Pigmented dermatofibrosarcoma protuberans with prominent meningotheelial-like whorls

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive skin tumor. In addition to the conventional type, several morphologic variants have been described. Recognition of these uncommon variants will facilitate the diagnosis. We report herein a peculiar case of pigmented DFSP (Bednar tumor) with prominent meningotheelial-like whorls, a distinctive pattern that has not been described previously in DFSP. The tumor occurred in a 40-year-old man who presented with a slowly growing mass on his left shoulder. The overall histological features were consistent with Bednar tumor. However, unexpected numerous meningotheelial-like whorls were found in some areas of the tumor. Like the tumor cells in typical areas of Bednar tumor these meningotheelial-like whorls were also positive for CD34 but negative for epithelial membrane antigen and S100 protein. The meningotheelial-like whorls in Bednar tumor represent an eccentric arrangement of the tumor cells. We propose the term ‘pigmented DFSP with prominent meningotheelial-like whorls’ to highlight the distinctive pattern of this novel DFSP variant.


Dermatofibrosarcoma protuberans (DFSP) is a skin tumor of low-grade or intermediate malignancy. DFSP is characterized by a locally aggressive growth and a high rate of recurrence. The typical DFSP is composed of dense uniform spindle cells arranged in a distinct storiform pattern with frequent infiltration into the subcutaneous adipose tissue. In addition to the conventional type, approximately eight uncommon variants have been described. These variants include pigmented DFSP or Bednar tumor,1 myxoid DFSP,2 fibrosarcomatous DFSP (FS-DFSP),3 granular cell DFSP,4 DFSP/FS-DFSP with foci of myoid/myofibroblastic differentiation,5 DFSP with areas of giant cell fibroblastoma,6 an atrophic and plaque-like form of DFSP7 and palisading and Verocay body-prominent DFSP.8 Among these variants, only the FS-DFSP variant has a prognostic significance, whereas the other variants simply represent morphological heterogeneity in DFSP with no distinct relationship to the clinical behavior. Nevertheless, recognition of these rare variants will facilitate the correct diagnosis of DFSP. We report herein a peculiar case of pigmented DFSP (Bednar tumor) with prominent meningotheelial-like whorls. To our knowledge, this distinctive histological pattern has not been described in DFSP.

Case report
A 40-year-old man went to the outpatient clinic in a local hospital because of a slowly growing non-tender mass on his left shoulder. The painless mass had been present for nearly 1 year. His past history was unremarkable. Physical examination revealed a subcutaneous mass, which measured approximately...
4 cm in diameter. A simple local excision was performed. Grossly, the lesion was described as a firm whitish-gray nodule, measuring 3 × 2 cm in size. The lesion was considered initially as DFSP and sent to cancer hospital for further confirmation.

Histological reexamination of the lesion showed typical features of DFSP, which was composed of uniform spindle cells arranged in compact storiform pattern (Fig. 1). Although appearing relatively circumscribed at lower examination, the tumor extended into the deep adipose tissue in a so-called honeycomb fashion. A few entrapped nerve fascicles were also noted at the periphery. In focal areas of the tumor, scattered pigmented dendritic cells were identified, compatible with Bednar tumor (Fig. 2). Unexpectedly, numerous concentric whorls were found in the central portions of tumor bulk, closely mimicking the whorls seen in meningiomas (Fig. 3). However, no psammoma bodies could be noted. There was a gradual transition between these meningothelial-like whorls and the typical storiform areas of the tumor (Fig. 4). The surgical margins of the specimen were positive with tumor cells. A wide local excision was performed later. No adjuvant therapy was administrated. The patient is well now with no evidence of recurrence at 4-month follow up.

Immunohistochemical study was performed on paraffin-embedded tissue sections cut from the

![Fig. 1. A and B) Typical feature of dermatofibrosarcoma protuberans showing uniform spindle cells arranged in a storiform pattern. Note the grenz zone between the tumor and the epidermis.](image)

![Fig. 2. A few pigmented dendritic cells scattered within the tumor.](image)

![Fig. 3. A and B) Numerous meningothelial-like whorls.](image)
submitted blocks. The tumor cells in both the typical areas of DFSP and the meningothelial-like whorls showed diffuse immunoreactivity for CD34 (Fig. 5). They were all negative for epithelial membrane antigen (EMA), cytokeratin (AE1/AE3), α-smooth muscle actin, muscle-specific actin (HHF35), desmin, S-100 protein and HMB45.

Discussion
Histological diagnosis of DFSP is not difficult in most cases of DFSP. However, in some instances, DFSP and its several morphological variants need to be distinguished from a variety of dermal spindle cell tumors, such as dermatofibroma (cutaneous fibrous histiocytoma), neurofibroma (including pigmented variant), myxoid malignant fibrous histiocytoma and rarely schwannoma.8

The histological feature in the present case was compatible with a pigmented DFSP (Bednar tumor), which was initially described by Bednar10 as pigmented storiform neurofibroma. In contrast to the preliminary assumption, Bednar tumor was considered as a pigmented variant of DFSP because the clinical manifestations of Bednar tumor were similar to DFSP and the morphological features were also indistinguishable from DFSP except for scattered non-neoplastic pigmented dendritic cells. In an early study, we also showed that Bednar tumor contained the DFSP-specific COL1A1-PDGFB fusion transcripts, indicating a common histogenesis with ordinary DFSP.11

Bednar tumor is an uncommon entity, accounting for fewer than 5% of all cases of DFSP. Histologically, Bednar tumor is characterized by scattered foci of melanosome-containing dendritic cells within an otherwise typical DFSP. The number of melanin-containing cells varied from case to case. Abundant pigmented dendritic cells can cause black discoloration of the tumor, whereas scant pigmented cells can be only identified microscopically, as shown in this case. Occasionally, Bednar tumor may undertake fibrosarcomatous transformation with rare examples of distant metastasis.12,13 The distinctive feature in this case of Bednar tumor is characterized by the presence of numerous meningothelial-like whorls.

Fig. 4. A and B: There was a gradual transition between the meningothelial-like whorls and the typical dermatofibrosarcoma protuberans area.

Fig. 5. A and B: Immunohistochemical staining of CD34.
which closely resembled a meningioma. In our personal experience with more than 300 cases of DFSP, we have never seen such whorls either in typical cases of DFSP or in its other variants. We initially considered the possibility of so-called palisading and Verocay body-prominent DFSP, which was described by Llatjos et al. However, unlike our case, their cases were characterized by conspicuous nuclear palisading and Verocay body formation. These Verocay bodies and palisades in DFSP implied neural differentiation of the tumor. Zamecnik described another peculiar pattern of so-called giant rosettes in a single case of FS-DFSP. These giant rosettes were found in the fibrosarcomatous areas and were similar to their counterparts in hyalinizing spindle cell tumor with giant rosettes, a variant of low-grade fibromyxoid sarcoma. As indicated by Zamecnik, the presence of giant rosettes in FS-DFSP suggested a possible histogenetic relationship between fibrosarcomatous areas of FS-DFSP and hyalinizing spindle cell tumor with giant rosettes. And again, the giant rosettes in FS-DFSP were different from the meningothelial-like whorls in the current case.

Meningothelial-like whorls in mesenchymal tumors are very rare. According to the literature, meningothelial-like whorls were only found in dedifferentiated liposarcoma and endometrial stromal tumors. The meningothelial-like whorls in dedifferentiated liposarcoma represent a mesenchymal proliferation that may undergo pericytic or myofibroblastic or occasionally osteoblastic differentiation in liposarcoma. On the other hand, the meningothelial-like nodules in endometrial stromal tumors represent an additional pattern of myoid differentiation of the tumor cells. In contrast, the meningothelial-like whorls in our case do not represent a mesenchymal myofibroblastic proliferation or myoid differentiation as they do not express actins or desmin. Neither do these whorls represent perineurial or meningothelial differentiation as they do not express EMA. In line with the gradual transition between the whorls and the typical components of DFSP, and the strong immunoreactivity for CD34, we consider that these meningothelial-like whorls represent a peculiar arrangement of the tumor cells.

The major differential diagnosis includes extracranial meningioma and soft tissue perineurioma. Unlike DFSP, extracranial meningioma usually occurs in the skin of the scalp or along the vertebral axis. Histologically, extracranial meningioma is composed of uniform oval cells arranged in nests or whorls. Although not common, psammoma bodies can be occasionally noticed in some lesions. Immunostaining of EMA may help to distinguish extracranial meningioma from DFSP. Soft tissue perineurioma is a spindle cell neoplasm composed of fibroblast-like cells arranged mostly in short fascicles. However, cellular perineurioma may sometimes form storiform pattern or whorls resembling DFSP. Nevertheless, perineurioma can be segregated from DFSP by its consistent EMA positivity. Another lesion that may enter the differential diagnosis is extranodal follicular dendritic cell sarcoma (FDCS), which can also exhibit whorls. In contrast to DFSP, extranodal FDCS typically involves the head and neck region, gastrointestinal tract and retroperitoneum. FDCS can be identified by immunohistochemistry using FDC markers, such as CD21, CD23, CD35 and clusterin.

In summary, we describe the clinicopathological features of an underrecognized variant of DFSP, which was characterized by prominent meningothelial-like whorls. Based on its unique histology and inspired by the nomination of liposarcoma with meningothelial-like whorls, we propose the term DFSP with prominent meningothelial-like whorls to highlight the distinctive pattern of this novel variant of DFSP.

References

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