Complex regional pain syndrome of the right wrist – Type I (CRPS-I)

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Abstract Complex regional pain syndrome (CRPS) is an intractable neuropathic painful disorder affecting one or less commonly more than one limb, disproportionate to or even without clear noxious event. It may occur without or with peripheral nerve injury, thus classifying such syndrome into type I or II, respectively. It was previously referred to as reflex sympathetic dystrophy (RSD) or Sudek’s atrophy. We had reported this case as right wrist CRPS-I in 62 year old male patient, presented with severe unexplained acute right wrist pain and periarticular distal forearm soft tissue swelling, preliminary clinically diagnosed as septic arthritis. After full radiological examinations, CRPS type I was reported after exclusion of all other similar clinico-radiological presentations. The patient was treated conservatively with successful response.

1. Introduction

Complex regional pain syndrome (CRPS) is an uncommon neuropathic painful disorder that usually involves the upper or lower limbs, less commonly other parts of the body. It is elicited as response to noxious event like trauma, fractures, or other pathologies e.g. myocardial infarction or stroke or spinal cord injury, that lead to prolonged hospitalization or immobilization. It had been reported that, it may occur in response to trivial unnoticed trauma. According to absence or presence of associated peripheral nerve injury, it is classified as type I or II, respectively (1–3).

This syndrome was formerly referred to as reflex sympathetic dystrophy (RSD) or causalgia (derived from the Greek words kausis which means burning and algos which means pain). Previously, sympathetic incrimination was agreed pathogenesis, which was supported by the relief of the pain...
by sympathetic ganglion blockade. However, recent researches showed that this syndrome is not totally mediated by such sympathetic reflex and even, the sympathetic nervous system is not necessarily involved. This was proven by a failure of sympathetic blockade to treat many other cases. Moreover, dystrophic changes are not always present (2).

CRPS has variable clinical presentation, rendering its clinical diagnosis to be difficult with high incidence of missing. Also, there is no clear published data about its incidence. The classic clinical picture, regardless it is not always present, includes the presence of an initiating noxious event (may be trivial unnoticed), followed by the development of severe disproportionate pain, soft tissue edema, local skin warmth with 1.1 °C difference from the homologous body part and abnormal motor activity in the region of interest (ROI). However, some other pathology can have some of these complaints e.g. septic arthritis, diabetic foot and fractures (2,3).

The diagnosis of CRPS is mainly based upon exclusion of these similar pathologies through imaging, which usually shows diffuse limb affection, soft tissue and patchy multifocal bone marrow edema and preserved articular surfaces. This is also, supported by the successful conservative treatment regimen including opiates and no steroidal anti-inflammatory drugs (2,3).

2. Case report

A 62 year old male patient had been presented with an acute onset of progressively increasing right wrist pain and a limited range of motion as well as periarticular soft tissue swelling and local warmth involving the distal right forearm and the hand with low grade fever. There was no clear history of trauma. Septic arthritis was the first clinical impression. The patient had been referred to radiology department- in Almana General Hospital, Hufuf, KSA- first for plain X ray, US and Doppler examination, shortly afterward, complementary non contrast CT scan of the right wrist was done to confirm X ray findings. In the next day, MRI with contrast was done.

X-ray and CT scan (Fig. 1) revealed subtle periarticular osteopenia with preserved bony articular surfaces. Incidental findings of scapholunate diastasis with proximal capitate migration (SLAC) and modest intercarpal and radiocarpal

Figure 1  (A) Right wrist plain X-ray AP (B–D) CT scan showing minimal periarticular osteopenia, radiocarpal and intercarpal DJD with marginal osteophytes, SLAC, no gross articular surfaces destruction.
osteoarthritic changes (DJD)—seen as relative narrowing of the joint spaces, subchondral sclerosis and fine marginal osteophytes—were noted as well. However, there is no articular surfaces destruction to confirm such clinical diagnosis of septic arthritis. Also, considerable diffuse periarticular soft tissue swelling was noted. Doppler US (not shown) documented uninterrupted arterial flow in the brachial, radial and ulnar arteries, thus ischemia was excluded. We had recommended MRI for further evaluation of such still unclear picture.

MRI with and without intravenous contrast of the right wrist (Figs. 2–5) was done on the second day, revealing severe diffuse periarticular hand, wrist and distal forearm circumferential-more extensive on the radial side- subcutaneous soft tissue edema (Figs. 2 and 3). Also, Scattered multiple carpal bone marrow edema, namely, the scaphoid, lunate, capitate and trapezium was present (Fig. 2). Considerable multiple flexor tendon synovial sheathes effusion (Figs. 3 and 4) was evident as well, showing avid postcontrast enhancement (Fig. 5). There was no considerable joint effusion or articular surfaces destruction or abnormal articular synovial thickening or enhancement. Also, the incidental plain X ray findings of SLAC and radiocarpal DJD were confirmed with significant

Figure 2  (A, B) Coronal STIR MRI images showing scattered multiple carpal bone marrow edema (scaphoid, lunate and capitates), periarticular radial soft tissue edema.

Figure 3  (A) Axial (B) coronal STIR MRI images showing periarticular radial soft tissue edema and flexors synovial sheath effusion.
triangular fibrocartilage (TFC) shrinkage and volume loss (Fig. 2). The sagittal sequences showed mid carpal dorsal intercalated segmental instability (DISI) (Fig. 4A).

Based on such multicompartmental non localized carpal bone marrow and distal forearm soft tissue edema as well as synovial sheathes effusion with lack of signs of the other previously mentioned differential diagnosis, CRPS-I was reported as the most suitable suggested radiological diagnosis, so, conservative treatment was applied.

3. Discussion

The published data about CRPS stated that X ray and CT scan – as we experienced in our patient- can only show periarticular osteopenia with no gross bony articular surface destruction, thus helping to exclude other types of arthritis, fractures and osteomyelitis. However, many types of arthritis or even osteomyelitis can show only such localized osteopenia in their early stages with still no articular or bony destruction. This is to say,
Plain X ray and CT scan can be helpful, but are of limited sensitivity and specificity in diagnosis of CRPS (2,3).

US can show the soft tissue swellings and the synovial sheathes or joint effusion, but still non specific findings. Studies published by Pekindil et al. (4), stated that there may be altered arterial wave forms in the region of interest as monophasic wave pattern or dropped resistive and pulsatility indices (RI&PI). The Doppler had helped us to exclude ischemia, as it showed normal arterial flow of the brachial, radial and ulnar arteries, however it showed no significant changes as regards PI and RI from the contralateral homologous sides. Other theoretical imaging tools, not widely practically applied, are thermography and 99mTc-pertechnetate scintigraphy, that are expected to show also no specific local periarticular increased temperature and radioactive uptake, respectively (2–4).

MR MRI is the stated diagnostic imaging tool of choice, with published high specificity and sensitivity scores. It clearly shows and localizes the different types of tissues involved within the affected limb; also, it helps to exclude other differential pathologies. It is reported to show patchy multifocal bone marrow edema, subcutaneous soft tissue edema, joints and synovial sheathes effusion as well as preserved articular surfaces. Also, MRI can be trusted to exclude the other differential diagnosis that may give similar or nearby clinical picture e.g. Septic arthritis, osteomyelitis, fractures, rheumatoid arthritis, diabetic foot, ischemic insults, frostbites……etc. In our patient, MRI examination had revealed similar findings to the CRPS reported signs, including diffuse subcutaneous circumferential soft tissue edema and patchy multiple carpal bone marrow edema as well as flexor tendons sheath effusion with lack of pathognomonic signs of any other differential diagnosis. So, CRPS-I was suggested as the most suitable radiological diagnosis. Also, it confirms the incidental findings of the right wrist radiocarpal DJD, SLAC and DISI (2,3).

The treatment-based on such radiological diagnosis was conservative with administration of analgesics including few times of opiates injection, non steroidal anti-inflammatory drugs and physiotherapy. Excellent pain relief and gradual swelling resolution was noted and documented by the orthopedic stuff, thus confirming the radiological diagnosis of CRPS-I (3,5).

4. Conclusion

- MRI is the imaging tool of choice for CRPS diagnosis.
- Multiple compartmental and different types of tissues affection with lack of clear causative pathology are good suggestive signs.
- The satisfactory response to conservative treatment measures strongly supports the radiological diagnosis.

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References