The best diagnosis is:

a. chondroma
b. mixed tumor (chondroid syringoma)
c. nerve sheath myxoma (neurothekeoma)
d. nodular fasciitis
e. solitary adult myofibroma
Myofibromatosis can be classified based on its clinical presentation as either solitary infantile myofibroma, congenital multiple myofibromatosis without visceral involvement, congenital generalized myofibromatosis with visceral involvement, or solitary adult myofibroma. Solitary adult myofibroma is a benign proliferation of myofibroblasts that most commonly presents as a firm superficial nodule on the skin or oral mucosa of young to middle-aged adults. In contrast, congenital generalized myofibromatosis shows a female predominance and typically presents at or near birth as multicentric soft tissue tumors with associated bone, oral, and potentially fatal visceral involvement. Histologically, solitary adult myofibroma and the infantile myofibromatoses are nearly identical. A well-circumscribed, often multinodular nodule is seen in the dermis or subcutaneous tissue with a characteristic biphasic pattern consisting of bland plump myofibroblasts and smaller, spindle-shaped mesenchymal cells (Figure 1). The spindle-shaped cells tend to be arranged around branching blood vessels that resemble hemangiopericytoma, also known as staghorn vessels, which may be seen in the center or periphery of the tumor (Figure 2). Unlike the infantile myofibromatoses, the adult variant tends to undergo hyalinization, resulting in a pseudochondroid appearance.

The histologic differential diagnosis may include other multilobular dermal or subcutaneous tumors with a mucinous or pseudochondroid stroma. The architecture and stroma of mixed tumor (chondroid syringoma) may simulate myofibroma from low-power magnification, but it can be distinguished by the presence of tubuloalveolar and ductal structures within the chondroid stroma (Figure 3). Nerve sheath myxoma (neurothekeoma) displays myxoid lobules containing spindle-shaped, stellate, or epithelioid cells (Figure 4), and the lobules are separated by fibrous septa as opposed to the hemangiopericytomalike vessels of myofibroma. Nodular fasciitis should be included in the differential diagnosis of myofibroma, as both are unencapsulated nodules with variable amounts of myxoid stroma.

**Figure 1.** Subcutaneous tumor composed of nodular aggregates of spindle-shaped cells with central pseudochondroid stroma and irregular vascular spaces peripherally (H&E, original magnification ×40).

**Figure 2.** Biphasic tumor of plump myofibroblasts within a pseudochondroid stroma and small mesenchymal cells around peripheral vessels resembling hemangiopericytoma (H&E, original magnification ×100).

**Figure 3.** Small nonbranching ducts resembling eccrine syringoma embedded within a myxoid and chondroid stroma of mixed tumor (H&E, original magnification ×100).
and thin-walled blood vessels. In contrast to myofibroma, nodular fasciitis is composed of plump, spindle-shaped cells arranged in a haphazard array (tissue culture appearance) with extravasated red blood cells and scattered lymphocytes (Figure 5). Chondroma is an encapsulated tumor of mature cartilage and can be distinguished from myofibroma by the presence of chondrocytes occupying individual lacunae (Figure 6).

**REFERENCES**