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Fibromatosis

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Abstract
Fibromatosis represents a diverse group of fibroproliferative tumours. Their behaviour and pathological qualities are situated in an intermediate position between benign and malignant disease. The following represents the pathological and radiological presentation of a 29-year-old female with fibromatosis.

Key words: Fibromatosis; Immunohistochemistry

Case report

The patient is a 29-year-old white female without a significant past medical history, who had symptom onset following a motor vehicle accident one year prior to presentation. At that point, the patient began complaining of minor pain in the right shoulder and trapezius areas, which seemed to increase in intensity over the next several months. She subsequently developed progressive weakness of the right arm. At the time of first presentation physical examination revealed a fullness in the right anterior neck and supraclavicular regions. The remainder of the head and neck examination was unremarkable. A thorough neurological exam showed reflex diminution in the right supinator and right biceps, with the triceps maintained. Wasting of the right deltoid and supraspinatus muscles was noted, with weakness of arm flexion and external rotation, and concomitant loss of pinprick laterally in the lateral three fingers. The remainder of the neurological exam was normal.

Computed tomography (CT) with contrast showed a 4×3.5 cm mass posterior to the sternocleidomastoid muscle, with some extension posteriorly to the trachea and anterior margin of the cervical vertebrae, with resultant thyroid displacement. Magnetic resonance imaging (MRI) with gadolinium (Figure 1) showed a right-sided lesion in the prevertebral space displacing the jugular and carotid vessels anteriorly and touching the vertebral bodies without bony destruction. The larynx was displaced leftward without luminal involvement. The patient was taken to the operating room for a neck exploration. The tumour was located and found to be abutting the brachial plexus and attached to the paraspinal musculature. The tumour was subsequently debulked in a near total fashion. Grossly, fragments of lesional tissue had a solid and firm, beige to grey glistening cut surface with trabeculation but absence of necrosis or haemorrhage. The interface with normal soft tissue was ill-defined. Microscopic examination revealed long sweeping fascicles of spindle cells within a collagen-rich stroma (Figure 2). Some variation in cellularity was present, including hypocellular zones with a hyalinized, sclerotic stroma or occasional myxoid areas with relatively less amounts of collagen. Nuclei had an ovoid or serpentine shape with a minute nucleolus, without atypia (Figure 2). Rare (non-atypical) mitotic figures were present. There was a discrete capillary vascular framework, with few microhaemorrhages, but without haemosiderin deposition. Immunohistochemical

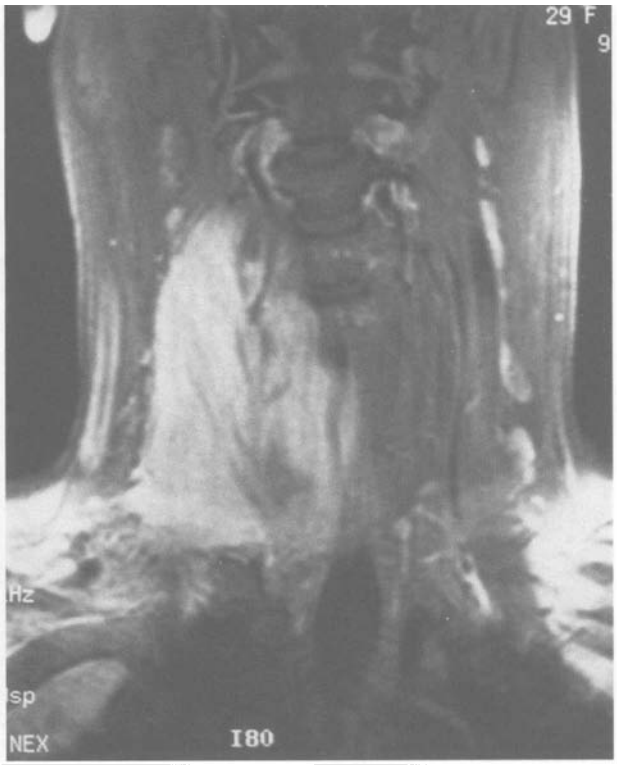

Fig. 1
T1-weighted MRI with gadolinium showing a 3×2 cm enhancing mass involving the right prevertebral space, extending from the inferior border of the C2 vertebra to the inferior border of the T1 vertebra.

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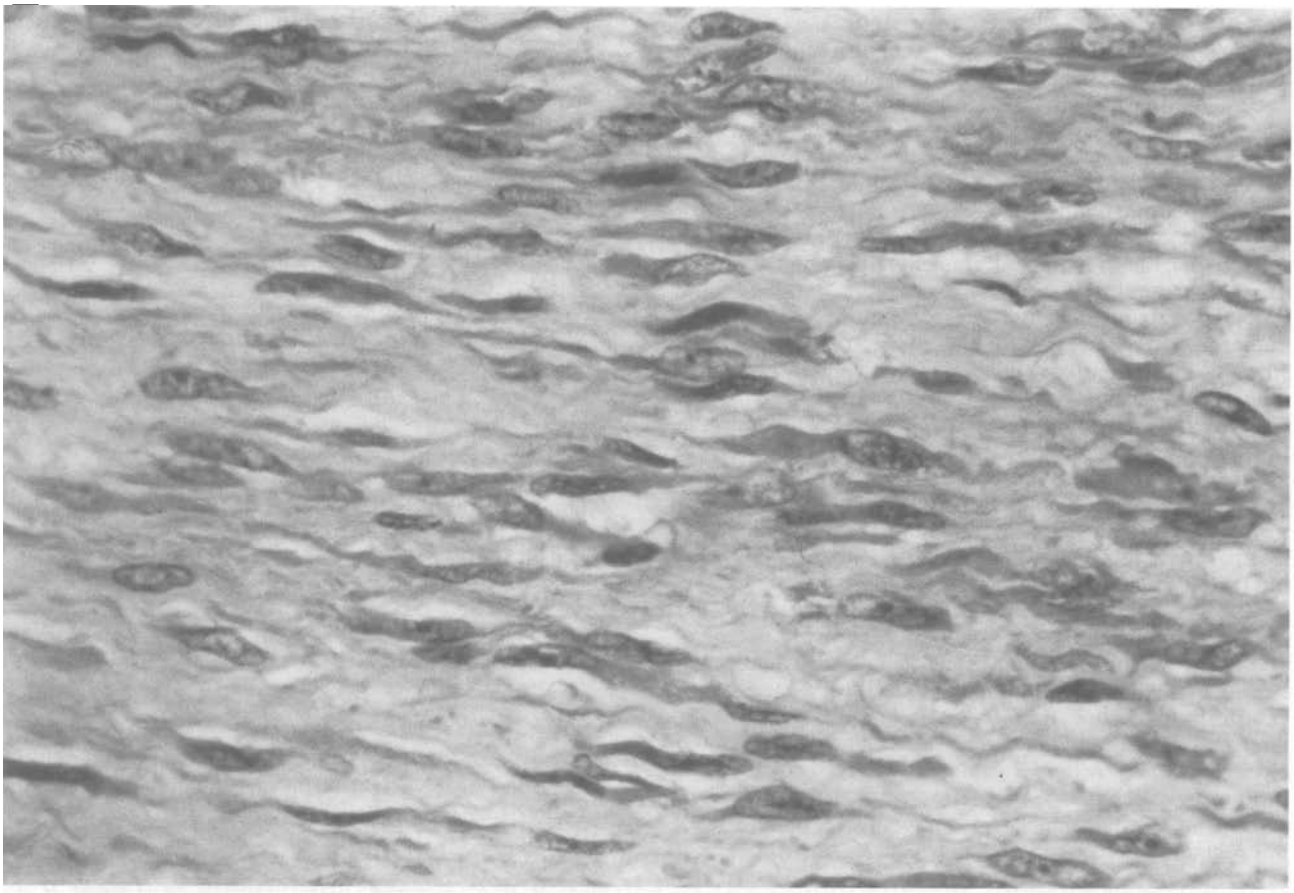

FIG. 2
Fascicle of spindle-shaped fibroblasts with parallel arrangement, and with abundant intercellular collagen. Note elongated nuclei with tapered extremities, and minute nucleoli without atypia (H & E; × 250).

staining revealed a strong cytoplasmic expression of vimentin but negativity for S-100 protein, epithelial membrane antigen, cytokeratin AE1/AE3, muscle-specific actin, desmin, CD34, and oestrogen/progesterone nuclear receptor protein. The pathological diagnosis was consistent with fibromatosis.

Discussion

The fibromatoses, also known as extra-abdominal desmoid and musculoaponeurotic desmoid, represent a heterogeneous group of fibroproliferative tumours situated in an intermediate histological and clinical spectrum between benign and malignant endpoints (Allen, 1997). Eleven to 15 per cent of all cases of fibromatosis occur in the head and neck (Fasching *et al.*, 1988; Plaat *et al.*, 1995), with an incidence of four cases per year per million population (Reitamo *et al.*, 1982). It is most common between the onset of puberty and 40 years of age (Enzinger and Weiss, 1995) and affects females to males in a ratio of 3:2 (Barnes, 1985; Plaat *et al.*, 1995). The aetiology of fibromatosis is not certain, but genetic mutation, heredity, alteration of sex hormone modulation, and radiation have been indicated (Hayry *et al.*, 1982; Enzinger and Weiss, 1995; Plaat *et al.*, 1995). As well, both gross trauma and microtrauma from frequent physical activity have been cited as possible causes (Hunt *et al.*, 1960; Enzinger and Shiraki, 1967; Fasching *et al.*, 1988; Enzinger and Weiss, 1995).

Clinically, the fibromatoses present as firm, poorly circumscribed gradually enlarging painless masses. Other clinical symptoms, such as dysphagia, stridor, facial pain, epistaxis, neuralgias, proptosis, and arm swelling may be related to involvement of contiguous structures (Fu and Perzin, 1976; Wolf *et al.*, 1989; Enzinger and Weiss, 1995; Plaat *et al.*, 1995). They most often occur in the supraclavicular and anterolateral regions of the neck (Barnes, 1985). Its often aggressive behaviour involving deep vital structures lends itself to being confused with malignant fibrous lesions, and differentiates it from more benign reactive states. Multicentric lesions can occur (Goellner and Soule, 1980). This entity's inability to metastasize differentiates it from malignant lesions.

The diagnosis of fibromatosis is based on needle or excisional biopsy, with histological confirmation. CT scan and MRI, though having utility in cases of recurrence and pre-operative margin assessment, may not be useful in primary case diagnosis because of similar surrounding tissue densities. On gross pathologic examination, the tumours are between 1 and 10 cm, have a firm consistency and a trabeculated cut surface (Barnes, 1985; Enzinger and Weiss, 1995). Microscopic examination reveals a poorly circumscribed mass, infiltrating the surrounding tissues, particularly skeletal muscle. The tumour itself consists of homogeneous elongated and slender spindle-shaped cells separated by vast amounts of collagen (Enzinger and Weiss, 1995). The nuclei are uniform and mitoses are sparse. The moderate cellularity, sparse mitotic activity and nuclear uniformity help distinguish fibromatosis from

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poorly differentiated fibrosarcoma, but may allow for confusion with well-differentiated tumours. The presence of myofibroblasts together with typical fibroblasts has been documented in fibromatosis (Gabbiani and Majno, 1972). The myofibroblast is a special type of fibroblast characterized by cytoplasmic extensions, segments of external lamina, well-developed rough endoplasmic reticulum and intracytoplasmic bundles of actin-type microfilaments with associated areas of condensation (dense bodies). Such cells, under the influence of vasoactive prostaglandin, are probably responsible for the contractile nature of the lesion (Badalamente *et al.*, 1988). Myofibroblasts are most numerous in the involutional phase of fibromatosis, whereas they are sparse in the early proliferative phase (Chung and Enzinger, 1981).

Proliferative conditions referred to as myofibroma (solitary) and myofibromatosis (multiple) differ from fibromatosis. The former conditions are characterized by a much more prominent myofibroblastic component including plump cells with abundant eosinophilic cytoplasm, resembling smooth muscle cells, and the presence of intervening richly vascular areas including haemangiopericytoma-like features. Myofibroma is localized in the dermis and subcutis, most commonly involving the head and neck region in infants and children (Chung and Enzinger, 1981). Myofibromatosis is a very rare multicentric proliferative disease, including involvement of internal organs and the skeletal system (Chung and Enzinger, 1981). Visceral lesions are already present at birth in most cases.

Treatment of fibromatosis is difficult. The tumour itself has a high recurrence rate of between 25 and 70 per cent (Fasching *et al.*, 1988; Enzinger and Weiss, 1995). The treatment of choice is surgical excision, with those tumours grossly involving muscle requiring wide local excision to obtain adequate tumour-free margins. Cases where complete excision is limited due to vital structure involvement may result in incomplete resection. The use of chemotherapy and radiation is controversial. Some authors report moderate benefit with chemotherapeutic agents (Goellner and Soule, 1980; Fasching *et al.*, 1988; Enzinger and Weiss, 1995), while others report beneficial effects of tamoxifen and non-steroidal anti-inflammatory agents (Enzinger and Weiss, 1995). Radiation therapy may prove useful preoperatively, in those instances where complete removal is not possible, or in recurrent cases (Mckenzie, 1972; Wara *et al.*, 1977).

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