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Radiologic-Pathologic Correlation

Nodular fasciitis: a retrospective study of 272 cases from China with clinicopathologic and radiologic correlation

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ABSTRACT

Our aim is to describe the largest series of nodular fasciitis (NF) with emphasis on the clinicopathologic and radiologic correlation. A total of 272 cases of NF were diagnosed between 2004 and 2014 at our institution. There were 160 males and 112 females with age ranging from newborn to 77 years (mean, 36 years). The upper extremity was the most common location (34%), followed by the head and neck region (24%), trunk (21%), and lower extremity (14%). By radiology, the lesion appeared as well-defined homogeneous mass with low or isodensity on computed tomography, homogenous hypointense or isointense on T1-weighted sequences, and heterogeneous intermediate-to-high signal on T2-weighted sequences. Although all cases were composed of short intersecting fascicles of uniform plump spindle cell, the cellularity and stromal components varied considerably between different cases. In intramuscular or deeply seated NFs, extension into adjacent skeletal muscles or structures was often noted. Immunohistochemically, all cases showed diffuse staining for smooth muscle actin and calponin, with consistent negativity for desmin, h-caldesmon, and β-catenin. Of patients with available followed up information, only 1 experienced local recurrence due to incomplete excision. Our comprehensive study further demonstrated that NF had a wide clinicopathologic spectrum. Correlation with the radiologic features may help pathologists in arriving at an accurate diagnosis.

1. Introduction

Nodular fasciitis (NF) is a benign myofibroblastic proliferation that occurs predominantly in the subcutaneous tissue of young to middle-aged adults [1]. It typically presents as a fast-growing nodule that usually reaches 2 to 3 cm in size within few weeks. The most common site is the upper extremities, followed by the trunk, head and neck region [2-6]. Occasionally, the lesion may arise in unusual locations including the salivary gland, breast, mesentery, and vulva [7-11]. In most instances, it develops in the subcutaneous tissue and underlying fascia and appears as a well-defined nodule. On rare occasions, it may also occur in the dermis or arises within the skeletal muscles, vessels, joints, and peripheral nerves [12-25]. Because of the rapid growth, high cellularity, remarkable mitotic activity, local infiltration into adjacent tissues, and unusual clinical settings in some instances, misdiagnosing NF as a sarcomatous lesion continues to be a challenge for pathologists, which may lead overtreatment to the patients. A multidisciplinary study combining the clinical, pathologic, and radiologic characteristics on NF is sparse. In this study, we undertake a retrospective review of the largest series of NF with emphasis on the clinical, pathologic, and radiologic correlation.

2. Materials and methods

A retrospective study design was formulated, and institutional review board approval was obtained. Between January 2004 and December 2014, a total of 272 cases with NF were diagnosed at our institution. Special variants of NF such as cranial fasciitis, intravascular fasciitis, and ischemic fasciitis were excluded from the study. Clinical presentations, tumor site and size, radiologic appearance, and treatment information were obtained from the medical record, radiology and pathology reports, and discharge summary. The follow-up information was conducted via medical records or from the referring pathologists. Four-micrometer-thick hematoxylin and eosin–stained section was reexamined. Representative paraffin blocks or unstained slides containing tumor material from each case were selected for immunohistochemical study. The primary antibodies used in the study include smooth muscle actin (1A4, dilution 1:200; DAKO), desmin (D33, dilution 1:100; Dako), calponin (CALP, dilution 1:150; Maixin), CD10(SP67, ready to use; VENTANA), h-caldesmon (h-CD, dilution 1:300; DAKO), CD34 (QBEnd 10, dilution 1:100; DAKO), CD68 (KP1, dilution 1:600; Changdao), β-catenin (β-catenin1, dilution 1:200; DAKO), and cytokeratin (AE1/AE3, dilution 1:50; DAKO). Pretreatment was

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carried out according to manufacturer's recommendation. Omission of primary antibody and substitution by nonspecific immunoglobins were used as negative controls. Appropriate positive controls were run concurrently for all antibodies tested.

3. Results

3.1. Clinical findings

There were 160 males and 112 females with a ratio of 1.4:1. The patient's age at presentation ranged from newborn to 77 years. The newborn presented with a mass arising in the dorsal aspect of ulnar side of his left hand, whereas the oldest is a 77-year-old man who had a mass in his left shoulder. The mean and median ages were both 36 years. Approximately 65% of patients were in their third to fifth decades, whereas children (aged <10 years, 9 cases) and elderly patients (aged >60 years, 16 cases) accounted for 3.3% and 5.9%, respectively (Fig. 1). Clinically, most patients presented with a solitary subcutaneous nodule, which grew rapidly within 2 weeks or 1 month before surgical excision. However, patients with a preoperative duration of several months were not uncommon. One patient presented with 1-year history. Approximately three-fourths of patients were associated with slight pain or tenderness on palpation. Two patients with intraneural NF presented with symptoms of peripheral neuropathy. The patient with a tumor arising within the lateral cord of the left brachial plexus complained of left shoulder pain associated with numbness of the left thumb and index finger. Another patient with a tumor of the right median nerve complained of pain and numbness with decreased sensation of the right hand. History of antecedent trauma was not documented in all cases.

With regard to the anatomic site, 92 cases (34%) were located in the upper extremities especially the forearm, 65 cases (24%) in the head and neck region, 56 cases (21%) in the trunk, 37 cases (14%) in the lower extremities especially the thigh, 9 cases (3.3%) in the breast, 8 cases (2.9%) in the groin, and 5 cases (1.8%) in the vulva. Of the upper extremities, 45 cases were located in the volar aspect of the forearms, followed by shoulder (n = 18), upper arm (n = 11), elbow (n = 8), hand (n = 7, 5 in the palm and back of hand, 2 in the finger), and axillary (n = 3). Of the head and neck region, tumor occurred in the neck (n = 36); cheek (n = 11); supraclavicular region (n = 5); external ear region (n = 4); orbit (n = 4); temple (n = 2); and forehead, eyelid, and oral cavity (1 case each). Of the trunk, 26 cases were located in the chest wall, followed by back (n = 20), waist (n = 5), and abdominal wall (n = 5). Of the lower extremities, 20 cases were located in the thigh, followed by hip (n = 9), knee (n = 4), leg (n = 2), and foot (n = 2, 1 each in the heel and plantar). All patients were treated by surgery. In patients with follow-up information (ranging from 2 to 110 months), none of them experienced local recurrence except one after incomplete excision.

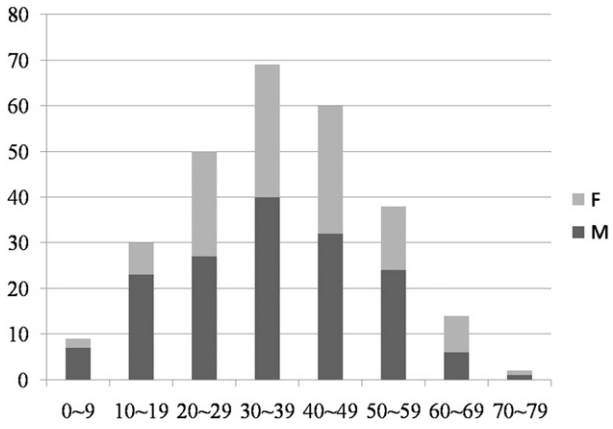

Fig. 1. Age and sex distribution in 272 patients with NF.

3.2. Imaging studies

Of 29 patients with recorded examinations, 15 patients had ultrasonography only before excision. Ultrasound scan usually showed a well-defined hypoechoic or isoechoic lesion situated in the subcutaneous tissues. Computed tomography (CT) or magnetic resonance imaging (MRI) was available in 14 cases. The lesion usually appeared as a fascia-based well-defined oval mass (Fig. 2A). A few intramuscular lesions were relatively large with ill-defined margins. On CT, it was low or isodense compared with adjacent muscles. In cases with cystic and mucoid degeneration, contrast-enhanced CT scan showed heterogeneous appearance with prominent peripheral rim-like enhancement in some cases (Fig. 2B). On MRI, it was homogenous hypointense or isointense on T1-weighted (T1WI) and heterogeneous intermediate to hyperintense on T2-weighted (T2WI) with enhancement after gadolinium administration (Fig. 2C-F).

3.3. Pathologic features

All lesions were solitary. Apart from 1 intradermal case, 1 periosteal case, and 16 intramuscular cases (5.9%), all the remaining cases were subcutaneous. They were described as well-circumscribed soft, rubbery-to-firm nodules or masses with glistening appearance on cut section (Fig. 3). The excised specimens measured from 0.5 to 7.0 cm in maximum diameter, with a mean and median size of 2.3 and 2.0 cm, respectively. Of note, the lesion in 238 cases (87.5%) was smaller than 4 cm. Those with a greatest dimension larger than 4 cm were mostly deeply seated or intramuscular lesions.

At low-power magnification, most were well circumscribed but unencapsulated (Fig. 4A and B), whereas a minority of cases had poorly defined borders with infiltration into the surrounding adipose tissue, skeletal muscle, or parotid gland (Fig. 4C-E). Entrapped nerve fascicles were noted in 2 intraneural NFs (Fig. 4F). On high power, typical cases were composed of uniform bland spindle cells that arranged in short intersecting fascicles within a loose stroma with extravasated erythrocytes and scattered lymphocytes (Fig. 5A). However, the cellularity and the stroma components varied considerably between cases, ranging from hypercellular with less stromal material, highly myxoid with abundant mucoid matrix to heavily hyalinized with deposition of keloid-like collagen fibers (Fig. 5B-D). Cases with combined areas were not uncommon (Fig. 5E). Mucoid degeneration was a frequent feature, which was prominent in a few cases creating a broken fish net-like appearance (Fig. 5F). Approximately 10% of cases contained scattered multinucleated giant cells. These giant cells had more eosinophilic cytoplasm. Compared with the classic osteoclasts, these giant cells were relatively smaller and could contain only 2 to 3 nuclei. Their presence ranged from scarce that could be easily ignored to numerous (Fig. 5G and H). Generally, mitotic figures were not difficult to find (ranging from 1 to 10/10 high-power field), but cytologic pleomorphism and nuclear atypia were absent.

3.4. Immunohistochemical study

The spindle cells showed diffuse expression of smooth muscle actin (Fig. 6A). They were also positive for calponin, muscle specific actin, and CD10. Intralesional histiocytes and small multinucleated giant cells were positive for CD68 (Fig. 6B). Other markers including desmin, h-caldesmon, β-catenin AE1/AE3, CD34, and S-100 protein were all negative.