Infantile nodular fasciitis of the hand: A case report and literature review

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ABSTRACT

Pediatric nodular fasciitis is uncommon and has a preference for the head and neck region. Occurrence in other anatomic locations is uncommon. We describe here a case of nodular fasciitis that arose in the hand of a newborn infant who presented with a rapidly growing mass. On MRI, it was heterogeneous isointense on T1-weighted and hyperintense on T2-weighted images. Histological examination showed short intersecting fascicles of uniform spindled myofibroblasts embedded in a myxoid to collagenous stroma, consistent with a nodular fasciitis. However, the lesion was initially diagnosed as an infantile fibrosarcoma due to the rapid growth, brisk mitotic activity and focally infiltrative architecture. This study illustrates that unusual presentation of nodular fasciitis may cause diagnostic confusion.

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1. Introduction

Nodular fasciitis is a benign proliferation of fibroblasts and myofibroblasts, which was first reported by Konwaler et al. in 1955 [1]. It is also called pseudosarcomatous fasciitis because it is not uncommonly mistaken for a sarcoma due to rapid growth, high cellularity, brisk mitotic activity, and infiltrative borders [2,3]. Although this benign entity has been well recognized for a long time, it continues to be a diagnostic challenge for pathologists in routine practice especially when dealing with those that occur in unusual clinical settings [4,5]. This benign lesion is common in young to middle aged adults and is infrequent in infant and young children. Pediatric nodular fasciitis typically occurs in the head and neck region [6,7], whereas occurrence in other anatomic locations is distinctively rare [8]. Although nodular fasciitis can occasionally occur in the hand of children [9], it is rarely reported to develop in the hand of a neonate.

2. Case report

The patient is a full term male neonate. He was delivered by spontaneous vaginal delivery at 40 weeks gestation to a 28-year-old primigravida woman. On the second day after birth, a nodule was found in the dorsal aspect of ulnar side of his left hand. There was no birth injury. As the mass grew rapidly and became more and more prominent, the baby was taken to a clinic at 29 days of age. Physical examination revealed a subcutaneous mass, which was firm and measured approximately 4 cm in diameter. The overlying skin was brown reddish with a glistening appearance (Fig. 1). The baby seemed to feel uneasy and cried when the mass was palpated. Magnetic resonance imaging showed a relatively well circumscribed mass that was heterogeneous isointense on T1-weighted images with peripheral enhancement and internal cystic degeneration and high intense with irregular areas of isointense on T2-weighted images (Fig. 2). The lesion measured 4.5 × 2.5 × 1.0 cm in size. It was considered as a hemangioma. He was admitted to a children hospital and preoperative preparation was carried out. At surgery, a non-encapsulated gray nodule was identified which was closely adherent to tendon and the fourth metacarpal bone. At intraoperative consultation, it was described as ‘a poorly circumscribed spindle cell neoplasm showing nuclear atypia and high mitotic activity. In the comment, it was stated that the lesion involved the tendon and adjacent fibroadipose tissues, with partial destruction of bone’. A wide local excision was performed. The mass was therefore completely excised together with the fourth metacarpal bone. The final histological diagnosis of the excised mass was an infantile fibrosarcoma. The pathological materials were later sent to our department for further confirmation.

Histologically, the lesion was composed of uniform spindled myofibroblasts that arranged in loose fascicular to storiform patterns, and embedded in a variably myxoid to collagenous stroma (Fig. 3a). In some areas, there was marked microcystic degeneration. The stroma contained scattered lymphocytes as well as small clusters of lipid macrophages (Fig. 3b). On high power, the spindled cells had eosinophilic cytoplasm with elongated nuclei, vesicular chromatin and small nucleoli. There were no cytological pleomorphism and nuclear atypia. Mitotic figures were not difficult to identify (average, 3/10HPF), but atypical forms were absent. The lesion encased the tendon and eroded the bone cortex (Fig. 3c, d). Immunohistochemically, the spindle cells...
were positive for smooth muscle actin and calponin. They were negative for desmin, h-caldesmon, β-catenin, AE1/AE3, CD34, S100 protein and WT1. Intralesional scattered histiocytes were positive for CD68. In addition, by fluorescence in situ hybridation (FISH) using breakapart probe, more than 50% of the counted tumor cells showed separated green and orange signal, indicative of the rearrangement of the USP6 gene (Fig. 3e), and no separated signals were detected with ETV6 breakapart probes. Both the histological features, immunophenotypes and molecular study were consistent with a nodular fasciitis. The baby remains well 9-months after surgery.

3. Discussion

Nodular fasciitis is a benign self-limited process which is commonly seen in young to middle aged adults with a peak incidence in the third and fourth decades. The principle location is the upper extremities (34%–46%), followed in frequency by the head and neck (20%–26%), trunk (15%–21%) and lower extremities (14%–18%) [2,3]. Infant and young children below the age of 10 years can also be affected, but are less common. It has been estimated that pediatric nodular fasciitis accounts for less than 10% of all cases [6]. In contrast to nodular fasciitis of adulthood which occurs predominantly in the upper extremities, pediatric nodular fasciitis has a preference for the head and neck region [7,10]. The lesion may involve various sites, including the forehead, eyelid, orbit, cheek, maxillofacial region, nasal dorsum, maxilla, mandible, external auditory canal, postauricular region, neck, larynx and parapharyngeal space [11]. Occurrence outside the head and neck region is extremely rare. Mazura et al. reported an intramuscular nodular fasciitis involving the rectus abdominus muscle of an 11-year-old girl [8]. In this study, we describe a rare case of nodular fasciitis that arose in the dorsum of the hand in a newborn infant. This case was briefly included in a recent series of nodular fasciitis, but the detailed clinical and pathological features have not described [12]. Although nodular fasciitis occurs most commonly in the upper extremities, hand is infrequently involved. Up to present, approximately 30 cases have been reported in the English literature [13]. Apart from two teenagers, all the other patients were adults with equal gender distribution. To date, nodular fasciitis has not been described in the hand of a newborn infant yet.

Like nodular fasciitis of other locations, patients with a hand lesion usually presented with a solitary rapidly growing mass which was often associated with mild pain and tenderness. The average size was...
about 2 cm ranging from 1 to 6 cm. Large and multiple lesions were very rare and should be diagnosed with caution [14]. It has been shown that the lesion tended to occur in the volar aspect of the hand, with the palm and fingers being mostly involved. Hypothenar and thenar eminence, first web space, and dorsum of the hand were occasionally affected. There seemed a predilection for the right hand.

Radiological features of nodular fasciitis were usually considered non-specific. By ultrasonography, the lesion appeared as a hypoechoic mass which was often well-defined. On CT scan, it was often fascia-based and was low to isodense. In cases with myxoid change, contrast-enhanced CT scan demonstrated heterogeneous appearance with prominent peripheral nodular or rim-like enhancement. Because of the varying histological components, the appearance on MRI varied between different cases. Whereas myxoid and cellular nodular fasciitis were typically iso- to hyperintense to skeletal muscle on T1 weighted images and iso- to hyperintense to fat on T2 weighted images, fibrous lesions were distinctly hypointense on T1 and T2 images. It is worthy to mention that nodular fasciitis of the hand could show infiltrative borders and or bone erosion, which might lead to a diagnostic pitfall [15,16].

Although the histological features of nodular fasciitis have been well described, the lesion continues to be a diagnostic challenge for pathologists. Due to the occurrence shortly after birth, clinically rapid growth, poorly defined borders, presence of brisk mitotic figures, locally infiltrative fashion, and unexpected presentation in the hand, the current case was initially misdiagnosed as an infantile fibrosarcoma. Unlike nodular fasciitis, infantile fibrosarcoma is composed of compact long fascicles or bundles of immature-appearing fibroblasts frequently accompanied by prominent lymphocytic infiltrate. It is highly cellular with notable nuclear atypia and a high rate of mitotic activity. In addition, herringbone arrangement a feature frequently seen in an infantile fibrosarcoma is rarely noted in a nodular fasciitis. Immunohistochemically, both nodular fasciitis and infantile fibrosarcoma express actins, although the staining is more diffuse and stronger in the former. A recent study showed that cytoplasmic expression of WT1 was found in infantile fibrosarcoma but not in nodular fasciitis [17]. Cytogenetically, infantile

Fig. 3. Histological features. a (10×) The uniform spindled cells are arranged in short intersecting fascicles within a loose stroma showing focal microcystic change (arrow). b (20×) Interstitial deposition of collagen fibers with small clusters of macrophages (arrow). c (10×) Encasement of the tendon (arrow). d (10×) Cortical erosion (arrow) (arrow) of the involved metacarpal bone. e Split signals (cells with one green, one orange signal) were considered positive by FISH, indicating USP6 rearrangement (arrow).
fibrosarcoma was characterized by a recurrent t(12;15)(p13;q25), resulting in an ETV6-NTRK3 fusion gene [18]. In contrast, nodular fasciitis harbored MYH9–USP6 fusion gene [19]. Molecular assay, such as FISH or RT-PCR, may serve as a helpful tool in the diagnosis of challenging cases. Other infantile fibroblastic and myofibroblastic lesions than may enter the differential diagnosis include fibroma of tendon sheath, fibrous hamartoma of infancy, infantile myofibroma, infantile fibromatosis and inflammatory myofibroblastic tumor. Fibroma of tendon sheath is a well-circumscribed nodular lesion with peripheral slit-like vessels. Compared with nodular fasciitis, it is hypocellular and more densely collagenous. Fibrous hamartoma of infancy occurs mainly in the maxilla, upper arm and trunk with rare involvement in the hand. It is composed of a admixture of fibromatosis-like fibrous trabeculae, mature adipose tissue and variable clusters of immature-appearing cells embedded in loose matrix, forming a characteristic organoid pattern. Infantile myofibroma differs from nodular fasciitis by its biphasic pattern characterized by a central high cellularity hemangiopericytoma-like component and in the hand. Misdiagnosing this benign lesion as a sarcoma could lead to unnecessary treatment, including amputation of a finger. Fine needle aspiration examination of nodular fasciitis is also very difficult especially when dealing with those lesions of unusual anatomic sites, including the hand. Extreme hypercellularity in the face of cellular isotropy is subject to being mistaken for a low-grade sarcoma, resulting in wide local resection [20]. Therefore, an excisional biopsy is considered as an optional approach before treating the fibrous lesions of the hand. If the histological features and immunohistochemical study strongly suggest the possibility of a nodular fasciitis, the more conservative ‘wait and see’ policy is recommended.

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References