

# Atypical Abnormalities of the Hand and Wrist in Children: The Utility of Magnetic Resonance Imaging (MRI)



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## Introduction

Common indications for performing MR of the hand and wrist are cartilage, tendon and ligament injuries; joint infection and evaluation for osteomyelitis; bony and soft tissue tumors; and carpal tunnel syndrome. The purpose of this exhibit is to show some atypical or uncommonly seen abnormalities of the hand and wrist, in which MRI showed characteristic imaging features and helped in the management.

## Cases

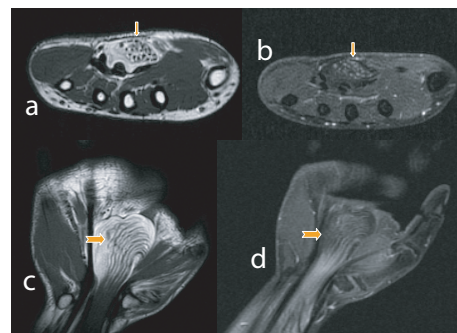
### Lipomatosis of Nerve<sup>2</sup>: (Fibrolipomatous Hamartoma)

Lipomatosis of nerve is non-hereditary, hypertrophy of mature fat and fibroblasts in the epineurium<sup>2</sup>.

Patients commonly present at birth or early childhood with a slow growing mass at the wrist, hand, or forearm. The upper extremity is affected in 78%–96% of cases, particularly the median nerve (85% of cases)<sup>1</sup>. The ulnar nerve is the second most commonly affected site.

MRI appearance of lipomatosis of nerve is pathognomonic showing longitudinally oriented cylindrical areas of low to intermediate signal intensity (nerve fascicles) surrounded by hyperintense adipose tissue in a diffusely thickened nerve both on T1- and T2W images<sup>3</sup>.

There is no curative treatment for lipomatosis of nerve, as complete resection results in severe sensory and motor deficits along the distribution. Carpal tunnel release may relieve some symptoms of compression in patients with median nerve involvement.



- 12-year-old female with enlarging swelling on the right palm for several months
- Axial T1W (a) and fat-suppressed, contrast enhanced axial T1W (b) images shows an enlarged median nerve in the carpal tunnel ↓, prominence of the nerve fascicles with interspersed fat and intense enhancement after contrast. The lesion extends into the palm as seen on T1W (c) and fat-suppressed, contrast enhanced coronal T1W (d) images →. A diagnosis of lipomatosis of nerve was made and the patient was managed conservatively.

### Macrodystrophia Lipomatosa

Macrodystrophia Lipomatosa is a form of localized gigantism characterized histologically by overgrowth of adipose and periosteal osteoblasts<sup>4</sup>. Bone overgrowth is typically more prominent volarly and distally, often resulting in osseous bowing. The lesion has a propensity to involve the second and third rays of the hand or foot. The osseous overgrowth does not progress after puberty.

MRI reveals high signal intensity on T1W in the soft tissues signifying overgrowth of adipose tissue with interspersed fibrous elements or septae<sup>4</sup>.

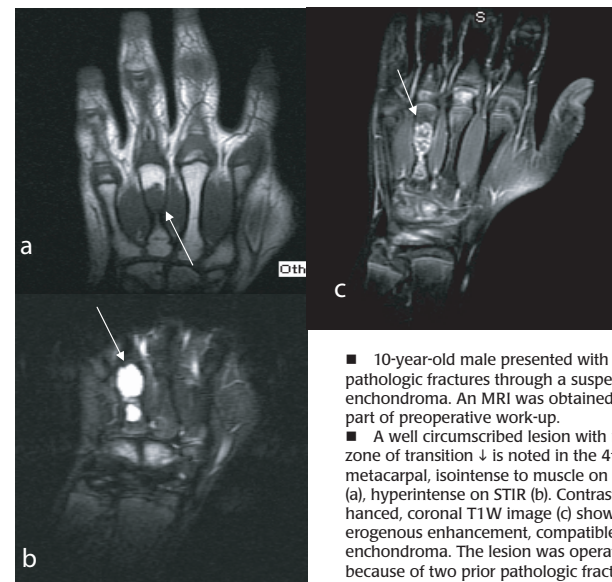


- 16 year old female with enlargement of the 4th and 5th digits
- Plain film (a) reveals prominence of the soft tissues around the 4th and 5th digits ↓ with subtle overgrowth of the middle phalanx of the 5th digit. Coronal T1W image (b) shows extensive deposition of adipose tissue with fibrous elements in the soft tissues of the 4th and 5th digits ↓. This is an atypical location for macrodystrophia lipomatosa, as the usual distribution is in the 2nd and 3rd rays. The patient was managed conservatively.

### Enchondroma

Enchondromas are common benign cartilaginous neoplasms that arise in the medullary canal due to continued growth of the residual benign cartilaginous rests that are displaced from the growth plate<sup>5</sup>. Enchondromas are common in the tubular bones of the hands and feet which comprise 50% of all enchondromas.

Enchondromas are monostotic, non-aggressive, central metaphyseal lesions showing a narrow zone of transition. They may cause cortical thinning and contain stippled cartilaginous matrix. Enchondromas, in the hands or feet, can be purely lytic. On MRI enchondromas are low signal intensity on T1W and very high signal on T2W images, with lobular margins, internal septations and punctuate signal voids representing calcified matrix. The high signal intensity is due to high water content of the mucopolysaccharide extracellular matrix. Gadolinium enhanced appearance is that of "arcs and rings" reflecting the lobular growth pattern and chondroid matrix<sup>6</sup>.



- 10-year-old male presented with two pathologic fractures through a suspected enchondroma. An MRI was obtained as part of preoperative work-up.
- A well circumscribed lesion with narrow zone of transition ↓ is noted in the 4th metacarpal, isointense to muscle on T1W (a), hyperintense on STIR (b). Contrast enhanced, coronal T1W image (c) shows heterogeneous enhancement, compatible with enchondroma. The lesion was operated on because of two prior pathologic fractures. Histology showed an enchondroma.

### Vascular Malformations

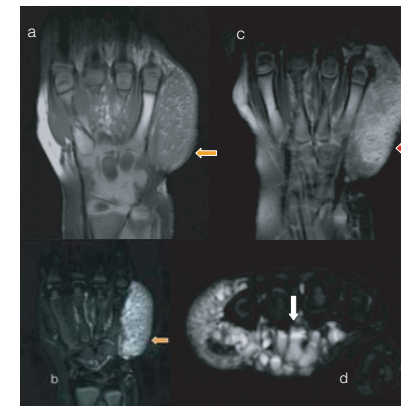
Vascular malformations are developmental abnormalities which are composed of a combination of vascular channels: venous, capillary, lymphatic, arterial or mixed, with or without fistula<sup>7</sup>. They can involve superficial soft tissues or deep tissues including bone.

MRI can define the channel type in a vascular malformation. The venous malformations are demarcated or infiltrative lesions with high T2 signal intensity with absence of arterial flow voids. Variable gadolinium enhancement (patchy or complete) is seen and is more conspicuous with fat suppression. Oval low signal phleboliths are found in 50%.

Lymphatic malformations are slow flow lesions also showing high T2 signal. The macrocystic malformations show characteristic fluid-fluid levels and may show high T1 signal due to hemorrhagic or proteinaceous content. Gadolinium enhancement occurs only in the septal components of the macrocystic lesions and may occur throughout in the microcystic lesions.

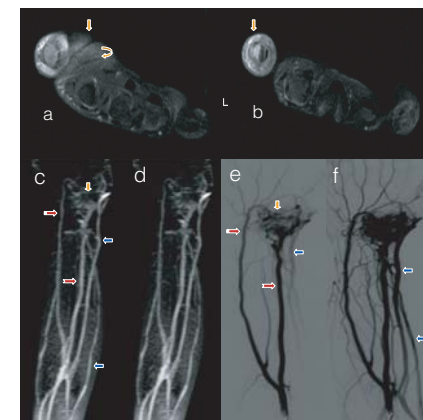
Arteriovenous malformations (AVM) are rare localized or infiltrative high flow malformations. The predominant MRI finding consists of serpiginous flow voids on T1- and T2W images which are better delineated on MR angiography. The enlarged arteries and veins are centered about the AVM nidus<sup>8</sup>.

### Lymphatic Malformation



- 22-month-old female presenting with a slow growing swelling over the palm of the left hand since birth.
- A mass → is seen lateral to the 5th metacarpal bone which is isointense on coronal T1W (a) hyperintense on STIR (b) images. The lesion is also hyperintense on fat-suppressed T2W axial image (d) and fluid-fluid levels are noted ↓. Diffuse enhancement in the microcystic component is noted on the fat-suppressed, contrast enhanced coronal T1W image → (d). The cystic components were treated with sclerosants.

### High-Flow Arteriovenous Malformation



- 4-year-old male presented with a localized swelling in the thenar eminence with a thrill and bruit.
- Fat-suppressed, contrast enhanced, axial T1W images (a, b) show a diffusely enhancing soft tissue mass around the 1st metacarpal bone and on the thenar eminence ↓. A flow void is also noted →. MRA images (c, d) show the ulnar → and radial → arteries feeding a nidus ↓ with early draining veins ←. A diagnosis of high flow AVM was made. The MRA findings of AVM were confirmed by an angiogram performed during therapeutic embolisation (e, f).

### Dysplasia Epiphysealis Hemimelica (DEH): (Trevor-Fairbanks Disease)

Dysplasia epiphysealis hemimelica (DEH), an uncommon, non-hereditary skeletal developmental disorder, is an osteochondroma arising from an epiphysis. The incidence is 1 in 1,000,000<sup>9</sup>.

This disease usually affects a single lower extremity, with rare involvement of the upper extremity. DEH is usually restricted to the medial or lateral side of the limb (hemimelic), with the former site affected twice as frequently as the latter. DEH is categorized into three different forms: a localized form (monostotic involvement), a classic form (more than one area of osseous involvement in a single extremity), and a generalized or severe form (disease involving an entire single extremity). Boys are affected approximately three times as often as girls<sup>9</sup>.

The disease progresses from stippled calcifications adjacent to one side of the epiphysis to ossification and eventually confluence with the epiphysis. This appears as a lobulated mass projecting from the epiphysis<sup>10</sup>.

MRI is useful in identifying the extent of epiphyseal involvement, joint deformity, and effect on surrounding soft tissue. The lesion and involved epiphysis have similar signal intensity, seen as intermediate signal intensity on T1W and high signal intensity on T2W images. Areas of low signal intensity on T1- and T2W images indicate areas of calcifications.

Surgical intervention is more frequently required for these lesions than for solitary osteochondromas as the epiphyseal location is often associated with pain, deformity, or loss of normal mechanical function<sup>11</sup>. Excision or even incomplete resection may result in reduction of symptoms.



- 8-year-old male with hard, slow growing knot on the posterior aspect of the wrist.
- Plain films (a) show enlargement of and a bony outgrowth from the lateral portion of the distal ulnar epiphysis ↓. Sagittal T1W (b) and coronal 3D GE (d) images show an enlargement of and an outgrowth from the lateral ulnar epiphysis with a thin cartilaginous cap. Coronal MPRG image (c) shows stretching and altered signal intensity in the TFCC →. Scapho-lunate dissociation and scapho-lunate ligament disruption → are also noted. These findings, which were confirmed at arthroscopy, were clinically unsuspected. They helped plan the surgery better and the TFCC and scapho-lunate ligaments were repaired and the bony outgrowth was shaved. DEH affecting the lateral aspect of the distal ulnar epiphysis is exceptionally rare.

### Granulomatous Infection in Acute Myelogenous Leukemia

Bacterial infections seen in patients with acute myelogenous leukemia include cutaneous infections such as staphylococcus and streptococcus, cat-scratch fever, tuberculosis and atypical mycobacteria<sup>12</sup>.

MRI features mimic those of chloroma (granulocytic sarcoma)<sup>13</sup>, appearing hypointense on T1W, hyperintense on T2W, and demonstrating homogenous enhancement after contrast administration.



- 4-year-old female with acute myelogenous leukemia presented with soft tissue swelling around her wrist
- Plain films (a) show soft tissue swelling → around the wrist and no bony abnormalities. Extensive soft tissue thickening is seen surrounding the wrist tendons on sagittal and axial STIR images (b, c). There is uniform, intense enhancement on the fat-suppressed, contrast enhanced sagittal T1W image (d) →. Initially a diagnosis of chloroma (granulocytic sarcoma) was made.

Histology showed an unusual granulomatous infection with bacilli suggestive of either atypical mycobacterium or cat-scratch fever. The infection was successfully treated.

## Summary

MRI provided additional information in all cases which helped plan management and surgery.

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