarging over a prolonged period of time, often years. Occasionally there is a history of antecedent trauma. Although typically located on the trunk or proximal extremities, a small percentage occur as scalp lesions.20

The initial classification of an aspirate as a STT, possibly DFSP begins with the identification of the neoplastic cell as fibrohistiocytic, rather than neural or muscular. This separation can sometimes be made on the basis of nuclear shape alone. Reactive fibroblasts often have angulated nuclei, while those of DFSP tend to have a round/oval shape. Nuclei from smooth muscle tumors are often described as cigar-shaped with blunt ends; those from neural lesions are twisted, slender, and elongate. In general, DFSP fibroblasts tend to have moderate amount of
cytoplasm and like reactive fibroblasts will have uni- or bipolar cytoplasmic processes. Unlike fibrotic/sclerotic lesions, aspirates of DFSP yield moderately cellular smears with discohesive cells as well as minute fragments of tissue. These fragments may be large enough to process as a cell block; in either case, they demonstrate the storiform arrangement of cells in the stromal fragments. These fragments are strikingly metachromatic on the Diff-Quik stain. In addition to the fibrohistiocytic cells arranged in a storiform array, background elements are also important and help narrow the differential. Chief among these features are the presence of entrapped adipose tissue, a delicate inconspicuous network of capillaries, and the absence of inflammatory cells (Table I).

The differential diagnosis for DFSP includes both benign and low grade, malignant spindle cell lesions. While immunochemistry can be used successfully to distinguish lesions of neural or muscular origin, it is less useful in attempts to distinguish the variety of fibrohistiocytic lesions that may be encountered as superficial dermal masses. The cytologic features of nodular fasciitis (NF), a troublesome soft-tissue pseudotumor that may be difficult to distinguish from DFSP, have been previously described.\(^{21,22}\) Although the spindle cells of both entities can vary in size and shape, the proliferating fibroblasts of NF tend to have nuclei with less rounded, more angular and prominent nucleoli. Both lesions also tend to have varying amounts of myxoid matrix; however, there is a more haphazard arrangement of the cells within the matrix in NF, as well as an intermingling of inflammatory cells (leucocytes, eosinophils, and macrophages). Another similar STT is a spectrum of lesions termed fibrous histiocytoma (dermatofibroma, histiocytoma, sclerosing hemangioma). Fibrous histiocytomas may occur in the deep (noncutaneous) soft tissues as well as skin and subcutis.\(^{23}\) Hemosiderin deposits within these cutaneous masses may result in
FNAB: DERMATOFIBROSARCOMA PROTUBERANS

Fig. 4. Details of loosely cohesive spindle cells with cytoplasmic processes, smooth nuclear membranes, homogeneous chromatin, and small nucleoli (Papanicolaou, ×1,000).

Fig. 5. Aspirate from case 6, demonstrating high cellularity with discohesive and more pleomorphic spindle cells having prominent but small nucleoli. These are mixed with the usual bland spindle-tumor cells most commonly seen in the other cases of DFSP in this series (Diff-Quik, ×400).

red to brown discoloration of the overlying skin. This may help clinically to distinguish them from DFSP. Aspirates from these masses produce stromal fragments of increased cell density and less intercellular matrix. In addition, background elements that may be present include plump histiocytes and multinucleate tumor giant cells that are often pigment laden, as well as inflammatory cells. The presence of entrapped adipose tissue suggests an infiltrative pattern as seen in DFSP, rather than the more circumscribed histiocytoma. Fibromatosis and low-grade fibrosarcoma are also problematic, especially since an adequate sample is sometimes difficult to obtain because of tight cellular cohesion. The cytologic features of these fibroblasts can vary considerably, from the deceptively bland low-grade sarcoma to the worrisome nuclear pleomorphism of aggressive fibromatosis. Other less likely entities that may be included in the differential diagnosis of DFSP include Kaposi’s sarcoma and amelanotic melanoma. Aspirates from Kaposi’s sarcoma typically contain delicate, discohesive, elongate spindle cells in a background of abundant blood. Even less likely to be confused cytologically with DFSP are melanomas. Aspirates from these lesions may show pleomorphic spindle cells, but with minimal stroma and no discernible storiform pattern. Although clinical correlation may be very helpful in a majority of cases, there are instances when a specific diagnosis may not be possible. In these cases, a more general diagnosis such as “spindle-cell tumor” or “mesenchymal lesion” with a list of probable diagnoses may have to suffice.

Because of the propensity of DFSP to recur, surgical treatment includes ample (3-cm) margins. Even with these margins the recurrence rate can be as high as 20%. Unfortunately, the likelihood of metastatic disease in-

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crease with multiple recurrences. An additional problem is the occurrence of frank sarcomatous change; areas of fibrosarcoma (FS) and malignant fibrous histiocytoma (MFH) can be found in initial, as well as recurrent DFSP and are associated with a less favorable prognosis. Because FNAB samples very discreet areas, sarcomatous change can be missed entirely on FNAB (sampling error) or can be diagnosed as MFH or FS. The variability of areas resembling giant cell fibroblastoma: report of two cases. Histopathology 1985;9:921-38.


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