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Desmoid Tumor of the Neck: CT and Angiographic Findings

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The term *desmoid tumor* (Greek *desmos* = band) was coined to designate tumors of a tendon- or ligamentlike consistency. They are derived from musculoaponeurotic supporting tissues and are histologically benign fibroblastic tumors. These rare lesions constitute only 0.03% of all neoplasms. Because of their predilection for the anterior abdominal wall, desmoid tumors have been categorically linked with "abdominal wall tumors" [1, 2]. Nevertheless, they are known to occur in virtually all parts of the body.

In 1856, Paget [3] reported a patient with this tumor in the arm, and, in 1923, Nichols [4] reported six patients with extraabdominal desmoid tumors. It was estimated that about two-thirds of them occurred in the abdominal walls [2]. Das Gupta et al. [5] reported a series of 72 extraabdominal desmoid tumors occupying the following areas in order of decreasing frequency: arms, legs, trunk, head and neck, and other. Only three patients in their series were reported to have desmoid tumors of the neck.

Radiologic description of these tumors has been lacking. Sacks et al. [6], in their presentation of two mesenteric desmoid tumors, showed lack of neovascularity in this lesion. Baron and Lee [7] emphasized sonographic and computed tomographic (CT) features in two patients with mesenteric desmoid tumors. We report one case of desmoid tumor in the neck with emphasis on the CT and angiographic appearance.

Case Report

A 54-year-old man noted a painless and slowly growing mass in the region of the right neck for several months. There was no history of direct or indirect trauma to the neck. His medical history was unremarkable and his family history noncontributory.

Physical examination revealed a hard mass occupying the right posterior cervical triangle in its lower half and extending beneath the sternocleidomastoid muscle to the anterior cervical triangle as well; the lower end extended beneath the clavicle by palpation and its borders could not be defined. The upper end of the mass was palpable 10 cm above the clavicle. The mass was firm and immobile. No abnormal pulsation or bruit could be heard.

A CT study of the neck and upper chest with intravenous contrast medium administration showed a solitary mass of greater density

than the surrounding soft tissues, about 8 cm in diameter, extending inferiorly to the right thoracic apex (fig. 1). We presumed that the hyperdensity was from contrast enhancement in the mass. The anterior and middle scalene muscles and the long muscle of the neck were incorporated into the mass lesion. Right common carotid angiography showed medial displacement of the right common carotid artery. Right subclavian arteriography demonstrated abundant neovascularity derived from the branches of the right subclavian artery in the mass (fig. 2A). A right axillary venogram after injection of the right basilic vein showed slight compression of the right subclavian vein. The other laboratory examinations were unremarkable.

At surgery the right sternocleidomastoid muscle was divided from its clavicular insertion to expose the mass. The mass showed a rough, reddish outer surface and was fairly well circumscribed. There was extension into the deeper structures medially and inferiorly with fixation to the musculature of the transverse processes from C6 to T1. The major arterial supply arose from the subclavian artery, which was torn during the process of dividing the vessels and was repaired and reconstituted with a no. 6 Gortex graft. The tumor was subtotally removed. The surgical specimen consisted of an 8 × 6 × 3.5 cm firm, nonencapsulated tumor with attached slips of skeletal muscle. On section, it was white to beige with focal hemorrhage and appeared to be incompletely excised. Microscopically, the tumor was moderately cellular and composed of plump spindle-shaped fibroblasts with bland nuclei in a collagenous matrix containing small blood vessels and mast cells (fig. 2B). There was no calcification. Mitotic figures were not seen. The tumor infiltrated skeletal muscle.

The patient had an uneventful postoperative course and was discharged in a stable condition. Regrowth of the tumor mass was clinically detectable within 5 months after surgery and cobalt-60 radiotherapy was given without any effect. At present, 1½ years after surgery, the patient is receiving Adriamycin chemotherapy. There is no evidence of distant metastases.

Discussion

Desmoid tumor occurs most often in the third to fifth decade of life, and about 70% of the patients are female. The etiology of this lesion is still unknown. Pregnancy, abdominal surgery, trauma, hormones, and heredity [8] have been implicated as causative factors. None of these can be traced in the clinical history of our patient.

These benign fibroblastic tumors usually infiltrate muscles and viscera and tend to recur if not radically excised. Spon-

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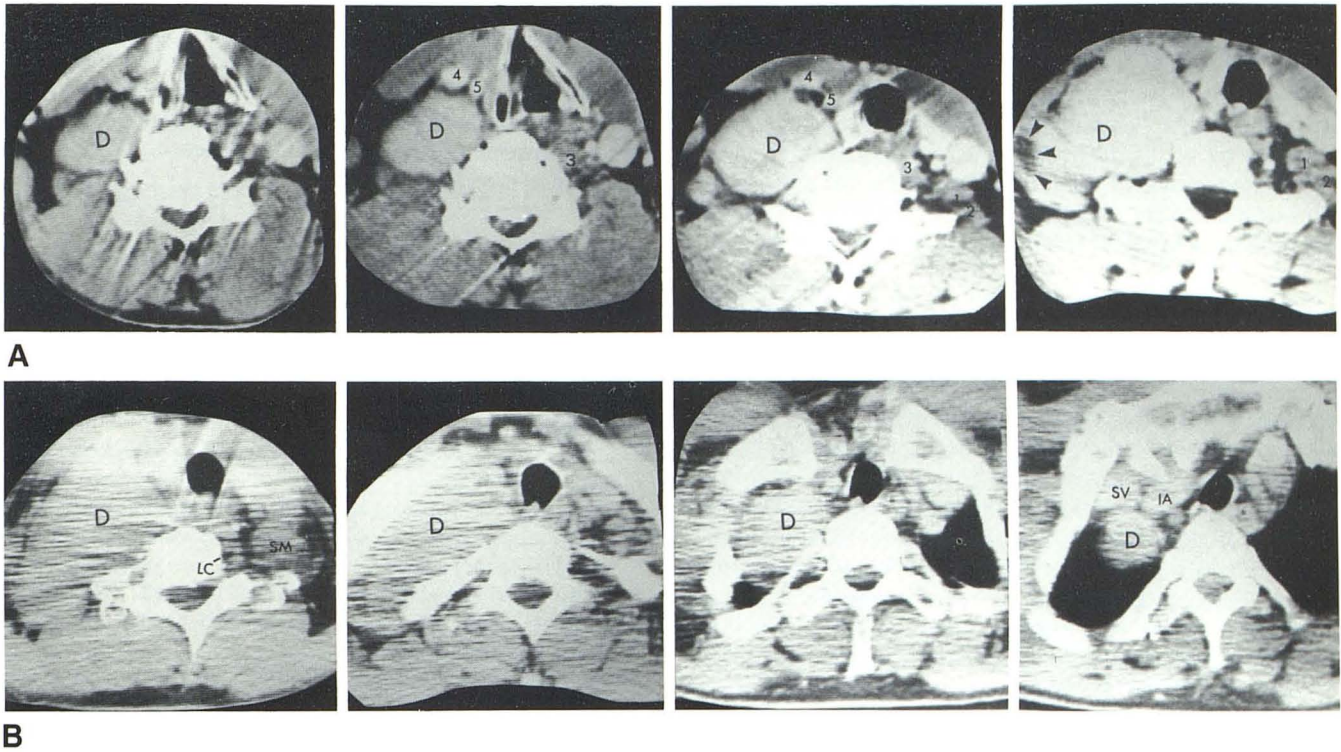
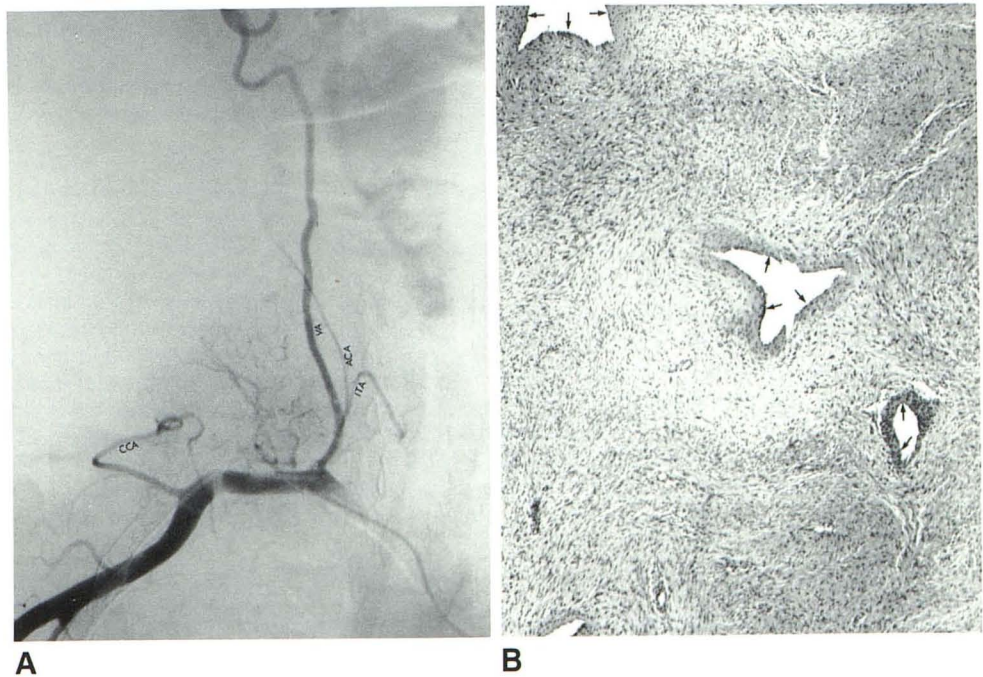


Fig. 1.—Postcontrast CT scans. **A**, Neck. Well defined right neck mass lesion (D) denser than surrounding muscles except for small area of fatty density (*arrowheads*). Left anterior scalene muscle (1), middle scalene muscle (2), and long muscle of neck (3) are identified, but these same muscles on right are incorporated in mass lesion. Right internal jugular vein (4) and right common

carotid artery (5) are displaced anteriorly and medially. **B**, Upper thorax and lower neck. Mass lesion (D) is behind right clavicle. Anterior, middle, and posterior scalene muscles on left are noted as confluent soft-tissue density (SM). LC = long muscle (longus colli); IA = innominate artery; SV = subclavian vein.

Fig. 2.—**A**, Right subclavian subtraction arteriogram, anteroposterior view. Neovascularity in mass lesion supplied by branches from right suprascapular, transverse cervical, and costocervical (CCA) branches of right subclavian artery. Medial displacement of proximal right vertebral artery (VA), ascending cervical artery (ACA), and inferior thyroidal artery (ITA). **B**, Photomicrograph of tumor demonstrates numerous spindle-shaped fibroblasts. Scattered small vessels (*arrows*).



taneous regression of a desmoid tumor has been reported [2]. However, experience in the literature indicates that total surgical removal is the treatment of choice [5]. Radiation [9] and antiestrogen therapies have been noted to produce some successful results. Postoperative recurrence of the tumor was estimated to be as high as 50% [10], presumably from inadequate removal of the tumor. Therefore, it is imperative to make a complete preoperative assessment of this tumor with regard to its location and extent. This is possible with CT.

Previous radiologic reports of this tumor emphasize the use of sonography [6, 7], which shows the lesion to be well defined and anechoic. Angiographically, Sacks et al. [6] did not demonstrate any neovascularity in the mesenteric desmoid tumor, despite scattered small vessels noted on the resected specimen. Hill et al. [9] were the first to show neovascularity of a desmoid tumor located in the neck similar to the finding described in our case. On CT, Baron and Lee [5] saw a few scattered fatty densities in an otherwise homogeneous, well defined soft-tissue mass in the mesenteric desmoid tumors. Unlike previous reports, we have shown incorporation of the neck muscles in the tumor owing to the infiltrative nature of this lesion, as confirmed on the histologic sections. Engulfment of skeletal muscles and scattered fatty densities appear to be the CT characteristics of this lesion.

CT provides useful information in defining both the osseous and soft-tissue extent of the cervical masses. Other neck masses, such as chordoma, neurofibroma, neurilemmoma, brachial cleft cyst, and lymphoma, rarely originate in skeletal muscles and usually exhibit certain CT features [11]. A tumor

harboring in the skeletal muscle, with smooth margins and scattered fatty densities, as in our case, has not been documented. Perhaps with that characteristic involvement of the skeletal muscle on CT, together with neovascularity on angiograms, desmoid tumor can be differentiated from other mass lesions. However, more documented cases are needed to substantiate this statement.

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