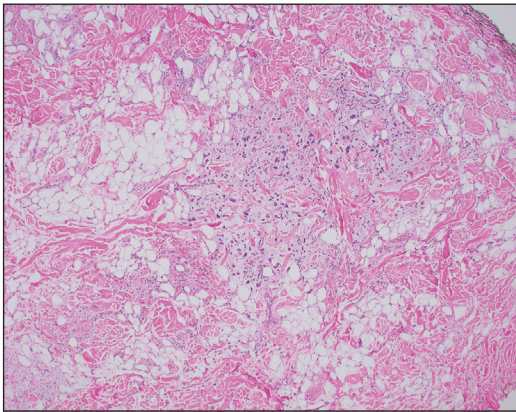


Subcutaneous Nodule on the Postauricular Neck

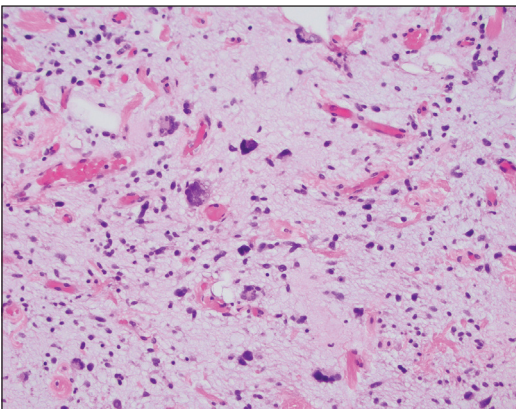
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H&E, original magnification $\times 100$.



H&E, original magnification $\times 200$.

An otherwise healthy 56-year-old man with a family history of lymphoma presented with a raised lesion on the postauricular neck. He first noticed the nodule 3 months prior and was unsure if it was still getting larger. It was predominantly asymptomatic. Physical examination revealed a 1.5 \times 1.5-cm, mobile, subcutaneous nodule. An incisional biopsy was performed and submitted for histologic evaluation.

THE BEST DIAGNOSIS IS:

- angiomyxoma
- dermatofibrosarcoma protuberans
- multinucleate cell angiohistiocytoma
- nodular fasciitis
- pleomorphic lipoma

PLEASE TURN TO **PAGE 239** FOR THE DIAGNOSIS

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The authors report no conflict of interest.

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THE DIAGNOSIS: Pleomorphic Lipoma

Pleomorphic lipoma is a rare, benign, adipocytic neoplasm that presents in the subcutaneous tissues of the upper shoulder, back, or neck. It predominantly affects men aged 50 to 70 years. Most lesions are situated in the subcutaneous tissues; few cases of intramuscular and retroperitoneal tumors have been reported.¹ Clinically, pleomorphic lipomas present as painless, well-circumscribed lesions of the subcutaneous tissue that often resemble a lipoma or occasionally may be mistaken for liposarcoma. Histopathologic examination of ordinary lipomas reveals uniform mature adipocytes. However, pleomorphic lipomas consist of a mixture of multinucleated floretlike giant cells, variable-sized adipocytes, and fibrous tissue (ropy collagen bundles) with some myxoid and spindled areas.^{1,2} The most characteristic histologic feature of pleomorphic lipoma is multinucleated floretlike giant cells. The nuclei of these giant cells appear hyperchromatic, enlarged, and disposed to the periphery of the cell in a circular pattern. Additionally, tumors frequently contain excess mature dense collagen bundles that are strongly refractile in polarized light. Numerous mast cells are present. Atypical lipoblasts and capillary networks commonly are not visible in pleomorphic lipoma.³ The spindle cells express CD34 on immunohistochemistry. Loss of Rb-1 expression is typical.⁴

Dermatofibrosarcoma protuberans is a slow-growing soft tissue sarcoma that commonly begins as a pink or violet plaque on the trunk or upper limbs. Involvement of the head or neck accounts for only 10% to 15% of cases.⁵ This tumor has low metastatic potential but is highly infiltrative of surrounding tissues. It is associated with a translocation between chromosomes 22 and 17, leading to the fusion of the platelet-derived growth factor subunit β , *PDGFB*, and collagen type 1 α 1, *COL1A1*, genes.⁵ Clinically, patients often report that the lesion was present for several years prior to presentation with general stability in size and shape. Eventually, untreated lesions progress to become nodules or tumors and may even bleed or ulcerate. Histology reveals a storiform spindle cell proliferation throughout the dermis with infiltration into subcutaneous fat, commonly appearing in a honeycomblike pattern (Figure 1). Numerous histologic variants exist, including myxoid, sclerosing, pigmented (Bednar tumor), myoid, atrophic, or fibrosarcomatous dermatofibrosarcoma protuberans, as well as a giant cell fibroblastoma variant.⁶ These tumor subtypes can exist independently or in association with one another, creating hybrid lesions that can closely mimic other entities such as pleomorphic lipoma. The spindle cells stain positively for CD34. Treatment of these tumors involves complete surgical excision or Mohs micrographic surgery;

however, recurrence is common for tumors involving the head or neck.⁵

Superficial angiomyxoma is a slow-growing papule that most commonly appears on the trunk, head, or neck in middle-aged adults. Occasionally, patients with Carney complex also can develop lesions on the external ear or breast.⁷ Histologically, superficial angiomyxoma is a hypocellular tumor characterized by abundant myxoid stroma, thin blood vessels, and small spindled and stellate cells with minimal cytoplasm (Figure 2).⁸ Superficial angiomyxoma and pleomorphic lipoma present differently on histology; superficial angiomyxoma is not associated with nuclear atypia or pleomorphism, whereas pleomorphic lipoma characteristically contains multinucleated floretlike giant cells and pleomorphism. Frequently, there

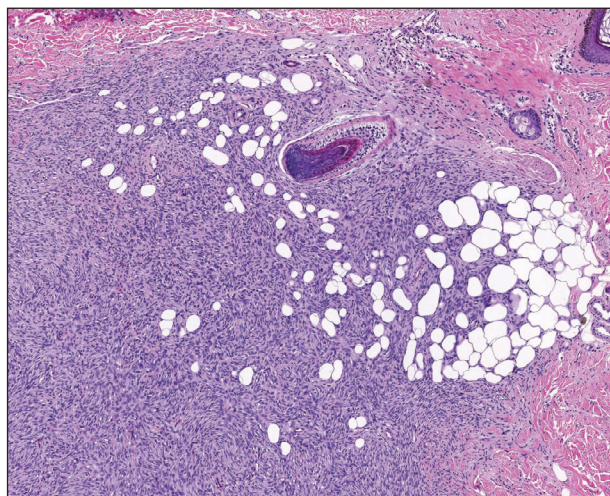


FIGURE 1. Dermatofibrosarcoma protuberans. Sheets of spindle cells are arranged in a storiform pattern proliferating within the dermis and infiltrating the fat in a honeycomblike pattern (H&E, original magnification $\times 100$).

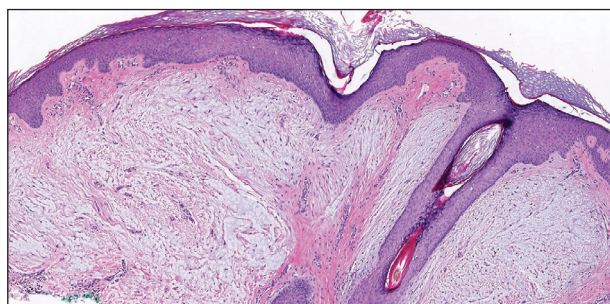


FIGURE 2. Superficial angiomyxoma. Abundant myxoid stroma and small spindled and stellate cells are present with minimal cytoplasm and surrounding thin blood vessels (H&E, original magnification $\times 40$).

also is loss of normal *PRKAR1A* gene expression, which is responsible for protein kinase A regulatory subunit 1- α expression.⁸

Multinucleate cell angiohistiocytoma is a rare benign proliferation that presents with numerous red-violet asymptomatic papules that commonly appear on the upper and lower extremities of women aged 40 to 70 years. Lesions feature both a fibrohistiocytic and vascular component.⁹ Histologic examination commonly shows multinucleated cells with angular outlining in the superficial dermis accompanied by fibrosis and ectatic small-caliber vessels (Figure 3). Although both pleomorphic lipoma and multinucleate cell angiohistiocytoma have similar-appearing multinucleated giant cells, the latter has a proliferation of narrow vessels in thick collagen bundles and lacks an adipocytic component, which distinguishes it from the former.¹⁰ Multinucleate cell angiohistiocytoma also is characterized by a substantial number of factor XIIIa–positive fibrohistiocytic interstitial cells and vascular hyperplasia.⁹

Nodular fasciitis is a benign lesion involving the rapid proliferation of myofibroblasts and fibroblasts in the subcutaneous tissue and most commonly is encountered on the extremities or head and neck regions. Many cases appear at sites of prior trauma, especially in patients aged 20 to 40 years. However, in infants and children the lesions typically are found in the head and neck regions.¹¹ Clinically, lesions present as subcutaneous nodules. Histology reveals an infiltrative and poorly circumscribed proliferation of spindled myofibroblasts associated with myxoid stroma and dense collagen depositions. The spindled cells are loosely associated, rendering a tissue culture–like appearance (Figure 4). It also is common to see erythrocyte extravasation adjacent to myxoid stroma.¹¹ Positive stains include vimentin, smooth muscle actin,

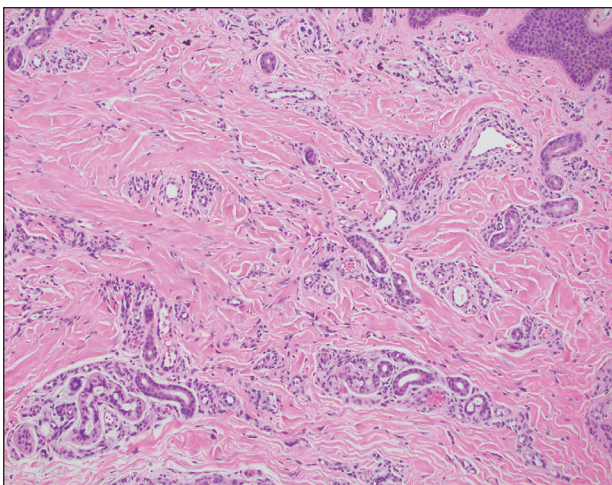


FIGURE 3. Multinucleate cell angiohistiocytoma. Fibrosis of the superficial dermis is present with scattered small-caliber vessels and fibrohistiocytic cells (H&E, original magnification $\times 40$).

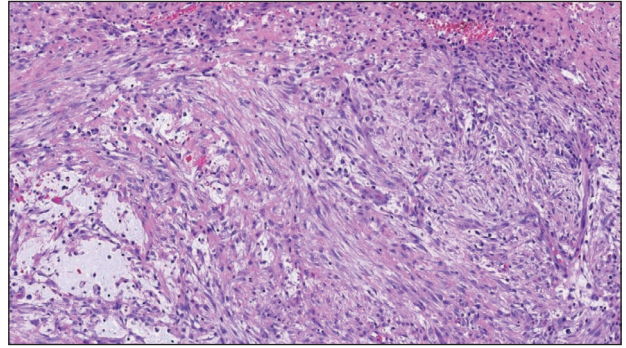


FIGURE 4. Nodular fasciitis. Slender, loosely aggregated, spindled cells with a tissue culture–like appearance can be seen (H&E, original magnification $\times 200$).

and CD68, though immunohistochemistry often is not necessary for diagnosis.¹² There often is abundant mitotic activity in nodular fasciitis, especially in early lesions, and the differential diagnosis includes sarcoma. Although nodular fasciitis is mitotically active, it does not show atypical mitotic figures. Nodular fasciitis commonly harbors a gene translocation of the *MYH9* gene's promoter region to the *USP6* gene's coding region.¹³

REFERENCES

1. Sakhadeo U, Mundhe R, DeSouza MA, et al. Pleomorphic lipoma: a gentle giant of pathology. *J Cytol*. 2015;32:201-203. doi:10.4103/0970-9371.168904
2. Shmookler BM, Enzinger FM. Pleomorphic lipoma: a benign tumor simulating liposarcoma. a clinicopathologic analysis of 48 cases. *Cancer*. 1981;47:126-133.
3. Azzopardi JG, Iocco J, Salm R. Pleomorphic lipoma: a tumour simulating liposarcoma. *Histopathology*. 1983;7:511-523. doi:10.1111/j.1365-2559.1983.tb02264.x
4. Jäger M, Winkelmann R, Eichler K, et al. Pleomorphic lipoma. *J Dtsch Dermatol Ges*. 2018;16:208-210. doi:10.1111/ddg.13422
5. Allen A, Ahn C, Sangüeza OP. Dermatofibrosarcoma protuberans. *Dermatol Clin*. 2019;37:483-488. doi:10.1016/j.det.2019.05.006
6. Socoliuc C, Zurac S, Andrei R, et al. Multiple histological subtypes of dermatofibrosarcoma protuberans occurring in the same tumor. *Rom J Intern Med*. 2015;53:79-88. doi:10.1515/rjim-2015-0011
7. Abarzúa-Araya A, Lallas A, Piana S, et al. Superficial angiomyxoma of the skin. *Dermatol Pract Concept*. 2016;6:47-49. doi:10.5826/dpc.0603a09
8. Hornick J. *Practical Soft Tissue Pathology A Diagnostic Approach*. 2nd ed. Elsevier Health Sciences; 2017.
9. Rato M, Monteiro AF, Parente J, et al. Case for diagnosis. multinucleated cell angiohistiocytoma. *An Bras Dermatol*. 2018;93:291-293. doi:10.1590/abd1806-4841.20186821
10. Grgurich E, Quinn K, Oram C, et al. Multinucleate cell angiohistiocytoma: case report and literature review. *J Cutan Pathol*. 2019;46:59-61. doi:10.1111/cup.13361
11. Zuber TJ, Finley JL. Nodular fasciitis. *South Med J*. 1994;87:842-844. doi:10.1097/00007611-199408000-00020
12. Yver CM, Husson MA, Friedman O. Pathology clinic: nodular fasciitis involving the external ear [published online March 18, 2021]. *Ear Nose Throat J*. doi:10.1177/01455613211001958
13. Erickson-Johnson M, Chou M, Evers B, et al. Nodular fasciitis: a novel model of transient neoplasia induced by *MYH9-USP6* gene fusion. *Lab Invest*. 2011;91:1427-1433. https://doi.org/10.1038/labinvest.2011.118