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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.

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

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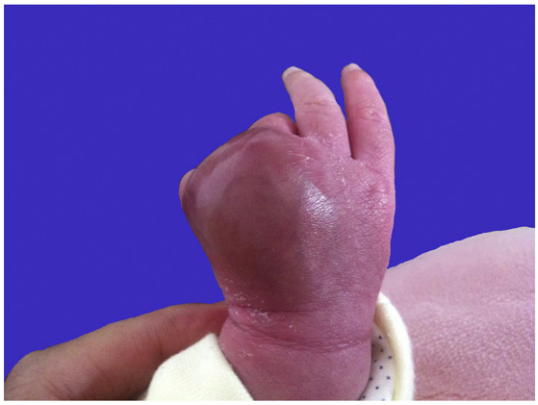

Fig. 1. The dorsum of the left hand. The lesion appears as an immobile, well-circumscribed, brown-reddish translucent mass involving the subcutaneous tissue of dorsal aspect of the hand.

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

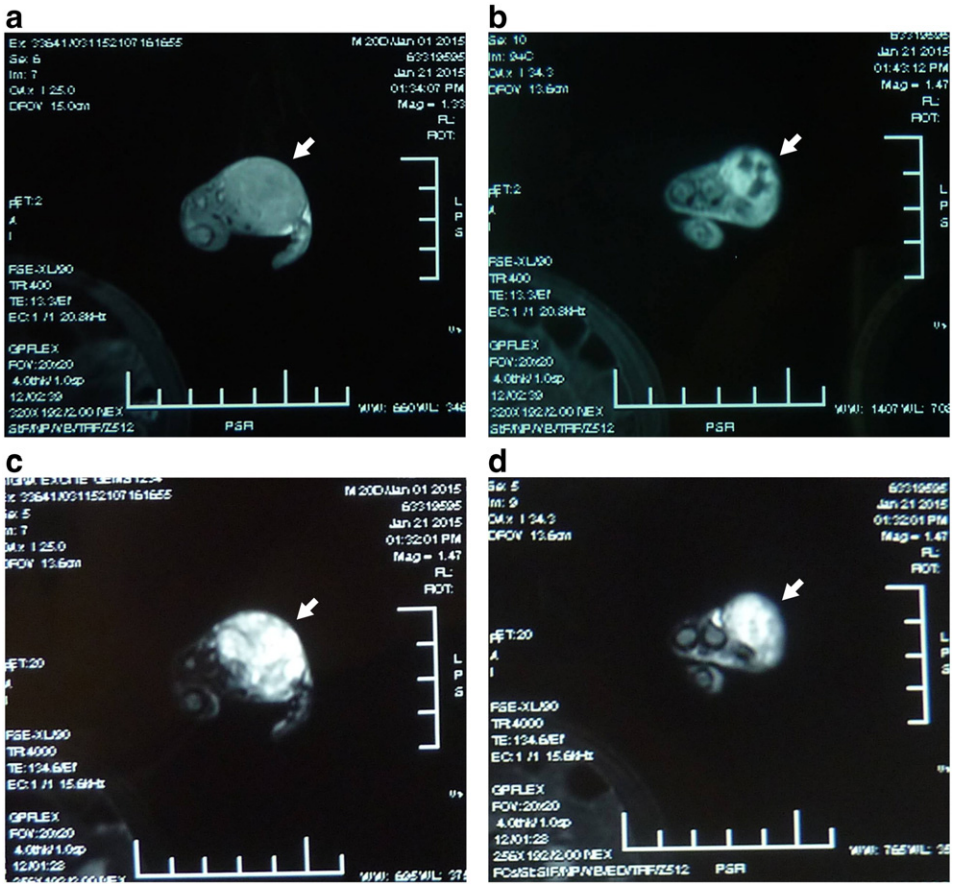

Fig. 2. MRI appearances. a Sagittally, T1-weighted sequence shows heterogeneous isointense signal (arrow). b Horizontally, T1-weighted sequence shows peripheral enhancement and internal cystic degeneration (arrow). c (Sagittal) and d (Horizontal) T2-weighted sequences demonstrate heterogeneous T2 signal with central hyperintensity (arrows).