IJRCI: 2020;(8)1:R1

REVIEWS

Polyarticular Charcot: A rare case report and a literature review in Indian context

Jeet Hemantkumar Patel[1],Ved Chaturvedi, Gurbir Singh Bhandari, Mayank Gupta

[1]Sir Ganga Ram Hospital, New Delhi, India

Abstract

Charcot or neuropathic arthropathies are a progressive form of destructive, erosive and generally painless arthropathies. Prevalence of neuropathic joints has decreased globally with reduction in the cases of leprosy and syphilis. However, syringomyelia and diabetes mellitus have emerged as the major causes for upper limb and lower limb Charcot joints respectively. Literature evidence shows lack of India data pertaining to these arthropathies. The present study describes a case of polyarticular Charcot in a patient with syrinx and Chiari malformation. The patient history revealed a provisional diagnosis of rheumatoid arthritis and Koch's elbow, and was treated with anti-tubercular treatment (ATT) and disease modifying anti-rheumatic drugs (DMARDS). Nervous system examination would have easily led to the diagnosis of syringomyelia. The present study also provides a review of Indian literature on neuropathic joints from 2001 to 2019. Diabetes mellitus, syringomyelia, leprosy and syphilis are major etiologies for Charcot joints.

Keywords: Charcot, arthropathy, polyarticular, Syrinx, syringomyelia

Introduction

Neuropathic arthropathy is a destructive form of progressive articular disease. It is also called neuropathic osteoarthritis or Charcot arthropathy. Jean-Martin Charcot was the first to describe arthropathies associated with tabs dorsalis in 1868.[1] Neuropathic joints have become far less common in India with the availability of more efficient treatment for DM, syphilis and leprosy.[2, 3] Moreover, recent years have witnessed a change in the prevalence of etiologies of arthropathies. In recent years, syringomyelia and DM have emerged as the major etiologies for neuropathic joints in upper and lower limbs respectively. Literature evidence from India on neuropathic joints is scarce. The present study discusses a rare case of polyarticular Charcot, which was provisionally diagnosed as rheumatoid arthritis, and the patient had undergone treatment with DMARDs and anti-tubercular therapy. The study also reviews the available Indian publications on the same.

Case report

A 48-year-old female presented with insidious onset heaviness in left arm/forearm and pain in multiple joints for 2 years. She gradually developed progressive numbness and pinprick sensations over left shoulder area, left elbow area, which gradually progressed to left forearm and left hand. She also had numbness over right hypochondrium and lower part of right breast. She also complained of multiple joints pain (both the shoulder joints, left elbow, left wrist, and left small joints of hand). She had mechanical pain in both the shoulders, knee and ankle joints. She noticed swelling in left elbow, left wrist and left small joints of hand (2nd and 3rd MCP joints and 1st CMC joints).

The patient history revealed that she had been operated earlier for left elbow swelling (synovectomy + ulnar neurolysis) and was on ATT for 4 months. The patient was treated with DMARDs (methotrexate, hydroxychloroquine), corticosteroids and non-steroidal anti-inflammatory drugs (NSAIDS). The history also revealed a wrong provisional diagnosis of rheumatoid arthritis (RA), Koch's elbow, and ulnar neuropathy.

Musculoskeletal examination revealed tenderness at bilateral shoulder joints with decreased range of motion (ROM) for overhead abduction. Swollen left elbow with decreased ROM, piano key movement at left wrist, swelling and radial deviation of 1st CMC joint left side, swollen 1st and 2nd left MCPs, and bilateral knee crepitus were other musculoskeletal findings (Fig. 1). Nervous system examination revealed hypotonia in both the upper limbs

with atrophied thenar and hypothenar muscles. There was a mild reduction in the hand grip (left more than right) and limited touch and temperature sensation over left upper limb. Sensory level was present up to T8. Biceps, triceps and supinator jerks were absent on left side. Lower limbs reflexes were normal. Ulnar nerves were palpable and thickened bilaterally.

Hemogram and biochemical blood investigations were normal, and rheumatoid factor (RF) was absent. Nerve conduction velocity testing of both upper limbs revealed distal mildly asymmetrical, large fiber predominantly motor, demyelinating axonal polyneuropathy/ polyneuoronopathy in a bilateral C5 to T1 distribution (left more than right). A moderate degree of carpal tunnel syndrome on the left side was present. X-ray of left elbow showed decreased joint space, erosions and destructive articular changes, and that of left hand showed decrease joint space, sclerosis,

Fig. 1: (A) Left swollen elbow with a surgical mark of synovectomy, (B) Left hand showing radial deviation of 1st CMC joint

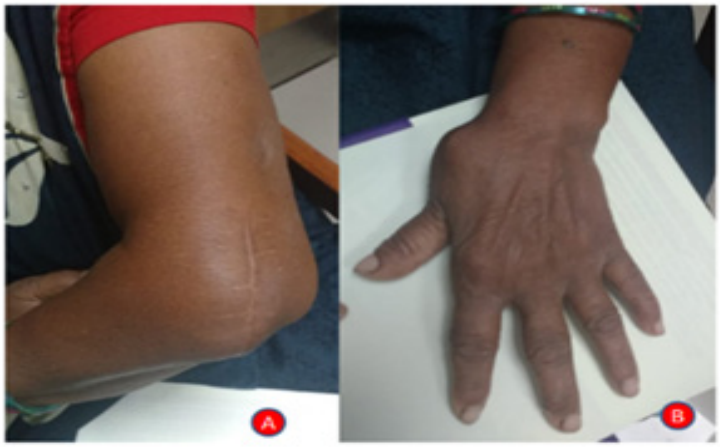


Fig. 2: (A) X-ray of knees showing changes of osteoarthritis (B) X-ray of left hand showing irregular joint margin, erosions and sclerosis at 1st CMC and radio-carpal joints, (C) X-ray of left elbow showing destroyed joint

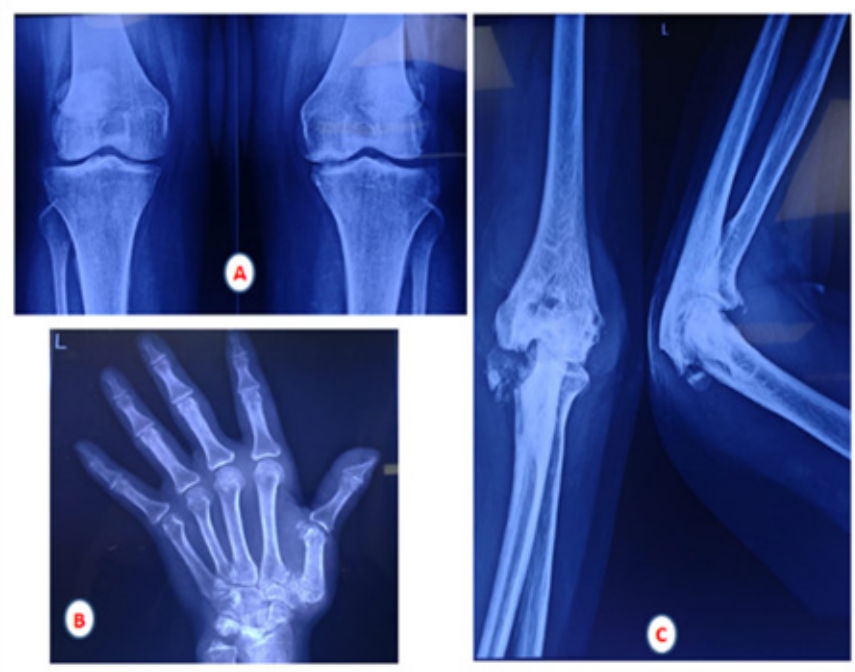


Fig. 3: Altered marrow signal intensities in humerus, radius, ulna, attached soft tissue and significant elbow joint effusion

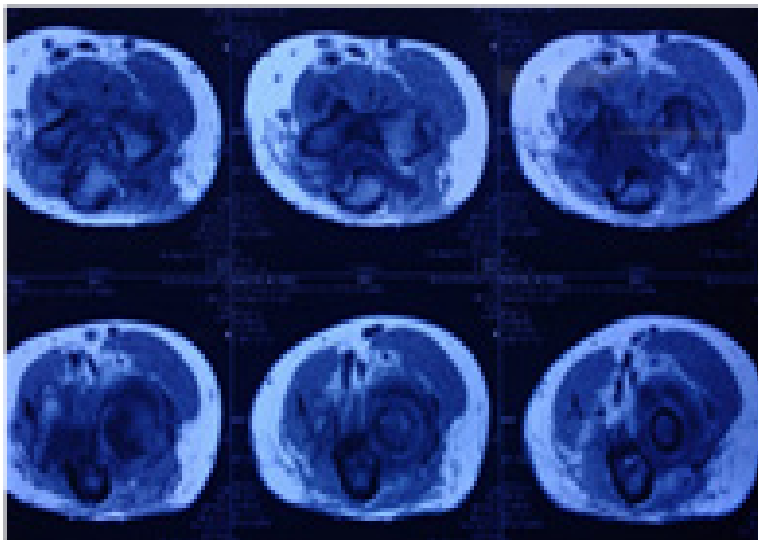
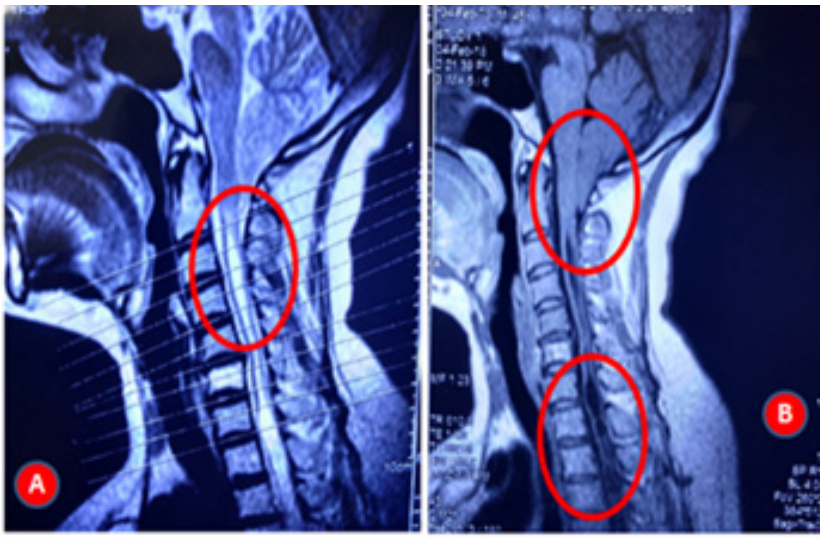


Fig. 4: (A) T2 image, (B) T1 image S/O Chiari malformation with syringomyelia of cervical-dorsal spine cord



osteophytes and degenerative changes at distal radioulnar, radiocarpal, 1st CMC, and MCP joints. X-ray of knee was suggestive of changes of osteoarthritis on both the sides (Fig. 2). MRI of left elbow revealed altered marrow signal intensities in humerus, radius, ulna, and attached soft tissue and significant elbow joint effusion (Fig. 3). MRI of cervical spine revealed syrinx involving cervical cord, extending to thoracic spine with longitudinally extended altered signal intensity changes with septae. There was also cerebellar tonsillar herniation up to C2 level. The MRI was suggestive of Chiari malformation with syrinx (Fig. 4). The patient was referred to undergo a corrective surgery for Chiari malformation.

Discussion

Neuropathic arthropathy is a minimally painful, destructive and progressive, arthropathy caused by a neurologic deficit. Common etiologies of neurologic deficit that can cause Charcot arthropathy are DM, syringomyelia, leprosy, and tabes dorsalis. Although William Musgrave, a British physician identified it for the first time in 1703, Jean-Martin Charcot, a French neurologist, first described the underlying pathology of this condition.[1]

The diagnosis of neuropathic joint is uncommon in day-to-day clinical practice, except in diabetes Charcot arthropathy. Although the data on neuropathic joints from India is

Table 1: Salient points that can be deduced from the case

1	Conducting neurological examination is important for upper limb asymmetric arthritis.
2	Mono or oligo-articular presentation is a common in syringomyelia. But this case shows that polyarticular involvement may also occur.
3	MRI of spine is helpful (not the affected joint) in proven case of syrinx.
4	Ulnar nerve may be thickened in syringomyelia, with or without neuropathic elbow joint, may mimic leprosy. But in leprosy, lower limb joints are usually affected, and not the upper limbs joints. Ulnar neurolysis is helpful to relieve symptoms due to nerve compression.
5	Syringomyelia associated neuropathic joints may mimic rheumatoid arthritis, psoriatic arthritis, tubercular arthritis, and articular manifestations of leprosy.

scarce, clinical experience shows that there is a significant decrease in the prevalence of neuropathic joint associated with syphilis and leprosy. The present study has carried out a literature review in PubMed and non-PubMed databases since 2001 using the keywords Charcot arthropathy, neuropathic arthropathy, syringomyelia, and India. Most of the publications on neuropathic joints from India were on DM and syringomyelia. The search identified 19 publications on syringomyelia leading to neuropathic joints and 7 articles, including certain reviews, on DM leading to Charcot joints (Table 2).

Several reviews on neuropathic joints have reported clinical and radiological characteristics of neuropathic joints and its pathogenesis.[16, 19, 32] The present study has tried to include all the published Indian literature on neuropathic joints. The study could not find any Indian review on syringomyelia-associated Charcot joints. One review was identified upon conducting a search for syringomyelia and neuropathic joints from countries other than India. Wang *et al.* reviewed 34 cases of syringomyelia with Charcot joints. The mean age noted was 45.21 years (ranging from 25 to 80 years) with increased female preponderance.[35] Causes of syrinx formation identified are Chiari malformation, spinal cord injury (trauma or post-spinal anesthesia), infective vertebral damage, and idiopathic syrinx. Meningitis, arachnoiditis, tethered spinal cord syndrome, spina bifida occulta and spinal cord tumors are other causes responsible for syrinx formation, albeit the current study could not find these etiologies for Indian syringomyelia cases. The management should be aimed at treating the primary etiology for syrinx, and neuropathic joint. Although there are studies reporting the usefulness of bisphosphonates like zoledronic acid and alendronate in diabetic Charcot foot, their use for managing syringomyelia-related Charcot joints have not yet evaluated.[20]

Posterior cranial fossa decompression has been identified as one of the treatment options for syrinx with Chiari malformation, no treatment is available for idiopathic syrinx. Ramnarayan *et al.* showed the beneficial effects of posterior fossa decompression with duraplasty in most of the patients with Chiari type 1 malformations.[36] Surgery is also useful for treating tethered cord syndrome, spinal tumors and post-spinal cord injury syrinx.

Syringomyelia was present in up to 25% of patients with Chiari malformation, particularly those with type 1 and Charcot arthropathy in 25% of syringomyelia cases.[37,38] Agarwal *et al.* found 63 cases of Chiari malformations. Increased male preponderance was noted and >50% of the subjects were younger patients (<20 years of age). Out of 63 cases, 46 cases were of type I and 14 cases were of type II Chiari malformations. Syringomyelia was present in 30 cases (47.62%) and was mostly associated with type I Chiari malformations.[39]

Prevalence of Charcot arthropathy in DM was 0.1 to 0.5%, and it involved tarsal, tarsometatarsal, metatarsophalangeal joints.[40] A direct association was noted between the prevalence and duration of disease. Shalini *et al.* reported that the prevalence rate of Charcot arthropathy in >50-year older diabetic patients with severe peripheral neuropathy was 9.8%. The mean age was 63 ± 8.36 years, and mean duration to develop neuropathic joints in DM was 18.01 ± 8.23 years.[7] Many surgical techniques and medical management were evaluated for usefulness in DM Charcot joint patients in India.[9, 16, 20] One Indian study by Durgia *et al.* reviewed bisphosphonates, calcitonin, and denosumab in management of acute Charcot arthropathies.[41] A systemic review by Richard *et al.* showed the effectiveness of bisphosphates in the management of acute Charcot joints, but the evidence was not strong due to insufficient data.[42] However, there is no study on the use of bisphosphonates for treating Charcot joint associated with non-diabetic causes

Table 2: Literature on neuropathic arthropathy from India since 2001

No.	Publication	Year of publication	Joints affected	Etiology
1	Choudhury *et al.*[4]	2019	shoulder	syringomyelia
2	Sebastian *et al.*[5]	2019	foot, ankle	DM
3	Karthik Yelamarthy *et al.*[6]	2018	spine	spinal cord injury
4	Salini *et al.* [7]	2018	foot, ankle	DM
5	Wakhlu et al.[8]	2017	shoulder	syringomyelia
6	Sundararajan et al. [9]	2017	foot & ankle	DM
7	Chandra et al. [10]	2016	knee	syringomyelia (post-spinal anesthesia injury)
8	Vemula et al.[11]	2016	elbow	syringomyelia
9	Singh et al.[12]	2016	1st CMC joint	idiopathic
10	Chakraborty et al.[13]	2015	shoulder	syringomyelia
11	Butala et al.[14]	2014	shoulder, 1st CMC joint	syringomyelia
12	Cps et al.[15]	2014	elbow	syringomyelia
13	Mascarenhas et al.[16]	2014	foot	DM
14	Sahoo et al. [17]	2014	elbow	syringomyelia
15	Nolkha et al. [18]	2014	shoulder	syringomyelia
16	Varma et al. [19]	2013	foot, ankle	DM
17	Bharath et al. [20]	2013	foot, ankle	DM
18	Murgai et al.[21]	2012	elbow	syringomyelia
19	Panagariya et al.[22]	2012	both shoulder	syringomyelia
20	Paliwal et al.[23]	2012	knee	syringomyelia (post-spinal anesthesia injury)
21	Kumar et al.[24]	2011	shoulder	syringomyelia
22	Panda et al. [25]	2011	shoulder	syringomyelia
23	Garg et al.[26]	2010	elbow	syringomyelia
24	Chauhan et al.[27]	2009	epiphyseal plate injury	meningomyelocele
25	Vaishya et al.[28]	2009	elbow	syringomyelia (post-tubercular)
26	Garg et al. [29]	2008	shoulder	syringomyelia
27	Raina et al.[30]	2007	knees	syphilis
28	Somalwar et al.[31]	2003	elbows	syringomyelia
29	Gupta et al.[32]	2003	foot	DM
30	Mittal et al.[33]	2002	shoulder	syringomyelia
31	Rao et al.[34]	2001	shoulder	idiopathic

like syringomyelia, leprosy etc.

Leprosy-associated Charcot joints are rare to find in daily clinical practice. Wakhlu et al. described 29 cases of rheumatological manifestations of leprosy and found only one patient with Charcot arthropathy. The current study could not find any other literature on leprosy- associated Charcot joints from India, but could identify one case of syphilis-associated disease.[30]

Conclusion

Neuropathic arthropathy is a rare cause of erosive joint disease. It may mimic Koch's joint involvement, psoriatic arthritis, Hansen's disease, rheumatoid arthritis and other forms of erosive arthritis, particularly in upper limb. Syringomyelia with or without Chiari malformations may present as upper limb asymmetric neuropathic arthropathy. Nervous system examination may assist in diagnosis, thereby to avoid unnecessary investigations, procedures and treatment. More data on the usefulness of medical agents like bisphosphonates, denosumab and calcitonin in syringomyelia Charcot arthropathy from Indian settings are needed.

Patient declaration statement

The authors certify that the patient had given her consent for images and other clinical information to be reported in the journal. The patient understood that her names and initials will not be published and due efforts will be made to conceal her identity.

Citation

Patel JH. Polyarticular charcot: a rare case report and a literature review in Indian context. IJIR. 2020;(8)1:R1

Submitted: 27 December 2019, **Accepted:** 6 February 2020, **Published:** 16 March 2020

*Correspondence: Dr. Jeet Hemantkumar Patel. Sir Ganga Ram Hospital, New Delhi, India
jeet5patel5@gmail.com

References

1. Shah, M. (2018). Charcot Arthropathy: Background, Anatomy, Pathophysiology. [online] Emedicine.medscape.com. Available at: https://emedicine.medscape.com/article/1234293-overview [Accessed 21 Jun. 2018].
2. Rao PN, Suneetha S. Current Situation of Leprosy in India and its Future Implications. Indian Dermatol Online J. 2018; 9(2): 83-89.
3. Kulkarni V, Parchure R, Darak S. Let's not let the guard down! – Early indications of syphilis resurgence? Indian J Dermatol Venereol Leprol 2019; 85: 246-7.
4. Choudhury P, Mohapatra A, Kharat A, Chauhan V. Neuropathic arthropathy of the shoulder joint caused by syringomyelia: report of a rare case. BMJ Case Rep. 2019;12(3).
5. Sebastian AP, Dasgupta R, Jebasingh F, Saravanan B, Chandy B, Mahata KM, et al. Clinical features, radiological characteristics and offloading modalities in stage 0 Acute Charcot's neuroarthropathy - A single centre experience from South India. Diabetes Metab Syndr. 2019; 13(2): 1081-1085.
6. Karthik Yelamarthy PK, Rustagi T, Mahajan R, Singh V, Das K, Bansal ML, et al. Infected charcot spine arthropathy. Spinal Cord Ser Cases. 2018; 4: 73.
7. Salini D, Harish K, Minnie P, Sundaram KR, Arun B, Sandya CJ, et al. Prevalence of Charcot Arthropathy in Type 2 Diabetes Patients Aged over 50 Years with Severe Peripheral Neuropathy: A Retrospective Study in a Tertiary Care South Indian Hospital. Indian J Endocrinol Metab. 2018; 22(1): 107-111.
8. Wakhlu A, Wakhlu A, Tandon V, Krishnani N. "Smouldering conditions of the shoulder" Lest we forget!. Indian J Rheumatol 2017; 12: 169-74.
9. Sundararajan SR, Srikanth KP, Nagaraja HS, Rajasekaran S. Effectiveness of Hindfoot Arthrodesis by Stable Internal Fixation in Various Eichenholtz Stages of Neuropathic Ankle Arthropathy. J Foot Ankle Surg. 2017; 56(2): 282-286.
10. Chandra SS, Harshavardhan JK, Ram GG, Vijayaraghavan PV. Neuropathic Knee Joint - A Complication of Syrinx Following Spinal Anesthesia: A Rare Case Report and Review of Literature. J Orthop Case Rep. 2016; 6(4): 77-79.
11. Vemula VR, Bodapati CP, Vuttarkar J, Vosuri BB. Bilateral ulnar neuropathy at the elbow secondary to Charcot-joint associated with Chiari malformation and syringomyelia. Neurol India. 2016; 64(5): 1099-100.
12. Singh M, Butala RR, Khedekar RG, Tripati M. A rare case of idiopathic thumb CMC joint neuropathic arthropathy complicated by blunt trauma. J Clin Orthop Trauma. 2016; 7(1): 138-141.
13. Chakraborty PP, Datta S, Ray S, Bhattacharjee R, Chowdhury S. Unilateral neuropathic arthropathy of the shoulder secondary to syringomyelia: Diagnostic challenges. World J Clin Cases. 2015; 3(12): 1017-20.
14. Butala RR, Arora M, Rao AA, Samant PD, Mukherjee S. A rare case of ipsilateral shoulder and thumb CMC joint neuropathic arthropathy. J Surg Case Rep. 2014; 2014(6).
15. Cps S, Pandey V, Acharya KV. Charcot's Elbow Following Syringomyelia: Revisited. JNMA J Nepal Med Assoc. 2014; 52(194): 822-4.
16. Mascarenhas JV, Jude EB. Pathogenesis and medical management of diabetic Charcot neuroarthropathy. Med Clin North Am. 2013; 97(5): 857-72.
17. Sahoo SK, Salunke P. Charcot arthropathy of the elbow joint as a presenting feature of Chiari malformation with syringomyelia. Br J Neurosurg. 2014; 28(6): 811-2.
18. Nolkha N, Srivastava D, Wakhlu A, Dhakad U. Neuropathic arthropathy and syringomyelia complicating a case of ankylosing spondylitis with peripheral arthritis. BMJ Case Rep. 2014; 2014.
19. Varma AK. Charcot neuroarthropathy of the foot and ankle: a review. J Foot Ankle Surg. 2013; 52(6): 740-9.
20. Bharath R, Bal A, Sundaram S, Unnikrishnan AG, Praveen VP, Bhavani N, et al. A comparative study of zoledronic acid and once weekly Alendronate in the management of acute Charcot arthropathy of foot in patients with diabetes mellitus. Indian J Endocrinol Metab. 2013; 17(1): 110-6.
21. Murgai A, Nair PP, Narayan S. Teaching NeuroImages: Neuropathic elbow arthropathy due to syringomyelia. Neurology. 2012; 79(12): e102.
22. Panagariya A, Sharma AK. Bilateral Charcot arthropathy of shoulder secondary to syringomyelia: An unusual case report. Ann Indian Acad Neurol. 2012;15(3): 202-4.
23. Paliwal VK, Singh P, Rahi SK, Agarwal V, Gupta RK. Charcot knee

secondary to lumbar spinal cord syringomyelia: complication of spinal anesthesia. J Clin Rheumatol. 2012; 18(4): 207-8.
24. Kumar S, Sharma V, Kumar S, Jain S. Imaging findings in Chiari I malformation with syringomyelia in a case of charcot shoulder. J Clin Imaging Sci. 2011; 1: 46.
25. Panda S, Madan VS, Sud S. Charcot's shoulder in syringomyelia. Neurol India. 2011; 59(5): 771-2.
26. Garg RK, Chaurasia RN. Charcot arthropathy of the elbow. Am J Med Sci. 2010; 340(6): 505.
27. Chauhan DK, Dhillon MS, Tripathy SK. Neuro-epiphyseal injury around the ankle: a case report. Foot (Edinb). 2009; 19(2): 133-6.
28. Vaishya R, Singh AP, Singh AP. Arthrodesis in a neuropathic elbow after posttubercular spine syrinx. J Shoulder Elbow Surg. 2009; 18(4): e13-6.
29. Garg RK, Kar AM. Charcot shoulder in syringomyelia. Intern Med J. 2008; 38(11): 868-9.
30. Raina S, Kaushal SS, Gupta D, Goyal A, Sood V. Charcot's knee joints. J Assoc Physicians India. 2007; 55: 786.
31. Somalwar AM, Sahastrabhojaney VS, Harkut P, Gautam S, Fusey SM. Extensive syrinx with Charcot's joints. J Assoc Physicians India. 2003; 51: 981.
32. Gupta PP, Mohan V. Charcot foot--an update. J Assoc Physicians India. 2003; 51: 367-72.
33. Mittal R, Gupta V. Swollen and stiff shoulder. Postgrad Med J. 2002; 78(916): 101, 108-9.
34. Rao P, Kotwal PP, Goel S. Painless destruction of the shoulder joint: a case report. Clin Rheumatol. 2001; 20(2): 143-6.
35. Wang X, Li Y, Gao J, Wang T, Li Z. Charcot arthropathy of the shoulder joint as a presenting feature of basilar impression with syringomyelia: A case report and literature review. Medicine (Baltimore). 2018; 97(28): e11391.
36. Ramnarayan R, Praharaj MS, Jayakumar PN. Chiari 1 malformations: an Indian hospital experience. Singapore Med J. 2008 Dec; 49(12): 1029-34.
37. Burina A, Smajlović D,Sinanović O, et al. arnold–chiari malformation and syringomyelia". Acta med sal 2009; 38: 44–46.
38. Riente L, Frigelli S, Sedie A. Neuropathic shoulder associated with syringomyelia and Arnold-Chiari malformation (type I). J Rheumatol 2002; 29: 638-9.
39. Agarwal S, Baruah P, Choudhury PR, et al. Study of chiari malformations in a tertiary care hospital in north-east india. Int J Anat Res 2016; 4(3): 2860-2867.
40. Rajbhandari SM, Jenkins RC, Davies C, et al. Charcot neuroarthropathy in diabetes mellitus. Diabetologia 2002; 45: 1085-96.
41. Durgia H, Sahoo J, Kamalanathan S, Palui R, Sridharan K, Raj H. Role of bisphosphonates in the management of acute Charcot foot. World J Diabetes. 2018; 9(7): 115-126.
42. Richard JL, Almasri M, Schuldiner S. Treatment of acute Charcot foot with bisphosphonates: a systematic review of the literature. Diabetologia. 2012; 55(5): 1258-64.
43. Wakhlu A, Sawlani KK, Himanshu D. Rheumatological manifestations of hansen's disease. Indian J Rheumatol 2018; 13: 14-9.