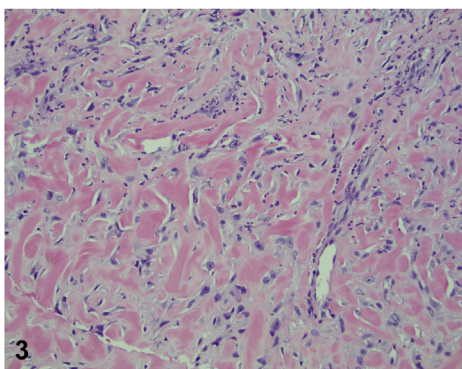
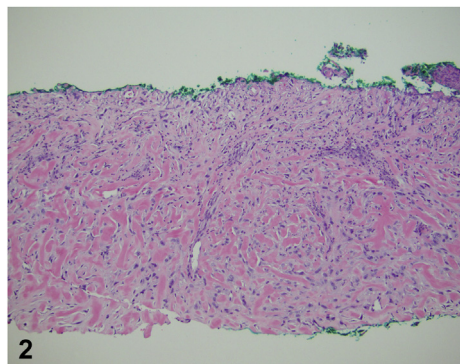


7-mm crusted papule on an elderly man's ear



Peter A. Young, MPAS,^a Nupoor A. Gajjar, MD,^b and Robert L. Burns, MD^c
Sacramento, Oakland, and Rancho Cordova, California

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PRESENTATION

A 73-year-old man presented for Mohs micrographic surgery with a biopsy-confirmed basal cell carcinoma on his right ear and requested the additional examination of a skin lesion on his left ear. The examination showed a 7-mm, thin, crusted papule on the midhelix (Fig 1), and a shave biopsy was performed to evaluate for basal and squamous cell carcinoma. Pathology showed a tumor composed predominantly of spindle-shaped cells with pleomorphic nuclei and multiple mitoses, with positivity to CD10 immunoperoxidase, transected at the base (Figs 2 and 3 [hematoxylin-eosin, original magnifications $\times 100$ and $\times 200$, respectively]).

Question 1: What is the most likely diagnosis?

A. Desmoplastic melanoma

B. Atypical fibroxanthoma

C. Morpheic basal cell carcinoma

From the Department of Dermatology, The Permanente Medical Group, Sacramento^a; Department of Dermatopathology, The Permanente Medical Group, Oakland^b; and Department of Mohs Micrographic Surgery, The Permanente Medical Group, Rancho Cordova.^c

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Correspondence to: Peter A. Young, MPAS, Department of Dermatology, The Permanente Medical Group, 2345 Fair Oaks Blvd, Sacramento, CA 95825. E-mail: peter.a.young@kp.org.

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- D. Clear cell sarcoma
- E. Squamous cell carcinoma

Answers:

- A. Desmoplastic melanoma — Incorrect. Histology for this would show strands of elongated, spindle-shaped cells surrounded by mature collagen bundles. The cells resemble fibroblasts, some with hyperchromatic and bizarre nuclei, and are typically CD10⁻.¹
- B. Atypical fibroxanthoma — Correct. Pathology showed that this tumor was composed predominantly of spindle-shaped cells with pleomorphic nuclei and multiple mitoses, characteristic of atypical fibroxanthoma. This is a rare, malignant, cutaneous neoplasm that exists along a clinicopathologic spectrum with pleomorphic dermal sarcoma (also known as undifferentiated pleomorphic sarcoma). Although these tumors share many similarities, the recognition of distinguishing characteristics may predict differences in clinical behavior and outcomes. A pleomorphic dermal sarcoma is often larger than an atypical fibroxanthoma and is more likely to recur or metastasize. An atypical fibroxanthoma, by definition, is confined to the dermis, whereas a pleomorphic dermal sarcoma involves the subcutis, fascia, lymphatics, or vascular structures.^{2,3} Because the lesion was transected at the base, pleomorphic dermal sarcoma cannot be ruled out with this biopsy alone.
- C. Morpheic basal cell carcinoma — Incorrect. Histology for this would show narrow 50 elongated strands and small islands of basophilic tumor cells embedded in a dense, fibrous stroma.¹
- D. Clear cell sarcoma — Incorrect. These tumors (“melanomas of soft parts”) are composed of nests and strands of oval or elongated cells separated by collagenous septa, with copious pale or granular amphophilic cytoplasm.¹
- E. Squamous cell carcinoma — Incorrect. Histopathology for this tumor would show nests of squamous epithelial cells, with abundant eosinophilic cytoplasm and large vesicular nuclei, arising from the epidermis and extending into the dermis.¹

Question 2: What is the appropriate next step?

- A. Reassurance
- B. Cryotherapy
- C. Repeat shave biopsy

- D. Mohs micrographic surgery
- E. Topical imiquimod

Answers:

- A. Reassurance — Incorrect. Reassurance is not appropriate for atypical fibroxanthoma or pleomorphic dermal sarcoma.
- B. Cryotherapy — Incorrect. Cryotherapy is not effective for either entity on the differential diagnosis.
- C. Repeat shave biopsy — Incorrect. Although this may confirm the diagnosis, both entities on the differential diagnosis would undergo Mohs micrographic surgery. A repeat biopsy would only delay the treatment without any benefit.
- D. Mohs micrographic surgery — Correct. The lesion was transected at the base, precluding the preoperative differentiation of atypical fibroxanthoma from pleomorphic dermal sarcoma. The treatment of choice for both is Mohs micrographic surgery.^{2,3}
- E. Topical imiquimod — Incorrect. This is not effective for either entity on the differential diagnosis.

Question 3: Mohs micrographic surgery was performed, and intraoperative frozen sections revealed that the tumor was confined to the dermis. Which of the following is true?

- A. Recurrence rates are as high as 90%
- B. Metastases are common
- C. When metastases occur, they are usually to the parotid gland, lymph nodes, or subcutaneous tissue
- D. The patient should be screened every 4 months
- E. Atypical fibroxanthoma stains negative for p53, S100A6, vimentin, and procollagen 1

Answers:

- A. Recurrence rates are as high as 90% — Incorrect. Following Mohs micrographic surgery, recurrence rates for atypical fibroxanthoma are 0% to 6.9%.
- B. Metastases are common — Incorrect. Metastases are rare but usually occur 1 to 2 years after the initial diagnosis of atypical fibroxanthoma.
- C. When metastases occur, they are usually to the parotid gland, lymph nodes, or subcutaneous tissue — Correct.
- D. The patient should be screened every 4 months — Incorrect. Patients with histories of atypical fibroxanthoma should be screened annually and counseled to limit ultraviolet exposure.

E. Atypical fibroxanthoma stains negative for p53, S100A6, vimentin, and procollagen 1 – Incorrect. Atypical fibroxanthoma is considered a diagnosis of exclusion histologically, in part because it stains positive for several nonspecific stains, including CD10, p53, S100A6, vimentin, and procollagen 1.²

Conflicts of interest

None disclosed

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