10 Tumors of Fibrous Tissue
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10.1 Introduction 149

Fibrous tissue consists of fibroblasts and an extracellular matrix containing both fibrillary structures (collagen, elastin) and nonfibrillary, gel-like ground substance. Both fibroblasts (spindle-shaped cells) and myofibroblasts, which are modified fibroblasts showing features common to fibroblasts and smooth muscle cells, produce procollagen and collagen. Collagen is the main component of the extracellular matrix, forming a heterogeneous group of closely related, noncontractile structures varying in function and distribution. Elastin is the main component of elastic fibers that are closely associated with collagen fibers. The ground substance of connective tissue is glycosaminoglycans (mucopolysaccharides), the most common types being hyaluronic acid and chondroitin 4- and 6-sulfates. Both elastin and glycosaminoglycans are also synthesized by fibroblasts [17, 48].

Tumors and tumorlike conditions of fibrous tissue are categorized into four groups according to their degree of malignancy: (a) benign fibroblastic proliferations, (b) fibromatoses, (c) fibrous tumors of infancy and childhood, and (d) malignant fibrosarcomas. Together they represent 5% of all cases of soft tissue tumors.

In the case of several fibroblastic proliferations (elastofibroma, fibroma of tendon sheath, extra-abdominal desmoids, fibromatosis coli), it is unclear whether these constitute reactive fibrosing processes or true neoplasms. Fibromatoses are aggressive, infiltrating lesions despite their histologically benign character, and aggressive fibromatoses or musculoaponeurotic desmoid tumors are by far the largest group of tumors of fibrous tissue. The terminology of childhood fibromatosis is confusing, and there are many classification systems based on different clinicopathologic factors such as age, localization, histology, and aggressiveness of the lesion [73]. Fibrous tumors of infancy and childhood and soft tissue fibrosarcomas are rare and have only rarely been reported in the radiologic literature. It is a constant finding that the majority of these tumors have a high recurrence rate after surgical resection, and recurrent lesions mostly have a more aggressive behavior than their primary counterparts. Another constant finding is the natural evolution of tumors of fibrous tissue, which are hypercellular in their initial stage and become more collagenous in later stages. Localization of the lesion and age of the patient are major diagnostic factors. Fibroma of ten-
don sheath, elastofibroma, all types of fibromatosis, and fibromatosis colli of infancy are characterized by typical localizations.

Ultrasound enables differentiation of solid and cystic masses, discrimination of circumscribed and diffusely infiltrating lesions, diagnosis of intraleSIONAL calcifications, and delineation of a mass from the surrounding structures. The combination of longitudinal and transverse scans allows accurate assessment of the localization of the lesion in three dimensions. On the other hand, since the density of many soft tissue tumors and of normal muscle is not very different on computed tomography (CT), the precise localization of the tumor margin and its relationship to neurovascular structures are difficult to assess using this modality.

Although magnetic resonance imaging (MRI) has the best capabilities in tumor characterizat ion, it is more important to estimate the extent and depth of these lesions correctly than to establish a possible histologic diagnosis. A correct histologic diagnosis of soft tissue tumors can be easily established, using percutaneous biopsy with or without the use of medical imaging techniques for guidance. The appearance on MRI will vary according to the relative proportions of the different tumor components [45]. In this regard, MRI parallels histology and allows a follow-up of the natural evolution or the evolution as a consequence of therapy of these lesions [33]. Good assessment of both the initial extent of disease and response to treatment will support new therapeutic strategies [4, 70, 72].

10.2 Benign Fibroblastic Proliferations

Benign fibroblastic proliferations constitute a heterogeneous group of well-defined entities. Some of these, such as nodular fasciitis, grow rapidly and are richly cellular. Others, such as fibroma of the tendon sheath and elastofibroma, grow slowly, are much less cellular, and contain considerable amounts of collagen [17].

10.2.1 Nodular Fasciitis

Nodular fasciitis, also called pseudosarcomatous fasciitis, infiltrative fasciitis, or proliferative fasciitis, is a benign soft tissue lesion composed of proliferating fibroblasts. It is characterized by rapid growth, which may arouse the suspicion of a sarcoma [39]. Although the cause is unknown, it is likely that it is triggered by local injury or a local inflammatory process. Most patients are asymptomatic or note only mild discomfort. Lesions are round to oval and mostly located in the upper extremity (48%), trunk (20%), head and neck (17%), and lower extremity (5%) [51]. Shimizu reported on a series of 250 patients with a mean age of 39 years and a peak in the fourth decade, in whom 44% of the lesions were located in the upper extremity, which is followed in frequency by the lower extremity and the trunk [64].

There are three subtypes of nodular fasciitis, defined according to their topography: the most common subcutaneous type, the intramuscular type, and the fascial type, which spreads along superficial fascial planes. Microscopically, younger lesions consist of fibroblasts embedded in a dense reticulin meshwork and birefringent collagen. There is a rich intervening myxoid matrix. Older lesions tend to have a more fibrous histology. In this regard, nodular fasciitis can be subdivided into three types based on the predominant histologic features: myxoid (type 1), cellular (type 2), and fibrous (type 3). However, since many different features may co-exist in one lesion, definition of a single fixed type is not always possible. There is no correlation between histologic subtypes and the subtypes classified according to their anatomic location. Histologic transformation from type 1 to type 2 and then to type 3 is indicative of the natural evolution of these lesions.

On ultrasound, lesions of this kind are mostly solid, and cystic change has been reported (Fig. 10.2a). On CT scan, nodular fasciitis has low attenuation values reflecting the myxoid character of the lesions [51]. The appearance on MRI reflects the gross morphology of the tumor. Myxoid and cellular lesions are iso- to hyperintense compared with skeletal muscle on T1-weighted images and iso-to hyperintense compared with fat on T2-weighted images (Fig. 10.1). Lesions with a more fibrous histology are markedly hypointense on all spin echo sequences [51]. On comparing these findings with the MR features of some neurogenic tumors (target sign) we named this appearance “the inverted target sign” (Fig. 10.2, 10.3). Three cases in our own series presented with the same MR appearance: a central area of low signal intensity and a peripheral area of higher signal intensity on T1-weighted images, (Fig. 10.2b), and an inverted signal intensity pattern