CURRENT CONCEPTS

Tumor-Like Conditions of the Hand and Upper Extremity

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Tumor-like conditions of the hand and upper extremity typically present as masses and can be confused as more serious conditions. The differential diagnosis of these lesions can cross over with many more commonly recognized benign and malignant upper limb tumors, and it is, therefore, important for the hand surgeon to be familiar with tumor-like conditions. The diagnosis of these lesions often can be made on clinical grounds supported by a careful physical examination and plain film radiography. Advanced imaging and excisional biopsy may be needed in many circumstances to secure the diagnosis given their similarity with bone and soft tissue sarcomas. *(J Hand Surg Am. 2017;■(■):■−■. Copyright © 2017 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Tumor-like conditions, tumors, lesions.

TUMOR-LIKE CONDITIONS OF THE HAND and upper extremity typically present as masses that may or may not be accompanied by pain. The differential diagnosis of tumor-like conditions of the hand and upper extremity is broad, and it is important to rule out other conditions. Identifying the correct diagnosis will guide medical and surgical treatment. In many cases, the appropriate history and physical examination, plain radiographs, and if needed, advanced imaging, most commonly magnetic resonance imaging (MRI), will lead to a diagnosis. Under most circumstances, an excisional biopsy will also be needed for the definitive diagnosis. In this article, we describe frequently encountered tumor-like conditions of the hand and wrist and list features to help differentiate these lesions from neoplastic conditions.

LIPOFIBROMATOUS HAMARTOMA

Lipofibromatous hamartomas are rare lesions that can infiltrate the peripheral nerves of the upper extremity, potentially leading to neurological dysfunction as well as deformity. Different terms have been used to describe this lesion, including fibrolipomatous hamartoma, fibrolipoma, lipofibroma, and intraneural lipoma.[1] In 1969, Johnson and Bonfiglio[2] introduced the term lipofibromatous hamartoma based on histological findings, but the etiology of this lesion remains unknown.

Patients with these lesions typically present within the first 3 or 4 decades of life.[3] Paresthesia is the presenting symptom in one-third of patients,[1] but the lesion is characterized by an expansive proliferation of fibrous and adipose tissue within the nerve epineurium, and it most commonly affects the median nerve in the wrist and forearm (Fig. 1).[4] This tumor can be associated with macrodactyly when the lesion involves the digital nerves. Lipofibromatous hamartomas of the brachial plexus as well as the other major nerves and nerve trunks are well recognized and can reach sizeable proportions (Fig. 2).

The role of surgery for the treatment of lipofibromatous hamartomas is controversial. For lesions of the carpal tunnel, carpal tunnel decompression alone may relieve symptoms. For lesions involving the brachial plexus, limited surgical debulking (Fig. 3) may be indicated for pain management or cosmetic concerns owing to the size of the lesion. Extensive surgical resection of lesions of the brachial plexus or other major peripheral nerves can result in irreversible neurologic injury.

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GANGLION

The word ganglion comes from a Greek word that, simply translated, means knot. The dorsal wrist is the most common location for ganglions and they are more common in women, encountered most often in the second through fourth decades of life. The volar radial ganglion is more common in the fifth through seventh decades.[5] Other locations for ganglions in the hand and wrist include the first dorsal compartment of the wrist, the carpal tunnel, Guyon canal, the carpometacarpal joints, the flexor tendon sheaths, the proximal interphalangeal joints, the distal interphalangeal joints, and the scapholunate interosseous ligament. The dorsal wrist ganglion accounts for approximately 70% of all ganglions of the hand, whereas volar radial ganglion cysts account for approximately 20%.[5] When ganglions arise in the proximal aspect of the flexor tendon sheath, they are typically referred to as retinacular cysts, whereas those located at the distal interphalangeal joint are termed mucous cysts. The etiology of the wrist joint ganglion remains poorly understood. Hypotheses for the development of ganglion cysts include (1) synovial protrusions of the joint, (2) degenerative cyst formation, and (3) trauma.[6–8] In an MRI study of 122 patients by el-Noueam et al,[7] 37 patients had identifiable internal derangement of the wrist. Ultrastructurally, the ganglion can be characterized by a cyst lined with fibrillated connective tissue and sparse mesenchymal cells, filled with fluid composed of necrotic debris.[9,10] A physical examination including transillumination is typically sufficient for a clinical diagnosis of ganglion cyst, but occasionally imaging is needed. Magnetic resonance imaging is very reliable for identifying ganglion cysts and is the preferred imaging study. Ultrasound has also been shown to be very reliable, particularly for the identification of occult ganglion cysts.[11]

In general, surgical treatment of ganglion cysts of the hand and wrist are considered more successful than nonsurgical treatments, with the exception of ganglion cysts in children in whom spontaneous resolution can be expected.[12,13] Nonsurgical treatment in adults including aspirations and injections often results in recurrence.[13,14] Although many favor open excision of ganglion cysts emanating from the wrist joint, arthroscopic removal has shown good results.[15]

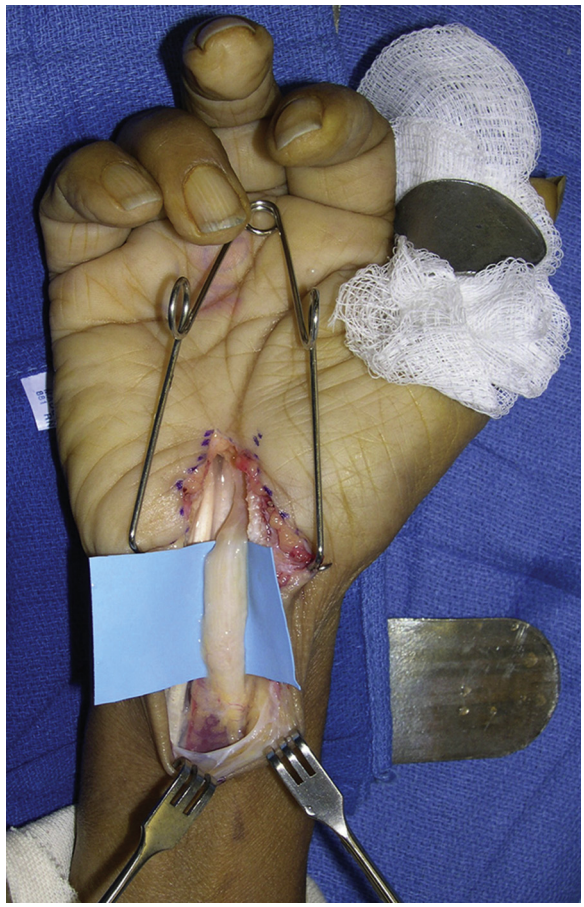

FIGURE 1: A 35-year-old man with a lipofibromatous hamartoma of the median nerve presenting with median nerve paresthesia.

SIMPLE CYST/UNICAMERAL BONE CYSTS

Simple bone cysts and unicameral bone cysts (UBCs) can occur in the carpal bones, distal radius and ulna, metacarpals, or phalanges. In the carpus, the lunate and scaphoid are the most common locations for development. Simple bone cysts/UBCs are most commonly seen in patients younger than 20 years, although these cysts have been seen in adults.[16] Simple bone cysts/UBCs are fluid-filled cavities lined by a thin membrane. Plain radiographs show a lucent lesion that is centrally located. Simple bone cysts/UBCs can be treated expectantly and often will spontaneously heal as the patient approaches skeletal maturity. The differential diagnosis includes aneurysmal bone cysts, posttraumatic cysts, avascular necrosis, intraosseous ganglion cysts, degenerative cysts, and neoplastic tumors.[17] Patients can present with a pathological fracture resulting from a simple cyst/UBC, often characterized by a fallen-leaf sign on plain radiographs.[18] If a patient is symptomatic or has had a pathological fracture, surgical intervention is warranted, and curettage with or without bone grafting is performed, with internal fixation as needed. Curettage alone without grafting has been successful in the carpal bones.[17] In larger lesions, bone grafting with allografts, calcium phosphate, or autografts can