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Case Report

A Juxta-Articular Myxoma of the Thumb

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Juxta-articular myxomata are benign tumors that are mostly encountered in the vicinity of larger joints. Few cases in the hand have been reported. We present a case of a juxta-articular myxoma at the metacarpophalangeal joint of the thumb in a 40-year-old man. The preoperative diagnostic work-up included 4-dimensional magnetic resonance angiography and ultrasound. The histochemical examination of the resected tumor established the diagnosis definitively. Follow-up magnetic resonance imaging scheduled with no clinical suspicion of tumor recurrence 9 months after surgery revealed no obvious recurrence. At 14 months, the patient had full motion without pain and declined further imaging.

Juxta-articular myxomata (JAM) are benign soft tissue neoplasms of mesenchymal origin. These cystic tumors resemble a ganglion and tend to recur after resection.[1] Because they sometimes behave in a locally destructive fashion and can cause pain, they can be difficult to distinguish from a myosarcoma.[2] Juxta-articular myxomata are mostly encountered in the vicinity of larger joints such as the knee joint.[3] Few cases involving the hand have been reported.[4,5]

Case Report

A 40-year-old male patient reported a mass at the metacarpophalangeal joint of the left thumb. Initially, this tumor was hardly noticeable, but it grew slowly over the past year. Because it was situated in the commissure between the thumb and the index finger, the pressure when grasping larger objects provoked mild pain.

The clinical examination revealed a solid, circumscribed round mass of 25 mm that was adherent to the ulnar-palmar aspect of the metacarpal joint of the thumb. It was scarcely visible because the texture and color of the skin were normal. The tumor did not adhere to the skin or tendons (Fig. 1). The stability and mobility of the thumb joints were not compromised.

The clinical impression of close contact to the joint was verified by ultrasound. The final diagnosis of a JAM was suspected on the basis of a magnetic resonance imaging (MRI) scan (Figs. 2–4) and 4-dimensional magnetic resonance angiography. As a well-circumscribed, septated, scarcely vascularized (supported by duplex sonography) mass with no pathological tumor-feeding vessels, it could be distinguished from a myosarcoma. We forewent plain films because they seemed sufficiently replaced by MRI and would be useful only to rule out arthritic changes and bone infiltration.

Without suspicion of malignancy, we elected to proceed with marginal resection rather than a biopsy to avoid a second operative procedure in a delicate region.

At surgery, the tumor could be completely resected without destruction of the joint ligaments, tendons, vessels, or nerves (Figs. 5, 6).

The histological examination revealed nonmalignant spindle cells embedded in a myxoid matrix (Fig. 7). The diagnosis was confirmed immunohistochemically with the affirmation of CD34 and the exclusion of S100 and actin.

The postoperative healing process was uneventful. The thumb regained full motion and became completely pain-free.

Regular follow-ups including ultrasound and MRI examination were initially scheduled on a yearly basis. A first control MRI

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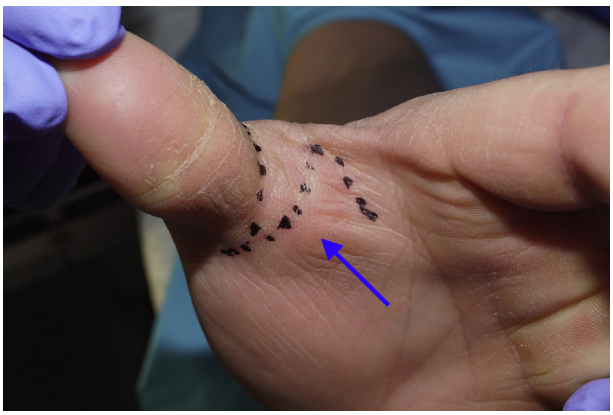

Figure 1. Juxta-articular myxoma at the ulnar-palmar aspect of the metacarpophalangeal joint of the left thumb (blue arrow).

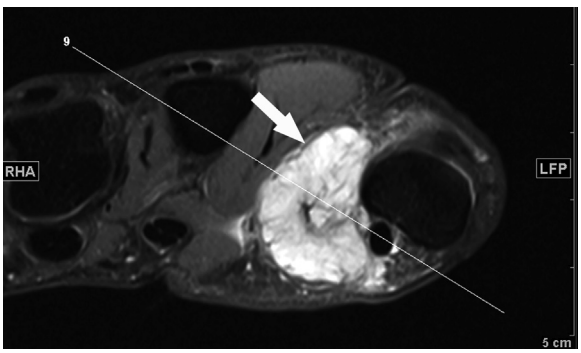

Figure 2. T2-weighted fat-suppressed axial MRI-image showing a strongly hyperintense, well-circumscribed, septated mass (arrow) adjacent to the metacarpophalangeal joint of the thumb. The line corresponds to the level of the coronal view of Figure 3.

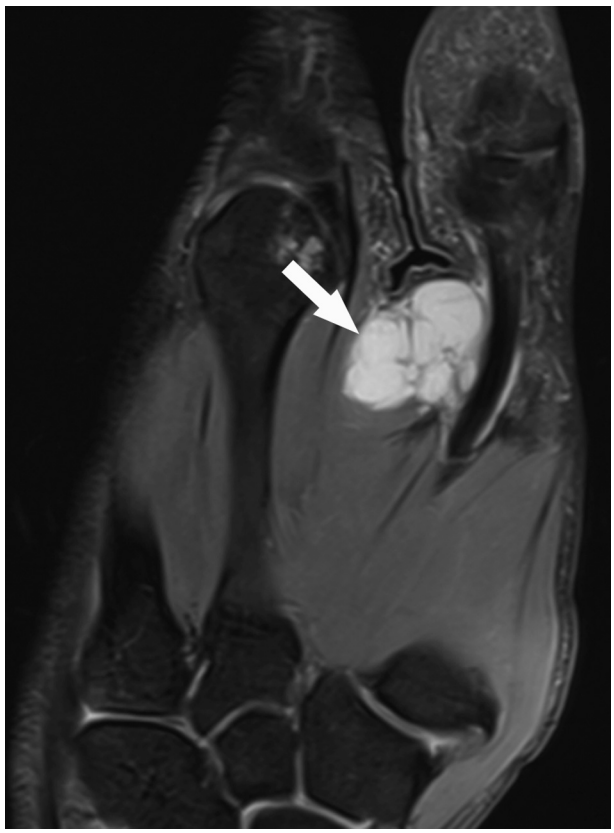

Figure 3. Coronal MRI scan of the first and second rays of the left hand corresponding to the section line in Figure 2. The mass (arrow) has well-defined margins. It is slightly displacing but not invading the surrounding (and normal) musculature and adipose tissue.

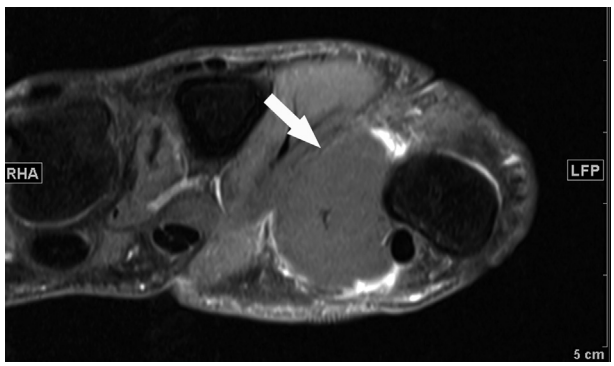

Figure 4. Nearly any contrast enhancement of the mass (arrow) surrounded only slightly dilated (for compressed) veins.

assessment, conducted without clinical suspicion of tumor recurrence 9 months after surgery, revealed no obvious recurrence. At 14 months after surgery, the patient remained pain- and symptom-free. Compared with the contralateral side, he had full range of motion, equal power, and normal sensibility in the thumb. The patient elected to defer additional imaging during the COVID-19 pandemic.

Compliance With Ethical Standards

All procedures performed in this case were in accordance with the ethical standards of the institutional and national research committee (Human Research Ethics Committee of Western Sydney Local Health District reference number 5452) and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. This article does not describe any studies with animals performed by any of the authors. Approval and informed consent were obtained from the patient of this case report.

Discussion

Less than 1% of all upper-limb tumors occur in the hand,[6] so tumors at this body site are not well-studied.[7] In the patient in the current report, the rare diagnosis of a JAM in the hand was established.

Myxomata clinically present as a slowly enlarging mass and do not necessarily provoke pain.[5,8] Most commonly, they are found in larger joints such as the knee,[1] shoulder, elbow, foot, and ankle, and only rarely in the hand and wrist.[9] Men aged 30 to 50 years seem to be affected more often.[2,5]

Myxomata can be identified as different types according to their tissue of origin (bone or soft tissue).[10] Deriving from the bone itself, myxomata are mostly seen in the jaw, whereas periosteal myxomata often occur in the long bones (eg, the femur).[11]

Treatment of a myxoma is complete surgical excision. Sometimes, additional procedures (eg, meniscectomy) are necessary.[5] Continued follow-up is important because the local recurrence rate after resection is high. Patients should be informed during

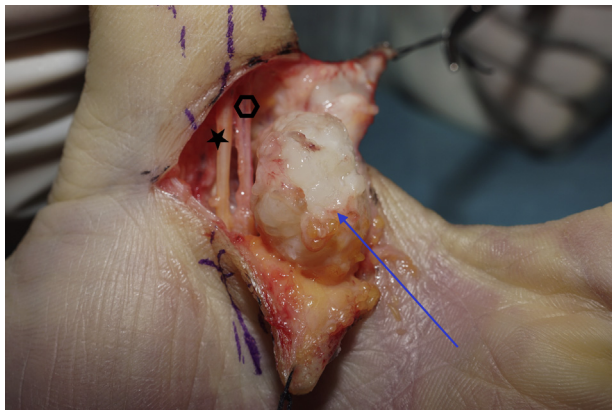

Figure 5. Intraoperative view of the left thumb. The juxta-articular myxoma (blue arrow) radially displaced the ulnar-palmar nerve (black star) and artery (black hexagon).

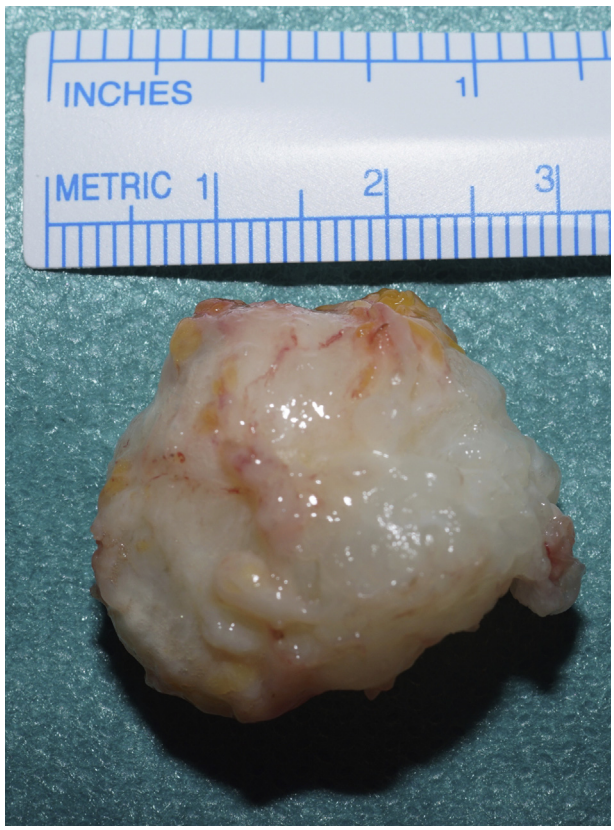

Figure 6. Juxta-articular myxoma of the left thumb.

preoperative counseling about the necessity for regular follow-up visits.[12,13] Previous studies described a recurrence rate of 34%, often within 18 months after surgery.[2] Incomplete resection may be the reason for this high recurrence rate.[1] In one patient, a recurrent myxoma led to amputation of the hand after 5 surgical attempts.[14] Usually, limb-sparing excision is possible in the hand, because tumors often become symptomatic when they are still small. The presence or absence of pain does not seem to be applicable for differentiating benign from malignant hand lesions.[7]

First described by Virchow in 1863,[14] the pathogenesis of myxomata is still incompletely understood, but the literature describes an association with previous trauma and osteoarthritis.

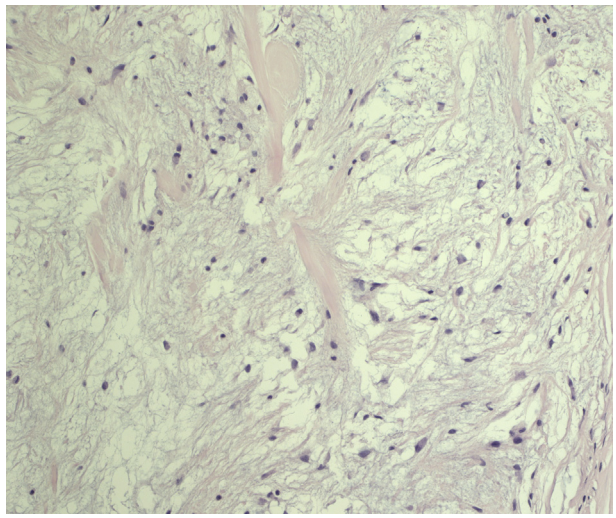

Figure 7. Hematoxylin eosin–stained histological section showing scantily distributed mesenchymal cells in a myxoid stroma, morphologically consistent with a juxta-articular myxoma.

Neither of these applied to the patient in the current study. Whether a neoplastic or a reactive process leads to proliferation of these mesenchymal cells remains unknown.[13,15] When looking for genetic abnormalities in DNA sequencing of myxomata, polymerase chain reaction shows Gs alpha mutations in intramuscular myxomata, but not in JAMs, so different underlying molecular mechanisms are postulated.[16]

Although classified as benign lesions, myxomata can be locally destructive and thus cause symptoms like pain or nerve palsy.[15,17] In the setting of local tissue destruction, one must consider malignancy in the differential diagnosis.[2]

Macroscopically myxomata present as cystic formations[5] of soft or friable consistency, white to yellow, ranging from 2 to 6 cm in size.[2] Microscopically these tumors can appear like fetal Wharton's jelly.[18] Few spindle-shaped fibroblast-type or stellate and fusiform cells are found within reticulin fibers and a myxoid matrix. The mucin consists of hyaluronic acid and mucopolysaccharides. Vasculature is usually poorly developed.[1,3,19] One report of an unusually large myxoma in the hand even showed hormone receptors.[20]

A few differential diagnoses should be considered when seeing a patient with a swelling on the hand or wrist as in our patient. The most probable diagnosis would be a ganglion cyst; however, these are smaller than JAM,[5] show no septation upon MRI, and have a less developed myxoid component.[8] A hibernoma of the hand should also be considered, even as a rare cause of carpal tunnel syndrome. This tumor of brown fat tissue shows spindle cells and myxoid structures like a myxoma but would be positive for PS100 and CD34.[21]

In cases of a rapidly growing mass, a sarcoma must always be considered.[12,13] Unfortunately, the diagnosis is often confirmed only by histologic examination after excision, so reoperation may be needed.[7] Careful preoperative planning including MRI and even core biopsy should be considered in these cases. Nodular fasciitis is a benign reactive myofibroblastic lesion that may occur after trauma. It also presents as a rapidly growing subcutaneous mass and is sometimes said to be a benign version of a sarcoma. Microscopic spindle-shaped cells in myxoid matrix with collagen are found. Nodular fasciitis is positive for α-smooth muscle actin and negative for many other markers (β-catenin, S100, CD34, High-Mobility Group AT-Hook 2, cytokeratin, epithelial membrane antigen, caldesmon, and desmin).[22] Finally, an intramuscular myxoma

should be considered in a differential diagnosis. It is more common in women around age 50 and often affects large muscles in the thigh or shoulder. In contrast to JAM, it shows only minimal cystic changes and has a low recurrence rate even with incomplete resection.[1,2,8] On computed tomography, MRI, and ultrasound, it presents with an intrinsic high water content and a surrounding rim of fat.[8] Activating missense mutations at the Arg201 codon of the Gs alpha gene leading to increased levels of cyclic adenosine monophosphate are detected in intramuscular myxomata (and are also found in McCune-Albright syndrome and sporadic fibrous dysplasia of bone) but not in JAM.[16]

In our patient, the diagnosis of a JAM was confirmed after immunohistochemical staining that was positive for CD34 but negative for S100 and actin.

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