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Case Report

Complex regional pain syndrome type I following pacemaker implantation

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ABSTRACT

A 70-year-old woman presented with burning pain and swelling over dorsum of right hand and small joints of the fingers, associated with redness, feeling of warmth, and stiffness of the fingers, with inability to bend the fingers since 2 months. The symptoms were progressively increasing in intensity for the past 1 month. There was no history of fever or trauma to the hand. Two months before her symptoms started, she had permanent pacemaker implanted for complete heart block with syncope. She was hypertensive and was on regular medication. Her X-ray of right hand showed decreased bone density (demineralisation), suggestive of osteopenia. A diagnosis of reflex sympathetic dystrophy syndrome or complex regional pain syndrome type I induced by pacemaker insertion was made. She was treated with amitriptyline and steroids, after which her symptoms improved dramatically.

1. Introduction

Reflex sympathetic dystrophy is an uncommon entity, which usually occurs secondary to nerve injury, surgery, high impact trauma (like gun-shot injury), bone fractures, trivial soft tissue injury, brain lesions like stroke, and in some cases triggers cannot be found.[1] It is rarely seen after permanent pacemaker implantation. We report a case of a patient who developed complex regional pain syndrome (CRPS) 2 months after pacemaker insertion.

2. Case report

A 70-year-old woman developed burning pain and swelling over dorsum of right hand and small joints of the fingers, associated with redness, feeling of warmth, and stiffness of the fingers with inability to bend the fingers since 2 months. The symptoms were progressively increasing in intensity for the past 1 month. There was no history of fever or trauma to the hand that she could recall. Two months prior to the development of her symptoms, she had permanent pacemaker (VVI) inserted for complete heart block with syncope. She was hypertensive for the past ten years and was on regular medication (Losartan 50 mg BD) for the same. There was no history of other significant medical ailment in the past. On admission, she was coherent, had pulse of 60/minute, regular, and BP-140/80 mm Hg. Examination of parameters in the cardiovascular, respiratory, abdomen, and nervous systems were within normal limit. Local examination revealed erythema and swelling in the right wrist, metacarpophalangeal joints, and proximal interphalangeal joints, as well as distal interphalangeal joints (Fig. 1). The above joints were extremely

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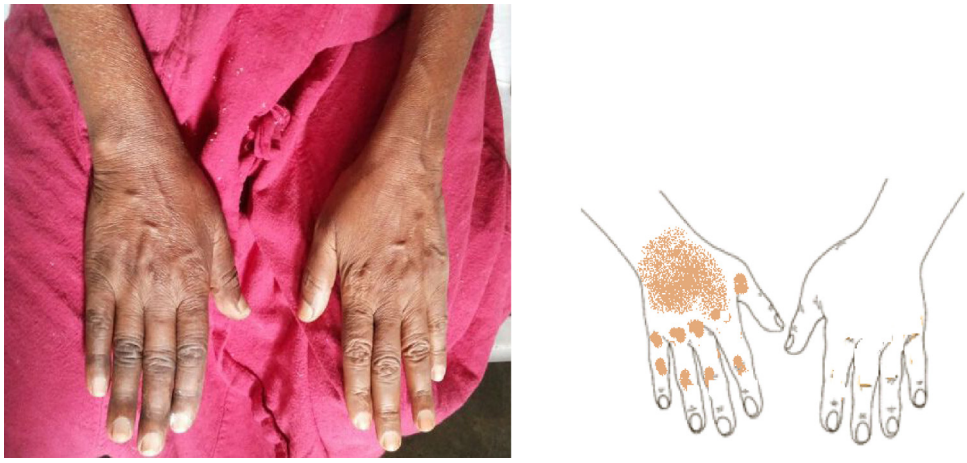

Fig. 1 – The swollen, painful right hand of early RSD with tender joints.

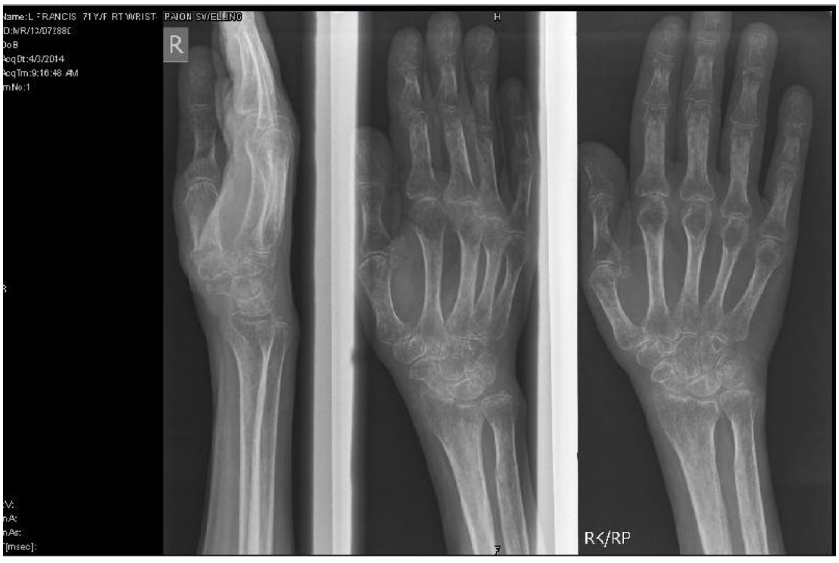

Fig. 2 – X-ray wrist joint showing osteopenia in the right hand; the joint spaces are normal. There are no erosions.

tender, and movements were restricted due to pain. All the other joints were normal. Neurological examination of the hand showed evidence of allodynia and hyperalgesia. Her blood investigations for parameters like hemoglobin, WBC count, liver and renal function tests, and fasting and postprandial blood glucose were normal. The erythrocyte sedimentation rate (Westergren) was 70 mm, at the end of 1 hour. X-ray of right hand including wrist, revealed severe demineralization (osteopenia) in the right hand, especially in the periarticular region (Fig. 2). There were no joint erosions. Her chest X-ray was normal showing the implanted pacemaker on the right (Fig. 3).

Considering the fact that pacemaker implantation done about 2 months back, blood examination being normal, the X-ray findings, which were apparent only in the involved hand, and there being no other episodes of trivial trauma or injury in the past 2 months following pacemaker insertion, which the patient could recollect, that could have triggered CRPS, diagnosis of CRPS secondary to pacemaker implantation

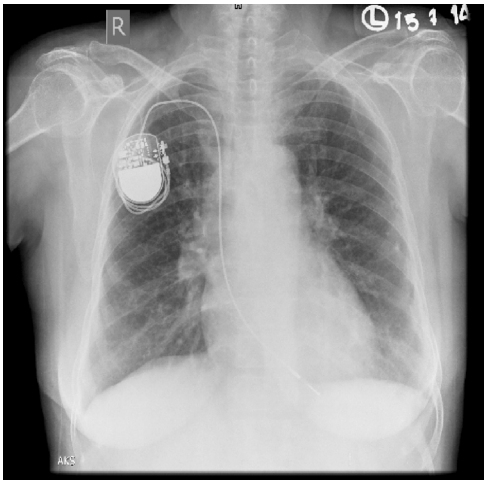

Fig. 3 – X-ray chest showing the implanted pacemaker.

was entertained. She was treated with oral amitriptyline 25 mg at bedtime, oral indomethacin 25 mg thrice a day, and physiotherapy of the hand. She, however, developed severe gastritis after two doses of oral indomethacin and it was stopped. She was started on intravenous hydrocortisone hemisuccinate 100 mg thrice a day. She responded to the above treatment, and her pain and swelling decreased significantly after one week of treatment. She was discharged on oral amitriptyline 25 mg at bedtime followed by oral prednisolone 20 mg once a day for 1 week, with gradual tapering over the next 2 weeks and was advised to continue physiotherapy. The swelling, stiffness, and pain had completely subsided after three weeks of treatment. She was asymptomatic when seen during the follow-up in the OPD.

3. Discussion

Complex regional pain syndrome (CRPS) (reflex sympathetic dystrophy syndrome [RSDS], algodystrophy, Sudeck's atrophy) is a debilitating, complex, poorly understood condition characterized by pain in a limb, in association with sensory, vasomotor, sudomotor, motor, and dystrophic changes.[1] It is often associated with limb dysfunction and psychological distress. Many different names have been ascribed to this condition and most recently the term 'complex regional pain syndrome' has been coined to emphasize the complex interaction of somatic, psychological, and behavioral factors.[2,3] The syndrome can affect anyone regardless of age or gender, however, women between the ages of 40 and 60 years are slightly more at risk. It commonly occurs after injury to the nerve either due to trauma (trivial or major) or surgery while in 9% of the cases there may not be an precipitating factor.[2,3] CRPS can be divided into two types based on the absence (type 1, much more common) or presence (type 2) of a lesion to a major nerve. The subtype of CRPS has no consequences for the general approach to management.[1] It also has been associated with various clinical conditions like diabetes mellitus and Parkinson's disease.[4]

The condition is diagnosed on the basis of clinical criteria given below.

Budapest Diagnostic Criteria[1,2]:

1. The patient must have continuing pain that is disproportionate to any inciting event.
2. The patient must report at least *one symptom* in at least *three* out of the following four categories in the affected extremity:
   - Sensory: reports of hyperesthesia;
   - Vasomotor: reports of temperature asymmetry and/or skin color changes and/or skin color asymmetry;
   - Sudomotor/edema: reports of edema (with or without joint stiffness) and/or sweating changes and/or sweating asymmetry; or
   - Motor/trophic: reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (nails, hair, skin).
3. The patient must display at least *one sign* in *two* or more of the following in the affected extremity:
   - Sensory: evidence of hyperalgesia (to pinprick) or allodynia (to light touch);
   - Vasomotor: evidence of temperature asymmetry and/or skin color changes and/or asymmetry;
   - Sudomotor/edema: objective evidence of edema (with or without joint stiffness) and/or sweating changes and/or sweating asymmetry; or
   - Motor/trophic: evidence of decreased range of motion (including joint stiffness) and/or motor dysfunction and/or trophic changes.
4. No other diagnosis can better explain the signs and symptoms.

With patients having at least three symptoms and ≥2 signs, the diagnostic accuracy is >80%. Bruehl et al. showed that the clinical diagnostic criteria for CRPS are the most effective criteria to rule or rule out CRPS across different populations.[11]

The clinical course is divided into three stages[1,2]:

Stage 1 (Acute): May last up to 3 months. The symptoms include pain, swelling, increased warmth in the affected part, excessive sweating, and limitation of movement due to pain.
Bone scan shows 'hot spot'.
Stage 2 (dystrophic): Can last from three to 12 months. Swelling is constant and skin wrinkles disappear. Pain becomes more widespread, stiffness increases, and there is hyperalgesia and allodynia. Fingernails become brittle.
Stage 3 (atrophic): Can occur from one year onwards. Atrophic changes predominate. The skin becomes dry, pale, and tightly stretched. There is stiffness and loss of joint function. X-ray of the joint shows osteopenia. These changes are now permanent. The symptoms may last for life.

Currently, no specific pathologic, histologic, or biochemical markers of this condition exist.[4–6] Although controversial, the presence of sympathetically mediated pain is an accepted etiology for many regional pain problems.

The condition is diagnosed on clinical ground. Standard X-rays typically demonstrate patchy demineralization in the affected part, which may be the result of disuse. Three-phase bone scintigraphy shows increased activity in both the blood pool and static phases. Bone scans appear to be sensitive to treatment and hence may be used for monitoring response to treatment.[2] MRI has more recently gained recognition in diagnosis, especially where radiology is contra-indicated, e.g., in pregnancy. Infra-red telethermography is reported to be sensitive and specific but is rarely used in clinical practice.[2,10]

The treatment of RSDS consists of physical therapies like active and passive range of motion exercises, transcutaneous electrical nerve stimulation, desensitization techniques, and sympathectomy.[4,9] Pharmacologic therapy consists of analgesics, antidepressants, anticonvulsants, membrane-stabilizing agents, adrenergic compounds, calcium channel blockers, corticosteroids, bisphosphonates, and newer agents like neurotropin.[4]

Till date very few cases of RSDS have been reported to occur after a pacemaker implant.[7,8] Our patient developed symptoms after 2 months of the pacemaker implantation. She had

pacemaker implanted in December 2013 and her symptoms started in February 2014. The patient described by Okada et al. had a pacemaker insertion in January 2000 and was diagnosed to have RSDS in September 2000.[7] She was treated with methylprednisolone and neurotopin.[7] Our patient responded clinically to steroid, amitriptyline, and physiotherapy within 3 weeks. She was able to completely flex her fingers and use her hand as earlier. There was no residual deformity. Prompt diagnosis and early treatment are required to avoid secondary physical problems associated with disuse of the affected limb and the psychological consequences of living with undiagnosed chronic pain. This case is presented for creating awareness of such a possibility, as early diagnosis and treatment can potentially prevent the progression of the disease, resulting in permanent disability.

Conflicts of interest

The authors have none to declare

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