Unusual Causes of Carpal Tunnel Syndrome

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Background

Carpal tunnel syndrome is the most common peripheral neuropathy in humans and has a prevalence of 3.7% of the US general population [1, 2]. While most cases are idiopathic, there are several well-known conditions which are often associated with carpal tunnel syndrome. These include pregnancy, hypothyroidism, diabetes, obesity, overuse, trauma, renal failure, and inflammatory arthropathy [3]. In all of these conditions, it is compression, entrapment, irritation, or global compromise of the nerve (as in diabetes) that leads to symptoms [3]. However, many of the more rare etiologies are not as familiar to most physicians, but are important to consider when caring for these patients. The mechanism of causing neuropathy can vary from compressive, inflammatory, infectious, or traumatic.

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Compressive

Tumors

Various space-occupying lesions have been identified as rare causes of carpal tunnel syndrome. Wu et al. reported a small series of patients [4] with carpal tunnel symptoms due to tophaceous gout (10), tenosynovitis (7), and tumors (8). In a recent study, Martinez-Villen et al. identified nonneural tumors or tumorlike lesions in 22 (5.3%) of 414 patients surgically treated for a nerve compression syndrome of the hand and forearm [5].

Benign tumors such as lipoma may cause secondary compression [6] and in rare cases may arise from the flexor tenosynovium [7, 8]. Intraneural lipomatous tumors have also been described [9]. These lesions will require decompression of the nerve from within the epineurium.

Fibroma of tendon sheath, though quite rare, is another benign tumor that should be considered. The typical presentation is a slowly enlarging, painless mass [10]. Males are more often affected, particularly in the 40–60-year-old range [11]. These tumors are usually well circumscribed, attached to the tendon or tendon sheath. Identifying the correct diagnosis and underlying cause are crucial, as release of the transverse carpal ligament alone may not predictably relieve symptoms or solve the primary problem in these cases. Excision of the mass is necessary to prevent recurrent symptoms.

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Ganglion Cysts

Ikeda et al. [12] describe a case of a 34-year-old patient who presented with rapidly progressive symptoms of numbness, pain, and weakness, as well as changes on EMG. Ultrasound revealed a ganglion compressing the median nerve. She underwent carpal tunnel release and ganglion excision; her symptoms resolved. Cysts may compress the motor or digital branch of the median nerve and in some cases may affect both the median and ulnar nerves [13]. Particularly in patients with rapidly progressive symptoms, the presence of a ganglion cyst should be considered. Physical exam may underestimate the size of these lesions as they often track down to the joint. Magnetic resonance imaging (MRI) or ultrasound can confirm the diagnosis and delineate the size of the lesion. Successful treatment hinges on decompression or excision of the lesion.

Foreign Body

It is possible for a foreign body to cause atypical synovitis and subsequently carpal tunnel syndrome, though only a few cases have been described. In one case report, a patient was found to have a piece of wood compressing the median nerve [14]. On exam, one should carefully inspect for signs of an entry wound and palpate for any abnormal masses. It is also important to clarify details of the history regarding exposure to specific materials. Ultrasound is a helpful tool to rule out such possibilities, bearing in mind that certain foreign objects are not visible on plain X-rays. Once the location of the object has been determined, removal is recommended.

Amyloidosis

Patients with amyloidosis may develop protein deposition in the flexor tendon sheath, resulting in median nerve compression. Deposition in the transverse carpal ligament is also possible, leading to thickening of the ligament. One recent study found that symptoms were bilateral in 97% of cases [15]. These patients usually respond poorly to steroid injections. MRI may show thickening due to amyloid deposition. Further assessment to confirm the diagnosis includes Congo red staining and immunohistochemical studies performed at the time of surgery. Amyloidosis should be considered in the differential particularly in patients with bilateral symptoms, refractory to conservative measures.

Calcific Tendinitis

Similarly, calcium accumulation (as seen in calcific tendinitis) can cause CTS. While most common in the flexor carpi ulnaris, calcium deposition may occur in a variety of areas, including the flexor tendons and carpal bones. Previous studies have found this condition to be the most common in patients between 30 and 60 years old [16]. Females are more often affected than males, and the dominant hand is more often involved [17].

Presentation may resemble infection or inflammatory conditions, with symptoms including erythema, localized pain, and swelling [18]. CRP may be elevated. AP and lateral X-rays are often unremarkable, depending on the size of the mass. Oblique views can be helpful. Advanced imaging, with CT or MRI, is often indicated to visualize the areas of involvement. In cases of acute CTS due to calcium deposition, treatment requires not only decompression of the nerve but also resection of the calcifications if possible.

Inflammatory Conditions

Gout

Gout is another less common but important condition to consider. Patients with chronic, long-standing gout are particularly at risk, as the large tophi may result in mass effect and subsequent symptoms [19]. Deposition in the transverse carpal ligament, median nerve, or carpal tunnel floor can occur. Tophaceous gout may also affect the flexor tendons themselves, causing stiffness, flexion contracture [20, 21], and tendon infiltration [22].

Serum urate levels aid in the diagnosis. Wrist joint or tendon sheath aspiration is also recommended. Fluid should be sent for crystal analysis with polarized light microscopy. Urate crystals are needle shaped and negatively birefringent. Preoperative MRI can help define the location and size of the lesions. CT is helpful if calcifications are present. Surgical decompression is recommended, in addition to medical management of the hyperuricemia. These patients are at increased risk of recurrence. It is also important to recognize that carpal tunnel syndrome may be the first manifestation of gout.

Calcium Pyrophosphate Dihydrate (CPPD) Crystal Deposition

Similarly, several case reports describe patients with CPPD crystal deposition or pseudogout [23]. In addition to the more common sites including the TFCC and DRUJ, calcifications of the flexor tendons may occur. The floor of the carpal tunnel and epineurium of the median nerve are other potential areas of involvement.

Calcifications are typically visible on plain films. The carpal tunnel can be particularly helpful. Chondrocalcinosis and degenerative changes of the joints are an associated finding in many of these patients and may be a clue to the correct diagnosis. Crystal analysis is recommended for further evaluation. In contrast to gout, CPPD crystals are rhomboid shaped and positively birefringent on polarized light microscopy.

Infection

A number of infectious organisms have been known to cause carpal tunnel syndrome. The mechanism appears to be similar regardless of the organism, due to the mass effect from the inflammatory process. Case reports of roundworm [24], atypical mycobacterium [25], and histoplasma capsulatum [26] [27] have been described. Adequate treatment hinges on eliminating the mass effect.

Trauma

Several types of direct and indirect trauma may lead to carpal tunnel syndrome, either acutely or in a delayed fashion. Distal radius fractures are perhaps the most common example, associated with acute carpal tunnel syndrome in 5.4–8.6% and delayed carpal tunnel syndrome in 0.5–22% of cases [28]. Possible causes include direct trauma to the nerve, entrapment of the nerve due to displaced fracture fragments, and/or increased canal pressures secondary to hematoma. A high index of suspicion is crucial when evaluating these patients, as a delay in diagnosis can have devastating consequences. Urgent surgical release and fracture fixation may be necessary in symptomatic patients or those with progression of symptoms.

A recent case control study by Dyer et al. examined risk factors for acute CTS in patients with a distal radius fracture [29]. These authors found that greater translation of fracture fragments, seen in higher energy injuries, was associated with a higher risk of developing acute CTS. A threshold value was identified in only one subgroup of patients, females younger than 48 years old, with 35% translation. This suggests that prophylactic carpal tunnel release may be indicated in some patients.

Less common traumatic causes include both bone forearm fractures and elbow dislocation. A thorough physical exam is necessary in all trauma patients. Indirect causes such as blunt trauma or hemorrhage are also important. For example, patients on chronic anticoagulation or hemophiliacs are particularly at risk [30].

Congenital/Anatomic Variants

A number of anatomic variants exist, involving both the bony structures and the soft tissues. Preoperative X-rays are helpful to identify patients with bony abnormalities potentially contributing to their symptoms. Accessory carpal bones [31], coalitions (pisiform-hamate) [32], and osteophytes (of the trapezium) have been described [33]. It is reasonable to obtain preoperative CT or

MRI in such cases, to further define the bony structure for planning purposes.

Several authors have identified anomalous muscles contributing to carpal tunnel syndrome. The palmaris longus is one example. The original cadaveric work was done by Reimann et al. in 1944. While distal variations of this muscle seem to be more common, Basu et al. report a case of a dual tendon, central muscle belly variant [34]. Reversed palmaris longus [35] and palmaris profundus are also possible [36]. Barutcu et al. report a series of three patients with bilateral carpal tunnel syndrome due to an anomalous transverse carpal muscle visualized intraoperatively [37]. Ultrasound and MRI may appear normal. In many cases, such variations are first identified intraoperatively, underlining the importance of careful dissection.

A bifid median nerve is another possibility. Due to the increased cross-sectional area, compression in the carpal tunnel can occur [38]. Advanced imaging, including MRI or ultrasound, is helpful though not routinely used by some authors. Median nerve variations are important to keep in mind particularly during endoscopic release of the carpal tunnel, to decrease the risk of iatrogenic injury.

Iatrogenic cases of carpal tunnel syndrome also occur in children. Batdorf et al. reported a series of 20 patients diagnosed over a 30-year period [39]. These authors recommend stepwise management, similar to adult patients, with activity modification and bracing, injections, and finally surgery for refractory cases.

Conclusion

Presented here are a number of rare causes of carpal tunnel syndrome. As described, these cases are often more complicated and difficult to diagnose. Therefore, appropriate treatment may be delayed in some cases as release of the transverse carpal ligament is not always the solution in these patients. These examples highlight the importance of a thorough history (including occupational, social, and travel) and detailed physical exam. In some cases, carpal tunnel syndrome may be the first manifestation of an underlying systemic condition, warranting further evaluation. Advanced imaging such as ultrasound or MRI should be considered in some cases.

A 62-year-old female patient with rheumatoid arthritis developed progressive numbness, tingling, and pain in the right thumb and index finger, 26 years following right total wrist replacement (Fig. 10.1a, b). She subsequently underwent tenosynovectomy and carpal tunnel release (Fig. 10.1c). Severe compression and flattening of the median nerve were observed intraoperatively (Fig. 10.1d). A significant amount of tenosynovitis and foreign body reaction due to metal wear was present (Fig. 10.1e–i). The patient went on to wrist fusion less than 1 year later.

A 45-year-old female with a history of Hodgkin's disease and long-standing carpal tunnel symptoms presented to the emergency department with acutely worsening, severe right wrist pain several days following an EMG. Physical exam and EMG findings were consistent with carpal tunnel syndrome. X-rays were unremarkable (Figure 10.2a). An ultrasound was also performed which showed no fluid collection. There was concern for evolving CRPS; therefore, thorough evaluation was performed including CT and MRI. Both studies showed abnormal calcification along the volar wrist (Fig. 10.2b–d, e–g).

The decision was made to proceed with debridement and carpal tunnel release. Intraoperatively, a significant amount of fluid was found surrounding the FPL tendon sheath and underneath the nerve. Calcification was also present along the FPL tendon sheath (Fig. 10.2h–k). The area was carefully debrided and specimens were sent for pathology and cultures. The pathology report showed calcifying synovitis. Cultures were negative. The patient's symptoms resolved by 3 months postoperatively.

A 48-year-old right-hand dominant female from Iowa with a history of Sjogren's syndrome and interstitial lung disease presented with a 5–6-month history of right wrist swelling and paresthesias. She described constant numbness and

tingling along the radial aspect of the hand, exacerbated by activities such as driving, typing, or holding a phone. She also reported weakness of the right hand. The patient had been previously treated with splinting and a carpal tunnel corticosteroid injection 2 months prior to presentation. The injection provided relief for approximately 1 month; however, her symptoms returned.

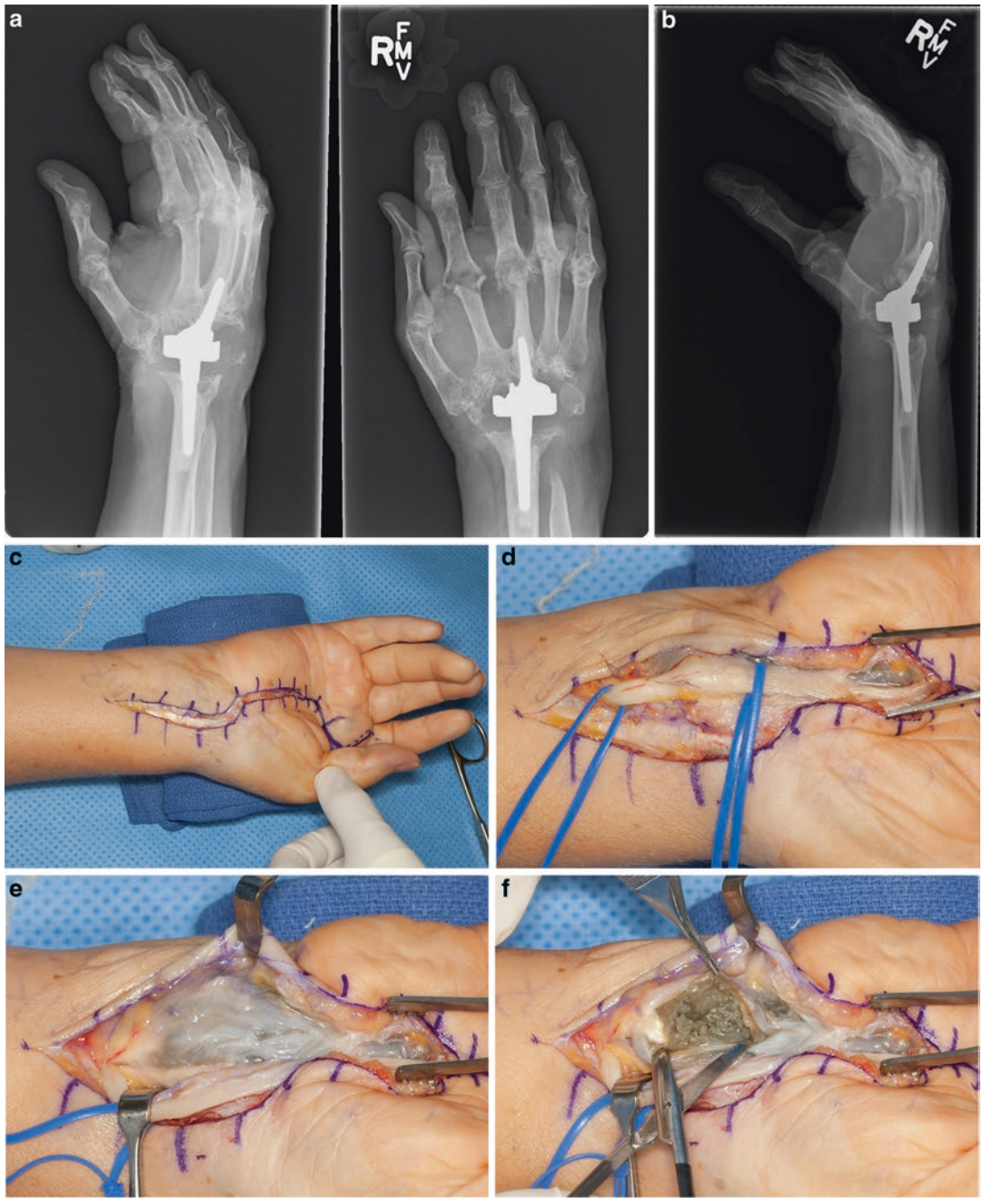

Fig. 10.1 (**a**–**j**) Foreign body reaction

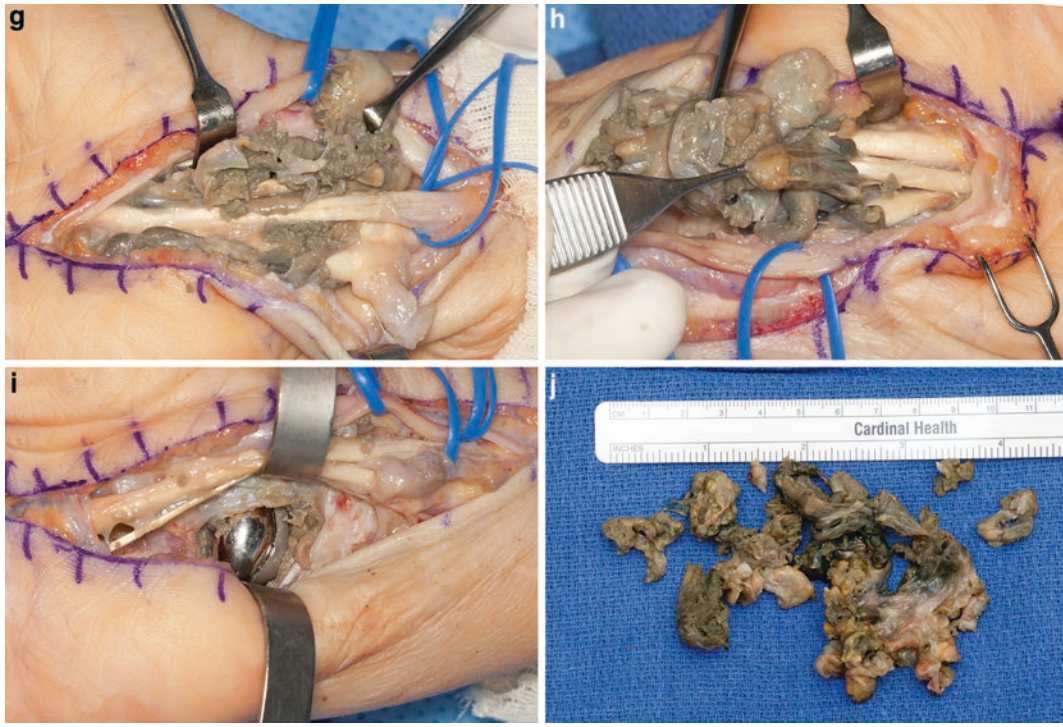

Fig. 10.1 (continued)

On physical examination, the patient had significant swelling along the volar wrist, extending through the palm and into the thumb. Carpal tunnel compression test was positive on the right and negative on Tinel's sign bilaterally. She was found to have weakness of thumb opposition (2.3 kg on the right, 4.6 kg on the left), as well as grip (4 kg on the right, 20 kg on the left). Two-point discrimination was 4–5 mm throughout all digits. Range of motion of the wrists was symmetric. EMG findings were consistent with carpal tunnel syndrome. X-rays were unremarkable (Fig. 10.3a, b). MRI showed extensive tenosynovitis (Fig. 10.3c–f).

The decision was made to proceed with carpal tunnel release and tenosynovectomy (Fig. 10.3g–i). Pathology and culture specimens were obtained at the time of surgery (Fig. 10.3j–k). The pathology report showed "non-necrotizing granulomatous inflammation of the right wrist flexor tenosynovium and FPL tenosynovium." Initial culture results were negative. However, 2 weeks later cultures revealed *Histoplasma capsulatum*. Therefore, the patient was subsequently admitted to the hospital for intravenous antifungals. After consultation with our infectious disease colleagues, the patient was treated with intravenous AmBisome for 2 weeks, followed by oral itraconazole for 3 months.

Fortunately, the patient's symptoms improved over time. The pain, swelling, and numbness had nearly completely resolved at the most recent follow-up visit. This case illustrates the importance of careful evaluation and consideration of these unusual conditions. A detailed occupational, travel, and social history is also essential.

A 13-year-old female was involved in an ATV accident and sustained distal radius/ulna fractures. She presented with numbness in the median nerve distribution. Initial X-rays showed 100% displacement and 80° of dorsal angulation (Fig. 10.4a, b). Closed reduction was performed in the emergency department (Fig. 10.4c, d). The decision was made to proceed with operative fixation. Intraoperatively, the median nerve was visualized and found to be tented and under a significant amount of stretch (Fig. 10.4e, f). The nerve was carefully dissected from its interposition between the radius and ulna fracture fragments. ORIF of the fractures was then performed (Fig. 10.4g, h).

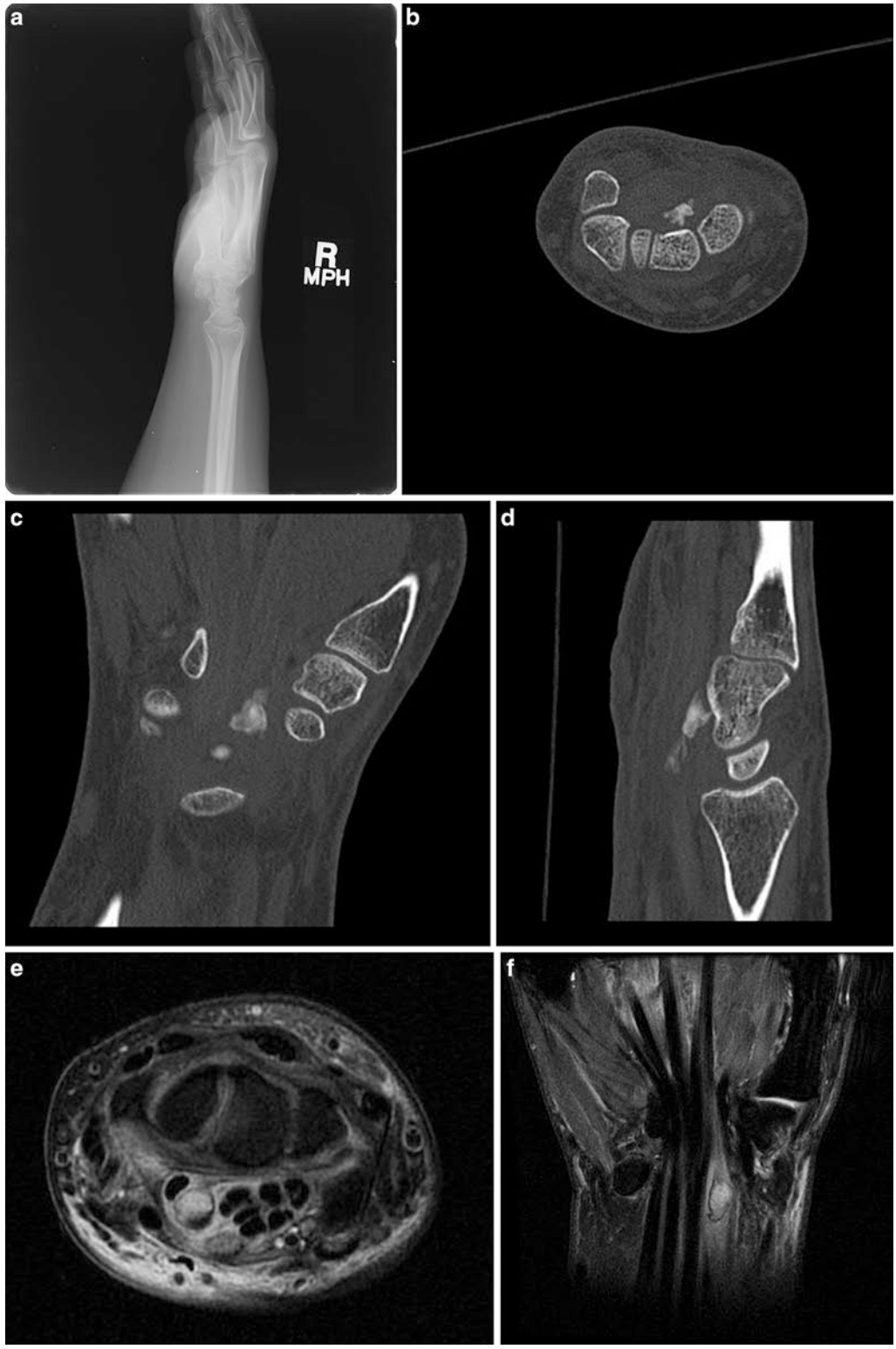

Fig. 10.2 (**a**–**k**) Calcific tendinitis

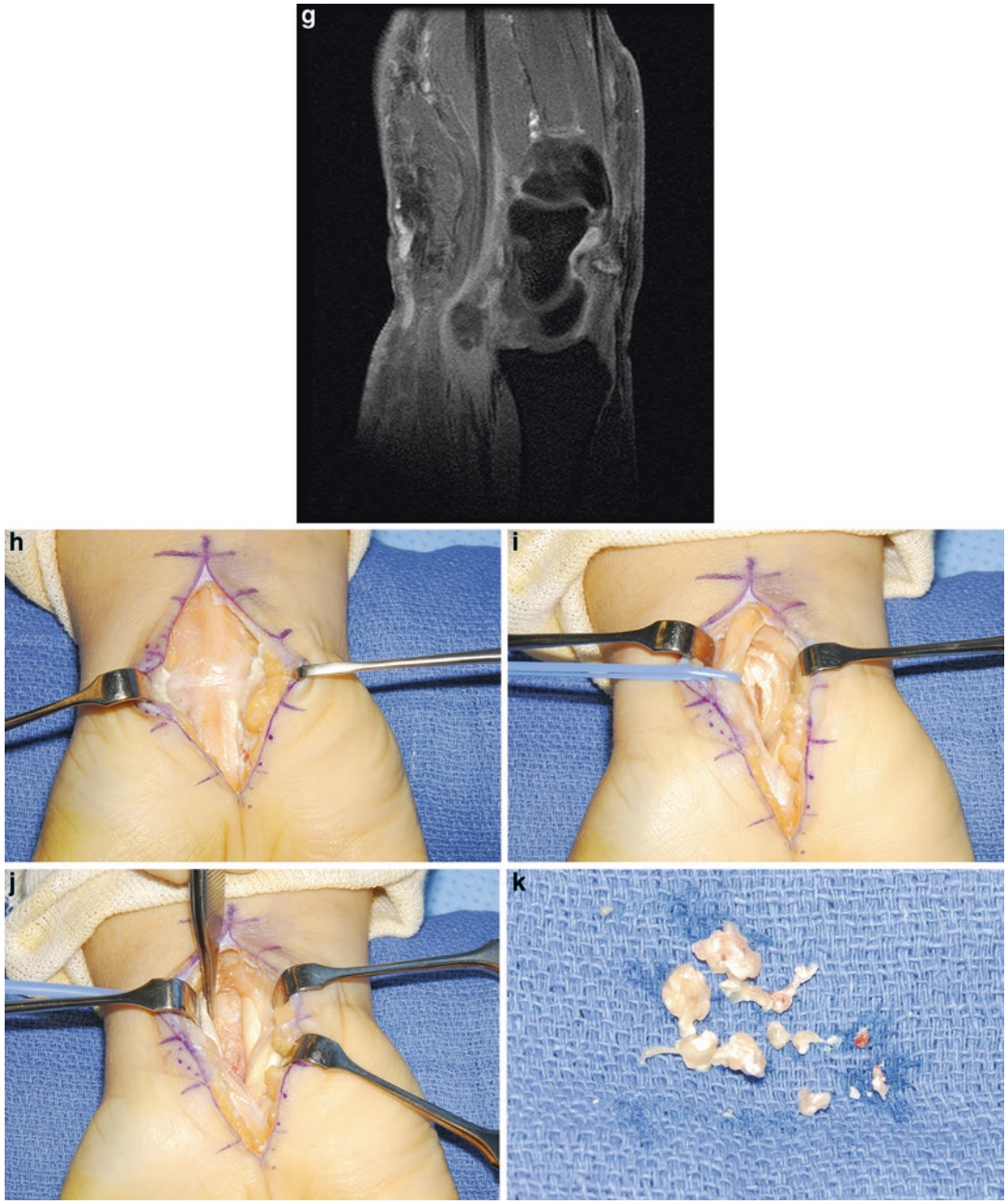

Fig. 10.2 (continued)

The fractures went on to heal uneventfully (Fig. 10.4i, j). The patient's nerve function gradually improved over time. At the time of the last follow-up (2 years from injury), two-point discrimination was 7 mm in the thumb and 5 mm in the other digits bilaterally. Tinel's sign over the median nerve was negative. She was able to make a full fist and had symmetric wrist flexion/extension as well as pronation/supination.

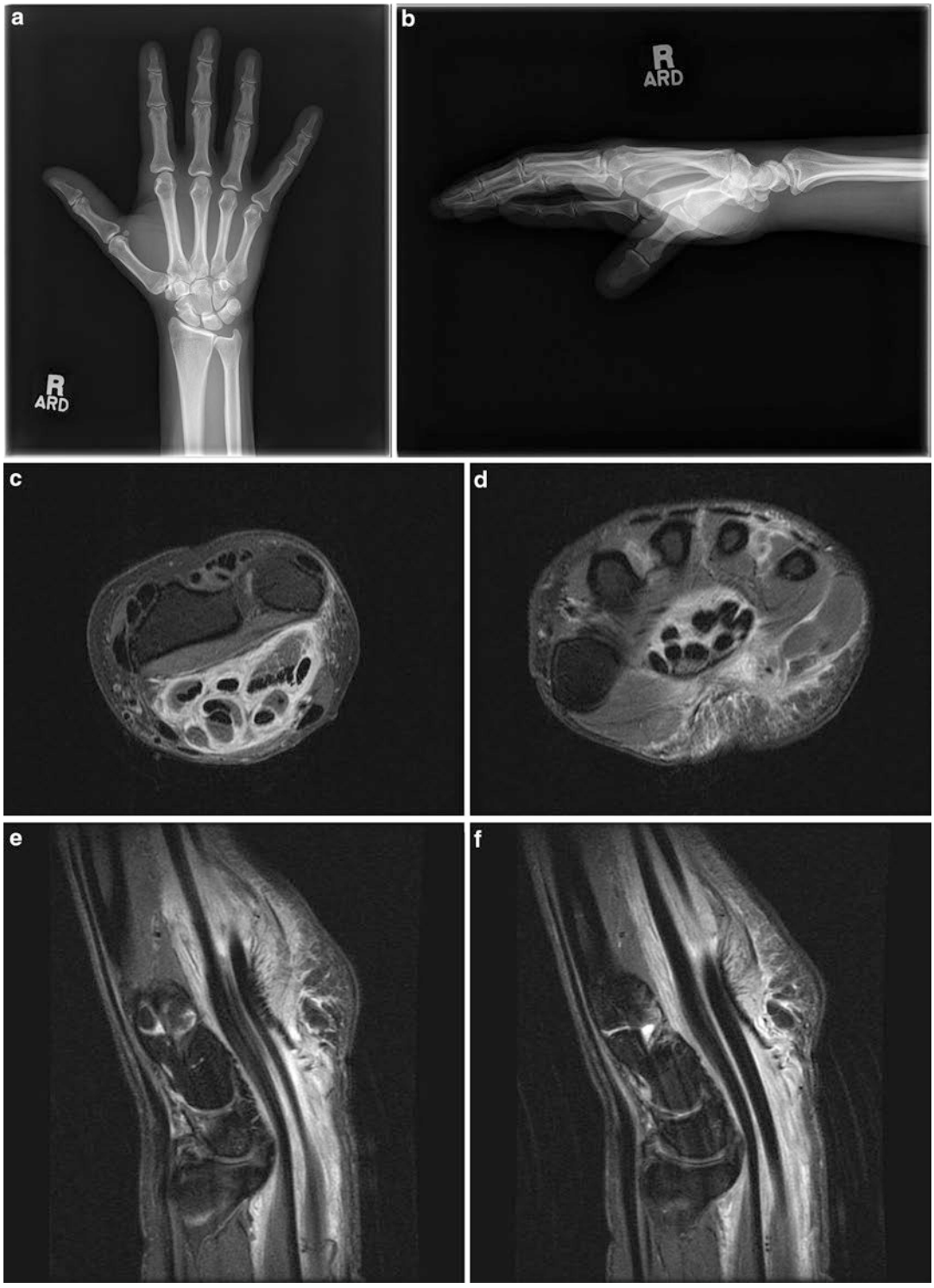

Fig. 10.3 (**a**–**k**) Histoplasma capsulatum

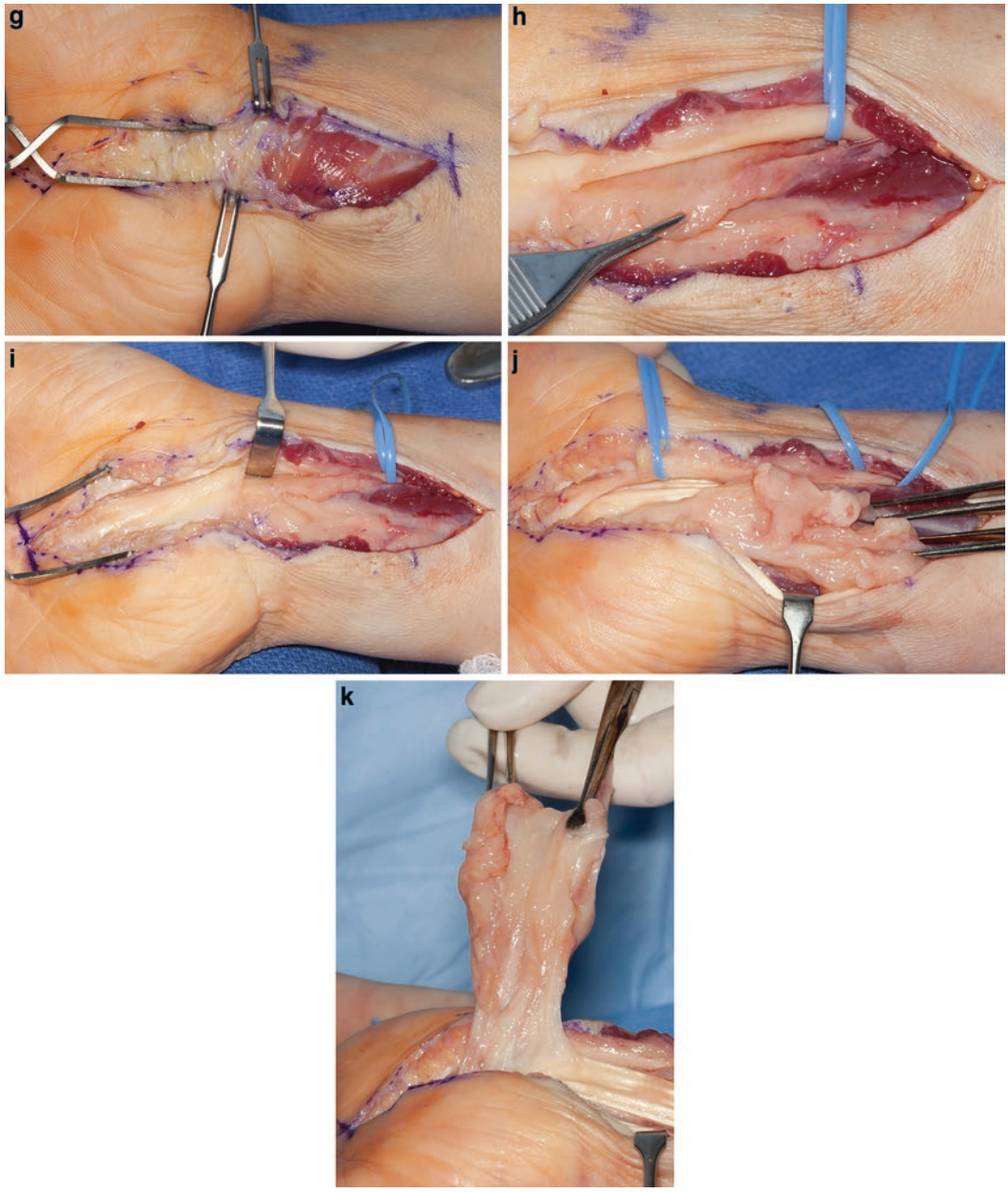

Fig. 10.3 (continued)

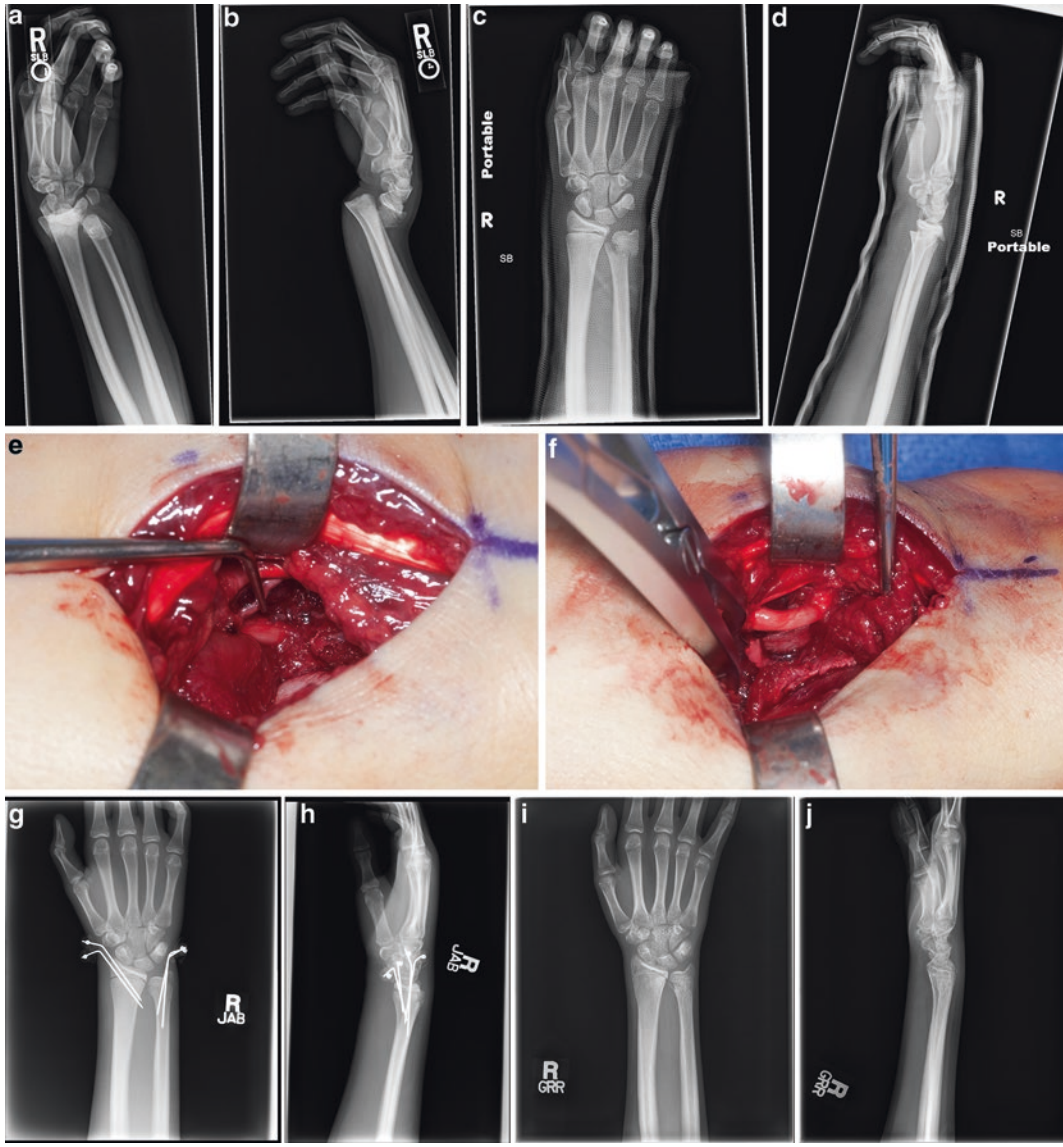

Fig. 10.4 (**a**–**j**) Trauma

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