Dermatofibrosarcoma Protuberans

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A 26-year-old woman with no prenatal care presented to the labor and delivery department in active labor at 38.4-weeks' gestation. A large multinodular abdominal mass suspicious for central necrosis and infected tissue was noted along her right lower quadrant. After successful vaginal delivery, computed tomography of the abdomen and pelvis demonstrated an enhancing multilobulated mass involving the skin and soft tissue, measuring 10.0 cm by 5.4 cm (**image A**, arrows). One week postpartum, the mass was surgically excised via wide local excision.

The histopathologic examination revealed a spindle-cell neoplasm showing a storiform type arrangement of the spindle cells (**image B**, circle; hematoxylin-eosin, original magnification × 100), with mild nuclear pleomorphism, invasion of the subcutaneous fat, and positive immunohistochemistry for vimentin and CD34 (not shown). These findings revealed a rare soft tissue sarcoma known as dermatofibrosarcoma protuberans, which has an estimated incidence of 0.8 to 4.5 cases per million per year.[1] These tumors comprise roughly 6% of all soft tissue sarcomas.[2] They are superficial, slow-growing, and have a low malignant potential, with roughly 85% to 90% being low grade with a relatively good prognosis.[3] Dermatofibrosarcoma protuberans has a high frequency of local regional recurrence, with rates ranging from 26% to 60%.[4]

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