A healthy 33-year-old man presented with a depressed, non-tender soft plaque on his back since 1 year. The lesion measured 0.7 mm in diameter and was hyperpigmented [Figure 1]. There was neither history of previous similar lesion, nor any history of antecedent trauma to this site. Upon skin biopsy and histological examination, storiform fascicles of uniform spindle-shaped cells with invasion to the subcutaneous fat tissue were seen [Figure 2]. A diagnosis of malignant spindle cell tumor was made from the history, clinical presentation, and pathological study, and excision of the lesion was advised. An immunohistochemical examination for Ki67, a nuclear antigen that reflected proliferation, showed 10%–15% of cells in mitotic phase [Figure 3].

**Question**

What is Your Diagnosis?

**Figure 2:** Histopathology shows characteristic storiform fascicles of uniform spindle cells) (H and E, ×400)

**Figure 3:** Immunohistochemical study for Ki67 showed 10%–15% proliferation phase in the lesion (+400)
Answer
Dermatofibrosarcoma protuberans (DFSP)

Discussion
Dermatofibrosarcoma protuberans (DFSP), which is known to be the most prevalent dermal sarcoma, is a malignant fibroblastic tumor most frequently arising in middle-aged adults, most commonly found on trunk. Presenting symptoms often include an indolent growth of an ill-defined, painless, indurated plaque, or nodule from the dermis. The color may be flesh-colored, violaceous, reddish, or slightly yellow-brown. In general, tumors at initial stages are only fixed to the overlying skin without involvement of deeper structures. However, infiltration process in more advanced stages can involve deeper structures of the subcutaneous tissue. The median size of the tumor is reported 4 cm, with greater chance of fibrosarcomatous changes in larger lesions.[1] Our case was notable for its unusual appearance of a DFSP lesion: a depressed hyperpigmented nodule and the lack of induration.

Clinical impressions according to presenting morphology of the typical DFSP lesions and pathological studies could be keloids and other neoplasms (because of the overlap of immunoprofile and microscopic morphology), such as dermatofibroma, solitary fibrous tumor, spindle cell/pleomorphic lipomas (SC/PL), and peripheral nerve sheath tumors (e.g., neurofibroma or schwannoma). It is notable that unlike DFSP, all of these differentials lack COL1A1-PDGFB fusion transcripts.

Because of the numerous differential diagnoses for DFSP, the diagnosis usually is made using pathological examination of the excisional or core needle biopsy of the suspicious lesion.

The histopathology studies of lesions show proliferation of uniform spindle tumor cells arranged in storiform fascicles with possible involvement of subcutaneous fat or of skeletal muscle with a characteristic "honeycomb" pattern, possessing little cytologic atypia and low mitotic activity (median <4 mitoses per 10 high power fields). However, mitotic rate is usually higher in areas of fibrosarcomatous change. Intercellular collagen deposition and diffuse vasculature may be present.[1,2] Spindle shaped cells are characteristically stained positively for CD34 and apolipoprotein D. The expression of CD99 and SMA (smooth muscle actin) is variable among lesions.

DFSP has morphological variants that share similar clinical, histological, and immunohistochemical features. Myxoid DFSP (Bednar tumor), giant cell fibroblastoma, fibrosarcomatous DFSP, granular cell DFSP, and atrophic and palisading variants are among them.[2]

Surgical treatment (classic wide excision or Mohs micrographic surgery) remains the most effective therapy for DFSP.

The targeted molecular therapy of DFSP by the PDGF receptor-selective oral tyrosine kinase-inhibitor (imatinib) and radiation therapy are substitutes of surgical resection for primary inoperable, recurrent, retractable, or metastatic tumors.[3]

The prognosis of primary local DFSP is generally very good, if adequate free margins are obtained during surgical excision.

Learning Points
• DFSP is rare but is the most prevalent dermal sarcoma
• Most frequently, it is reported in middle-aged adults. The most common affected location is trunk, followed by extremities, head, and neck. Presenting symptoms often include indolent growth of an ill-defined, painless, indurated plaque, or nodule. However, it may unusually present as a soft depressed hyperpigmented nodule as in our case
• Histopathologically, it is characterized by proliferation of uniform spindle tumor cells arranged in storiform fascicles
• It is often misdiagnosed as keloids and other neoplasms such as dermatofibroma, solitary fibrous tumor, and peripheral nerve sheath tumors
• The prognosis with wide excision of the lesion is excellent with minimal chance of recurrence.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal.

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Conflicts of interest
There are no conflicts of interest.

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