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Photo Quiz

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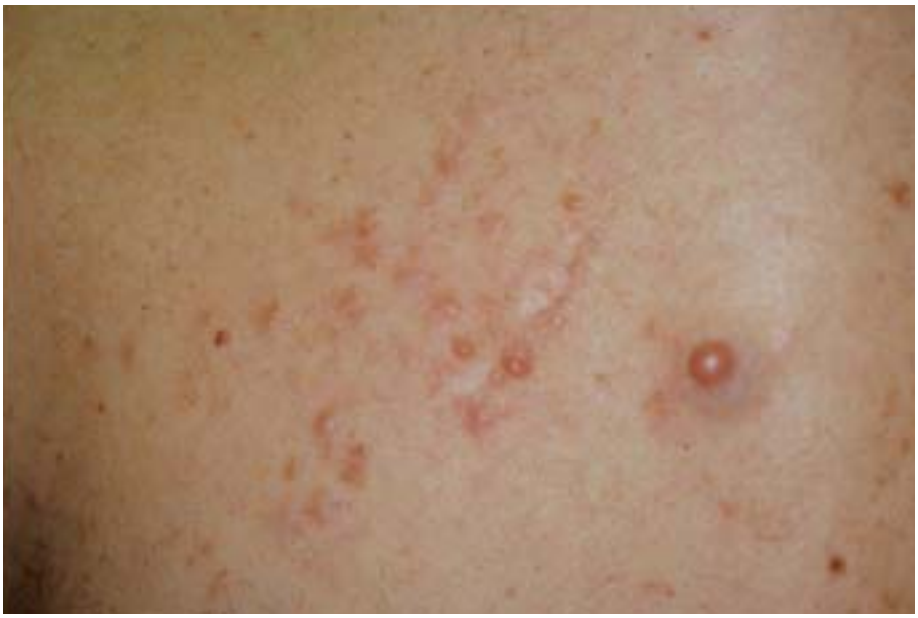

A 39-year-old man presented with a large asymptomatic plaque on his left upper back, which had been previously excised but recurred shortly thereafter and continued to enlarge. On physical examination, there were numerous 2-mm to 1.5-cm, reddish-brown, firm, dermal papules and nodules forming a large 6-cm plaque on his left scapular area.

What is your diagnosis?

PLEASE TURN TO PAGE 127 FOR DISCUSSION

The Diagnosis: Piloleiomyomas

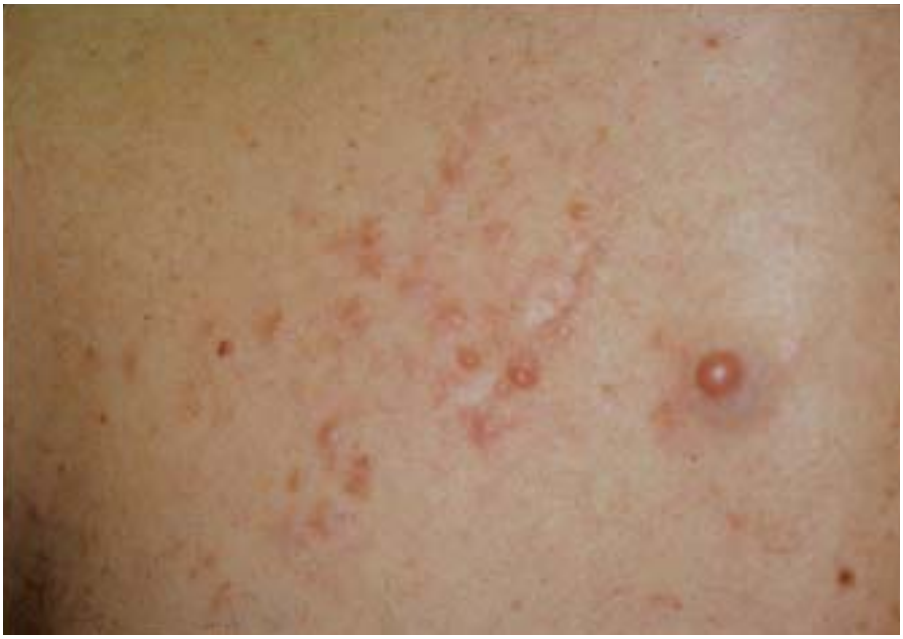

Figure 1. Numerous 2-mm to 1.5-cm papules and nodules forming a large 6-cm plaque on the patient's left scapular area.

Piloleiomyomas are benign smooth muscle tumors derived from the arrector pili muscles. These slow-growing tumors are seen primarily in young adults (male-female ratio of 1:1) and are most often located on the extensor surfaces of the limbs, trunk, and sides of the face and neck. The skin overlying the tumors is reddish-brown. Tumors may be solitary, but multiple tumors are more common. Solitary tumors may be several centimeters in size. As seen in the case presented, multiple lesions usually are smaller and often coalesce to form large plaques (Figure 1). The lesions are fixed to the skin but are freely movable and may be spontaneously painful and sensitive to touch or cold.[1,2] These lesions usually are distinctive, but the clinical differential diagnosis can include dermatofibroma, xanthogranuloma, steatocystoma, and carcinoma (either primary or metastatic).

Histologically, leiomyomas are composed of bundles of haphazardly arranged, poorly circumscribed, bland-appearing, spindle-shaped cells with oval or cigar-shaped nuclei and abundant eosinophilic cytoplasm.[1] On cross section, the smooth muscle bundles demonstrate slight vacuolation, which is the result of the perinuclear clear zone[3] (Figure 2). There are 2 other subtypes of leiomyomas that are similar histologically to piloleiomyomas: angioleiomyomas (derived from vascular smooth muscle) and dartoic leiomyomas (derived from the dartos muscle of the genitalia, areola, or nipple).[4]

The histologic differential diagnosis of piloleiomyomas includes neurofibroma, neurilemoma, and leiomyosarcoma, which can be differentiated with special stains when necessary. Smooth muscle can be stained with either phosphotungstic acid or Masson trichrome stain, which turn muscle either purple or pinkish-red, respectively. The leiomyomas express both smooth muscle actin and desmin, which highlight the muscle bundles.[1,2] Neurofibromas differ from piloleiomyomas in that they do not have an eosinophilic cytoplasm and, unlike leiomyomas, both neurofibromas and neurilemomas stain positive with S-100 protein.

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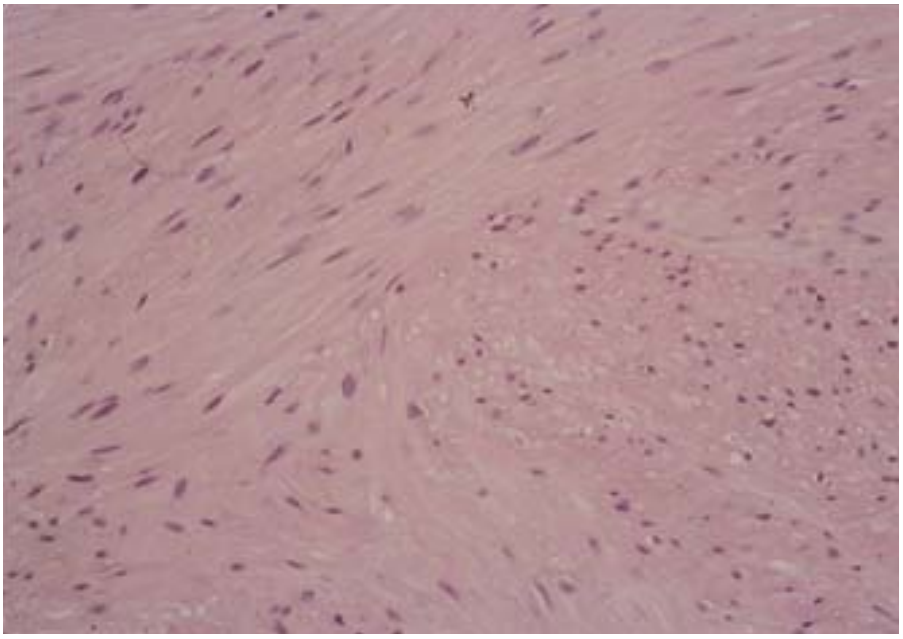

Figure 2. Smooth muscle bundles demonstrating slight vacuolation (H&E, original magnification ×40).

Histologically, leiomyosarcomas have many more mitoses and demonstrate nuclear pleomorphism.

Although piloleiomyomas are benign tumors, patients may seek treatment for either pain relief or aesthetic reasons. The treatment of choice for piloleiomyomas is excision. However, excised lesions often recur, and excision of an entire tumor may be problematic for cosmetic reasons because of size or location.[2,4] Pain from these tumors may be significant and debilitating. Effective pain relief treatments include nitroglycerin, phenoxybenzamine, and nifedipine.[5,6]

Although most cases of cutaneous piloleiomyomas are sporadic, there are reports of patients with multiple tumors who have a family history of leiomyomas consistent with autosomal dominant inheritance.[7] Women with cutaneous tumors also may develop uterine smooth muscle tumors, Reed's syndrome, or familial leiomyomatosis cutis et uteria.[8] There are isolated case reports of cutaneous piloleiomyomas being associated with polycythemia, and erythropoietic activity has been demonstrated in some tumor extracts.[9] There is one case report of a set of twins with multiple cutaneous leiomyomas with chromosomal translocation (9p trisomy/18p distal monosomy).[10] Gardner's syndrome (gastrointestinal polyposis, osteomas, fibromas, lipomas, and epidermal inclusion cysts) also has been associated with multiple leiomyomas.[7]

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