CASE REPOSITORY

Unusual Presentation of Infantile Myofibroma in the Deep Palm of a Child: A Case Report and Discussion of the Differential Diagnosis

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Infantile myofibroma or myofibromatosis is a myofibroblastic and fibroblastic proliferation that is most commonly reported in children younger than 2 years of age. It is a benign process composed histologically of a biphasic pattern of spindle-shaped cells surrounding a zone of less differentiated cells in a hemangiopericytoma-like pattern. We report this tumor in a unique presentation in the deep palm of a 2-year-old child without skin ulceration and with an intimate association with the median nerve. The well-circumscribed nature of the tumor facilitated complete excision with neural preservation. Final pathology was consistent with an unusual type of myofibroma or myofibromatosis. Conservative management with partial excision has been advocated for these masses because of potential surgical morbidity and its benign nature. This case report highlights the differential diagnosis of uncommon soft tissue tumors in the pediatric hand as well as the importance of a surgeon's surgical assessment in guiding treatment. *(J Hand Surg Am. 2016;■(■):■−■.*

Key words Infantile myofibroma, median nerve, myofibromatosis, palm.

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INFANTILE MYOFIBROMA OR MYOFIBROMATOSIS is a myofibroblastic and fibroblastic proliferation that is most commonly reported in in the head and neck of children younger than 2 years of age.[1,2] Rare cases in the hand have been reported, and these may be associated with skin ulceration and contracture.[2–8] Subtotal resection has been advocated by some authors to reduce surgical morbidity,[2–8] but no definitive treatment guidelines are established.

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CASE REPORT

A 26-month-old male infant presented with a mass in the palm that had been enlarging over the course of 3 months. The parents sought treatment after noting the increasing size of the palm. The child had remained clinically asymptomatic and appeared to use the hand without discomfort.

He was initially taken to another surgeon who planned an excisional biopsy. During exploration, the mass was found to be attached to the median nerve extending from the transverse carpal ligament to the common digital nerves. The surgeon elected to perform an incisional biopsy of the volar portion of the mass. The preliminary pathology diagnosis from

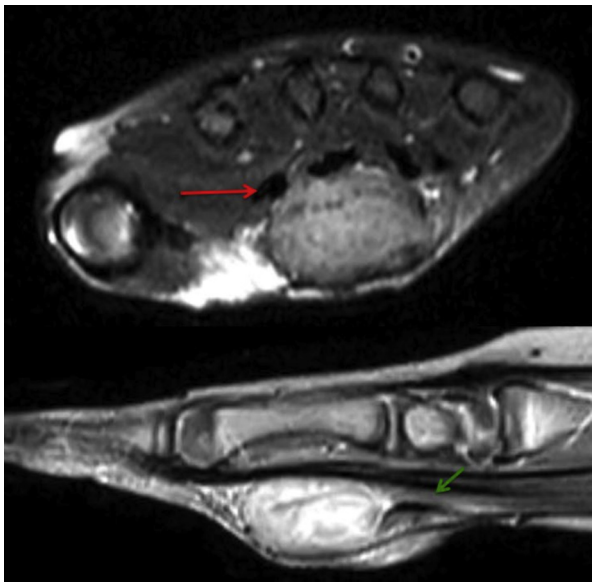

FIGURE 1: Coronal (top) and sagittal (bottom) views of the magnetic resonance imaging study. The mass was intimately associated with the median nerve and fascicles distally (green arrow). It was superficial to the flexor tendons without direct attachment (red arrow).

the biopsy was myofibroma, but no detailed immunohistochemistry was performed. The child was subsequently referred to our institution for further evaluation and treatment.

A magnetic resonance imaging study of the hand demonstrated a mass in the volar surface of the palm just deep to the palmar aponeurosis. The mass was hyperintense on T2-weighted images and was found to be solid without cystic components (Fig. 1). The median nerve was seen proximal to the mass and deep to the transverse carpal ligament, but distally, the nerve could not be distinguished from the tumor. After discussion with family, we proceeded with a plan to excise the tumor. The patient's previous incision was opened and extended distally into the palm and proximally across the wrist (Fig. 2A). The median nerve was identified at the level of the wrist and traced distally. The ulnar nerve, artery, and palmar arch were identified along the ulnar aspect of the mass (Fig. 2B). All the branches of the median nerve, including the motor branch, were identified distally. Individual fascicles and digital branches were traced and found to be splayed across the radial, ulnar, and deep capsular surfaces of the mass. Each fascicle or nerve branch was separated from the mass as it was reflected in an ulnar to radial direction (Fig. 2C). The flexor tendons were noted deep to the mass and the nerve fascicles, but no direct extension from a tendon to the mass was observed. The child had no residual neurological deficit after the tumor excision and had a well-healed scar at 1-year follow-up (Fig. 2D).

Gross examination showed a well-circumscribed soft tissue mass that had a gray-white whorled appearance to the cut surfaces. Histological sections demonstrated an encapsulated spindle cell proliferation. The neoplastic cells were arranged in whorled bundles with short to intermediate length fascicles and occasional myxoid foci. Mast cells and small slitlike arborizing blood vessels were observed interspersed within the neoplastic cells (Fig. 3). The neoplastic cells had eosinophilic cytoplasm with plump, oval nuclei and occasional cigar-shaped forms. The chromatin was fine and vesicular with small nucleoli. No significant atypia, hyperchromasia, or nuclear pleomorphism was recognized. Mitotic features were seen in the hypercellular areas; no atypical forms or necrosis was observed. Multiple immunohistochemical stains were performed to further characterize the spindle cell neoplasm. The neoplastic cells were strongly and diffusely positive for vimentin and desmin (both common to myofibroblastic tumors) and negative for S-100 staining, which is positive in tumors of neural origin. The remainder the immunohistochemical findings are displayed in Table 1.

DISCUSSION

Williams and Schrum[1] first described a myofibroblastic proliferation in a newborn in 1951, naming it a congenital fibrosarcoma. Chung and Enzinger[2] later isolated this subtype of myofibroblastic tumor and described the first large series of 61 cases, coining the term infantile myofibroma. In their series, myofibromatosis or myofibroma typically presented before the age of 2 years with almost two-thirds present at birth. They are most commonly found in the head and neck in both solitary and multicentric forms. The classic microscopic description is a well-circumscribed nodule with short bundles of spindle-spaced cells with a center of necrosis or hemangiopericytoma-like pattern.

From our review of the literature, only 16 cases of solitary myofibromas have been reported in the upper extremities of children, with 2 specific cases in the palm (Table 2).[2–8] In the largest reported series, Chung and Enzinger[2] described 6 cases of infantile myofibroma in the upper extremity, with the most distal case occurring at the wrist. Netscher et al[5] described myofibromatosis occurring in the dorsum of the hand of a child who presented with a thick scar