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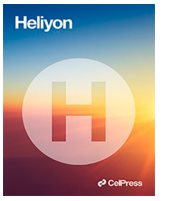

Case report

Charcot arthropathy of the knee accompanied by tethered cord syndrome and lumbosacral fur sinus

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ABSTRACT

Charcot arthropathy is a rare disease in clinic, which is easy to be misdiagnosed and delayed diagnosis. Imaging examination plays a key role in the diagnosis of Charcot arthropathy. It is important to improve the early diagnosis rate and strive for early treatment to improve the quality of life of these patients. Here we reported a rare case of charcot knee (CK) accompanied by tethered cord syndrome and lumbosacral fur sinus, who presented with joint destruction, joint deformity and multiple free bodies and received joint free bodies removal and joint replacement surgery with acceptable short and midterm follow-up results.

1. Introduction

Charcot arthropathy, also known as neurotrophic arthropathy, is a kind of arthropathy caused by central or peripheral nerve damage caused by a variety of causes, resulting in joint bone destruction, absorption, and even joint structure and function disorder. It was first discovered and reported by French doctor Charcot [1]. It is a rare disease that can occur in shoulder, spine, hip, knee, ankle and other joints [2,3]. We have herein repored a case of charcot knee (CK) accompanied by tethered cord syndrome and lumbosacral fur sinus.

2. Case presentation

A 68-year-old female patient presented at our hospital complaining of "progressive pain and deformity of the right knee with limited mobility for more than 20 years". Twenty years ago, the right knee was painful when walking, with no obvious limitation of movement and no fever. It can be relieved by plaster and analgesic treatment, but have recurrent attacks. A week ago, the pain was getting worse, the knee was swollen and deformed, and walking was obviously limited. She had an operation history of congenital spina bifida with meningocele 60 years ago, but the operation was unsuccessful. The patient had no history of heart disease, diabetes, allergy, infectious disease or blood transfusion. She had no other systemic illness. The mass was soft, well-defined and with no tenderness and few hair growth. The flexion contracture, swelling and deformity of the right knee was obvious, and several hard masses of different sizes could be reached, about 6 × 4 cm (Fig. 1A). The tenderness around the knee was obvious, and the range of extension and flexion of the right knee joint was 10°–95°. The floating patellar test (−), anterior and posterior drawer test (−), medial and lateral stress test (−). The sensation was lost below the tubercle of the right tibia. The muscle strength of both lower limbs was

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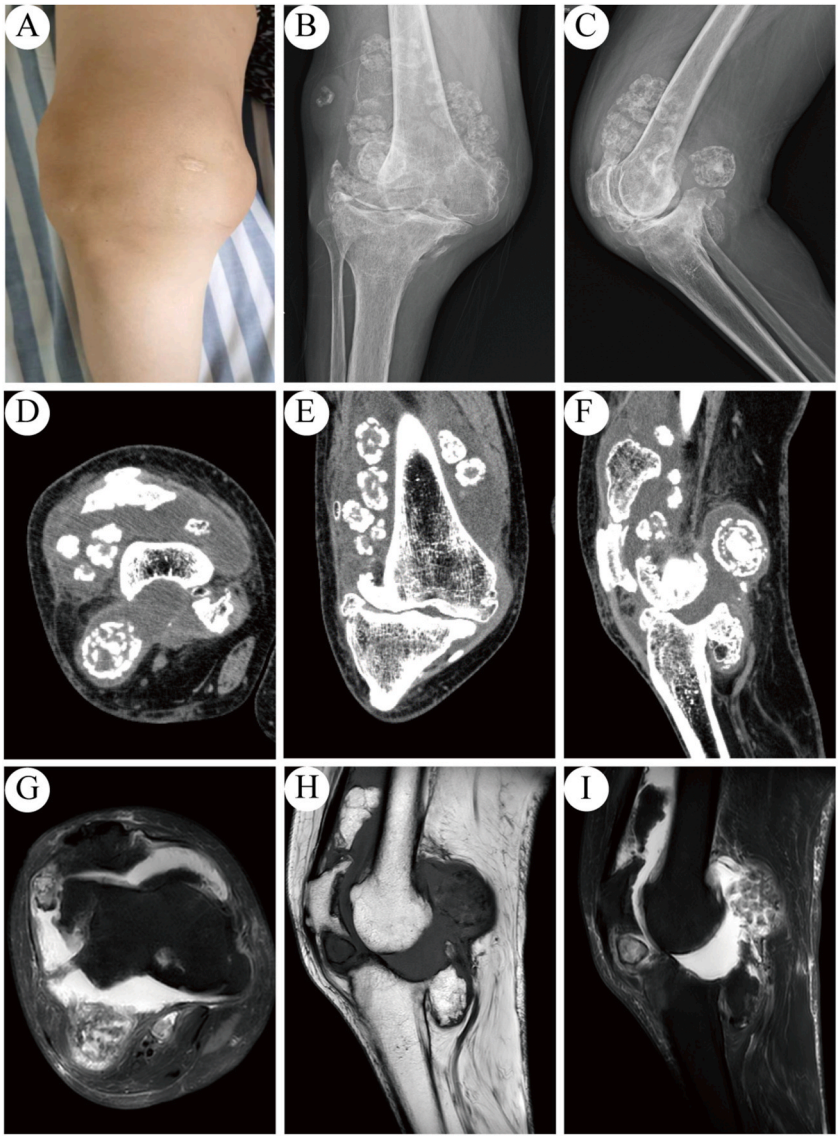

Fig. 1. Preoperative imaging of the right knee. (A) Physical examination. (B–C) Anteroposterior and lateral radiograph. (D–F) CT scan image (Tra, Cor, Sag). (G–I) MRI image (T2 Tra, T1 Sag, T2 FS Sag).

grade III +, the blood circulation and movement of both distal lower limbs were good, and there were no pathological signs. The muscles of both lower limbs were well nourished. The sensation and motor function of intestine, bladder, perineum, perianus, bilateral groin area and bilateral thighs were normal. The sensation below the tibial tubercle of the left knee joint was missing, the left knee joint has no redness, swelling and tenderness, and the extension and flexion activity was normal. X-ray and CT scan of the right knee showed extensive hyperosteogeny, narrow joint space, structural disorder, massive free body hyperplasia with joint deformity (Fig. 1B–F). MRI image of the right knee joint showed joint deformity, massive effusion in articular cavity with multiple free body formation, cartilage injury of patella and lower femur, meniscus and ligament injury in different degrees (Fig. 1G–I). Lumbosacral sagittal MRI showed low position of spinal cord cone, widening of lumbosacral spinal canal, spina bifida of L4-5 vertebra, and protrusion of sinus structure in the corresponding horizontal dorsal midline area (Fig. 2A–E). Physical examination showed a 15 × 10 cm mass in the waist and back (Fig. 2F). According to the above imaging findings, the radiologist diagnosed the patient as charcot knee, tethered cord syndrome and low back fur sinus.

The specialist approved the diagnostic conclusion and performed surgery for the patient. The free body in the joint cavity was removed, and the right knee joint rotation hinge artificial knee arthroplasty was performed. Intraoperative findings were as follows: the patellofemoral and tibiofemoral joint of the right knee were severely worn, most of the cartilage surface was stripped, and the subchondral bone was exposed. The internal and external condyles of the femur were flat and broad, and the fracture of the posterior tibia collapsed was about 1.5cm. The formation of osteophyte was obvious, there was much yellowish effusion in the joint cavity, and

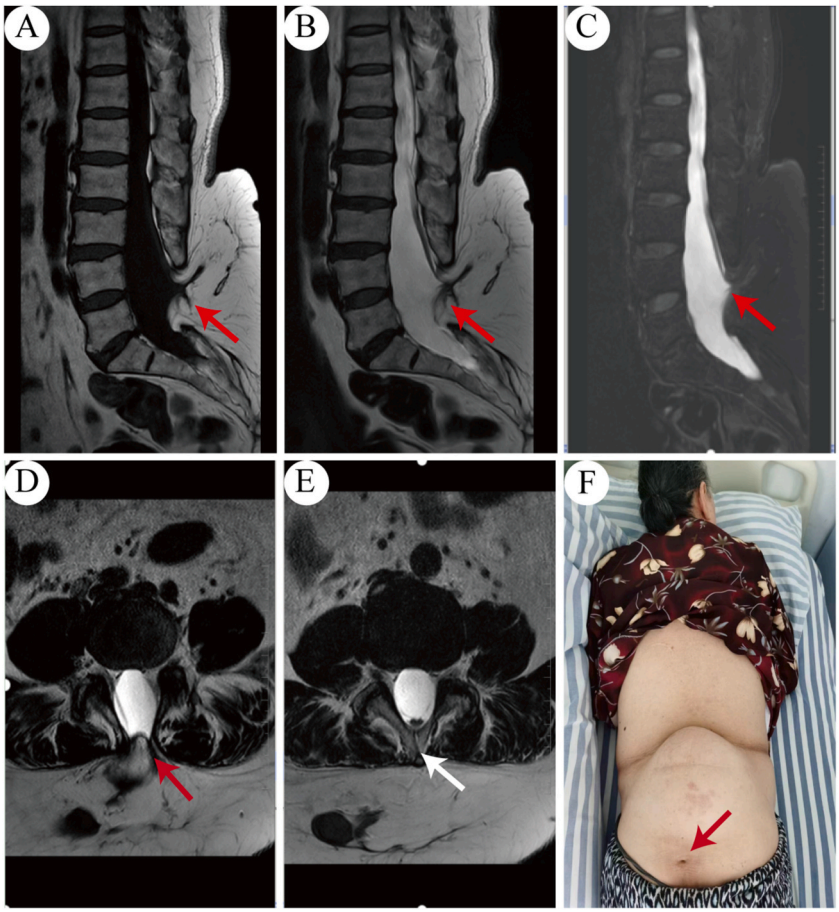

Fig. 2. Tethered cord syndrome and Lumbosacral fur sinuses. (A–E) MRI imaging (T1 Sag, T2 Sag, T2 FS Sag, T2 tra). (F) Physical examination of the lumbosacral fur sinus.

the anterior and posterior cruciate ligament was missing. Multiple free bodies of varying sizes in the joint cavity (Fig. 3A–B). The postoperative pathological findings were consistent with the characteristics of neuroarthropathy. Hematoxylin and eosin staining identified that the cartilage covered with thin fibrous tissue was partially calcified, and no other specific pathological abnormalities were found (Fig. 3C). The operation was successful, and the comparison of preoperative and postoperative X-rays was shown in Fig. 3 (Fig. 3D–G). After 3 months of follow-up, the patient's joint stability and function were significantly improved without any complications.

3. Discussion

The primary mechanism of Charcot's knee arthropathy is not clear. The initial changes may be hyperemia and active bone resorption caused by sympathetic nerves that control bone blood supply, as well as circulatory abnormalities caused by secondary nerve trauma. Common causes include diabetes, syringomyelia, syphilis, spinal cord tuberculosis, spinal cord injury, leprosy, severe meningocele, intra-articular use of hormones, severe alcohol, amyloidosis and so on [4]. In addition, spinal surgery can also induce iatrogenic Charcot neuropathicarthropathy [5]. It has been reported that inflammation is particularly critical in the pathogenesis. The imbalance of OPG/RANKL/RANK signaling pathway promotes osteoclast maturation and osteoblast decay, and aggravates inflammatory reaction, which is one of the important mechanisms leading to Charcot's joint [6]. This patient has a history of congenital spina bifida meningocele complicated with tethered cord syndrome and fur sinus, so the etiology was considered to be a combination of factors. It has been reported that there is no obvious pain in Charcot's arthropathy, and usually, the joint damage is very serious when patients see a doctor [7]. The knee pain of this patient was repeated and lasted for many years. When finally diagnosed, the disease had progressed to a late stage and could only be treated with joint replacement. Although the joint damage is serious, the conscious symptoms are relatively mild, and the serious joint damage is not commensurate with the patient's conscious symptoms, which leads to difficult early diagnosis of this disease and often leads to improper treatment and delay of treatment. Serious cases can lead to the risk of amputation. It is difficult to establish a standardized treatment strategy because of its complex and diverse etiology and individual differences in presentation and progression [8]. For patients with a clear primary disease, the treatment principle is to give priority to

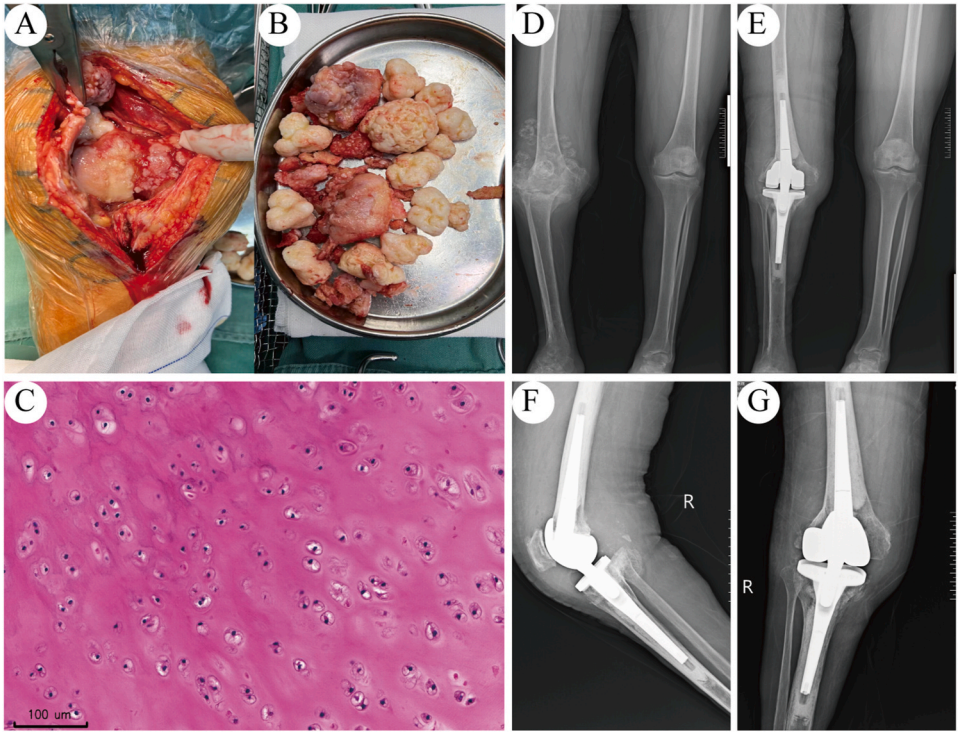

Fig. 3. Intraoperative findings and Postoperative imaging. (A–B) Large amount of synovial hyperplasia and numerous bone fragments. (C) Pathological findings: ematoxylin and eosin staining, 400× magnification. (D–E) Preoperative and postoperative comparison of X-rays of lower limbs. (F–G) Anteroposterior and lateral radiograph of the right knee postoperative.

the primary disease, and then local symptomatic treatment. It is difficult to diagnose CK in its early stage and there is no special treatment. It has been reported that the principle of distal tibial expansion osteogenesis can increase the fusion rate of CK [9]. With the continuous improvement of joint replacement technology, it is gradually used as the main treatment of CK, the complication rate is low, and the long-term prognostic effect is recognized by patients [10].

Tethered cord syndrome (TCS) refers to congenital or acquired factors that cause the spinal cord to be pulled, conical low, resulting in spinal cord ischemia, hypoxia, nerve tissue degeneration and other pathological changes, and clinically appear lower extremity sensory and motor dysfunction or deformity, bowel and urine disorders and other nerve damage syndrome. Tethered cord syndrome can occur at any age. Due to different pathological types and ages, its clinical manifestations vary. It is more common in children than in adults [11]. There are many causes of tethered cord, such as congenital spina bifida, intramural and extramural lipoma, meningomyelocele, spinal cord adhesion after lumbosacral surgery, diastematomyelia and so on. MRI is the preferred method for the diagnosis of TCS. MRI can clearly show the position of the spinal conus and the thickened terminalis. It is generally considered that the conus medullaris is lower than the lower margin of the lumbar 2 vertebral and the diameter of the terminalis $>$ 2mm is abnormal. The lipoma and fatty infiltration of the terminalis show high signals in both T1-weighted and T2-weighted images. Sagittal imaging can determine the relationship between the conus and lipoma, and MRI can also detect other abnormalities such as spina bifida, spinal division malformations, and syringomyelia [12]. Congenital fur sinuses are malformed and can occur anywhere from occipital to sacrococcygeal, most commonly in the lumbosacral region. Because the occurrence of congenital fur sinus is related to abnormal closure of neural tube, this disease is often accompanied by spinal tether, diastematic spinal cord malformation, spinal cord congenital tumor and other diseases. Common symptoms and consequences include: defecation disorder, unilateral or bilateral lower limb muscle weakness, unilateral or bilateral lower limb varus foot, high arch foot, lumbosacral or lower limb pain, numbness, decreased sensation and so on [13].

Imaging examination plays a key role in the diagnosis of Charcot arthropathy. X-ray examination can be used as a preliminary screening method to judge joint stability. CT scan can clearly show bone changes, and through three-dimensional reconstruction, the degree of bone and joint destruction can be evaluated more accurately. MRI can sensitively detect early bone abnormalities and reflect the progress of the disease, which is very helpful for the differential diagnosis of CK. Besides, PET-MRI, as well as new tools of artificial intelligence, including machine learning models, may provide some value in diagnosing, predicting joint replacement needs, and improving surgical accuracy [14]. More importantly, if there is a suspicious diagnosis of Charcot arthropathy, it is recommended that spinal cord MRI examination should be performed to evaluate whether there are spinal cord-related lesions. The diagnosis of Charcot's joint should be combined with clinical history, clinical manifestations, and imaging findings to distinguish it from other diseases such as osteoarthritis or other inflammatory arthritis, synovial osteochondroma, and chondrosarcoma. There are two important features

worth noting in the diagnosis of Charcot's arthritis: the joint destruction is much more serious than the clinical symptoms of the patients, and the affected limbs usually have neurosensory disorders [7].

4. Conclusion

Clinically, patients with myelopathy, such as tethered cord syndrome and lumbosacral cutaneous sinus, accompanied with joint destruction and multiple free bodies, regardless of whether they are associated with joint pain, swelling and deformity, the possibility of Charcot's arthropathy should be considered. It is important to improve the early diagnosis rate and strive for early treatment to improve the quality of life of these patients. In addition, clinicians should guide patients with spinal cord diseases such as tethered cord syndrome and lumbosacralfur sinus to prevent neuroarthropathy, remind patients to avoid joint overuse, and emphasize disease prevention over treatment.

Ethical approval and informed consent

The authors declare that the work described has been carried out in accordance with the Declaration of Helsinki of the World Medical Association revised in 2013 for experiments involving humans. Written informed consent was obtained from the individual for the publication of any potentially identifiable images included in this article. All data present in the manuscript are anonymised and cannot be associated to the patient.

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Data availability statement

The clinical data supporting the conclusions of this manuscript will be made available by the authors.

Additional information

No additional information is available for this paper.

CRediT authorship contribution statement

Hong Hu: Writing – review & editing, Writing – original draft, Formal analysis, Data curation, Conceptualization. **Xian Zhang:** Writing – review & editing, Writing – original draft, Formal analysis, Data curation. **Junping Li:** Writing – review & editing, Supervision, Funding acquisition, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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References

[1] R. Gupta, A short history of neuropathic arthropathy, Clin. Orthop. Relat. Res. (296) (1993) 43–49.
[2] B.F. Kocyigit, B. Kizildag, Neuropathic arthropathy of the shoulder secondary to operated syringomyelia: a case-based review, Rheumatol. Int. 43 (4) (2023) 777–790.
[3] S. Mateen, K.Y. Kwaadu, Management of midfoot charcot: minimally invasive techniques and improved fixation, Clin. Podiatr. Med. Surg. 40 (4) (2023) 593–611.
[4] T. Hester, V. Kavarthapu, Etiology, epidemiology, and outcomes of managing charcot arthropathy, Foot Ankle Clin. 27 (3) (2022) 583–594.
[5] S.H. Kim, W.J. Kim, E.S. Park, et al., Iatrogenic ankle charcot neuropathic arthropathy after spinal surgery: a case report and literature review, Medicina (Kaunas) 58 (12) (2022).
[6] T. Greco, A. Mascio, C. Comisi, et al., RANKL-RANK-OPG pathway in charcot diabetic foot: pathophysiology and clinical-therapeutic implications, Int. J. Mol. Sci. 24 (3) (2023).
[7] D. Pitocco, G. Scavone, M. Di Leo, et al., Charcot neuroarthropathy: from the laboratory to the bedside, Curr. Diabetes Rev. 16 (1) (2019) 62–72.
[8] V. Lu, J. Zhang, A. Thahir, et al., Charcot knee - presentation, diagnosis, management - a scoping review, Clin. Rheumatol. 40 (11) (2021) 4445–4456.
[9] G. Botek, S. Figas, S. Narra, Charcot neuroarthropathy advances: understanding pathogenesis and medical and surgical management, Clin. Podiatr. Med. Surg. 36 (4) (2019) 663–684.

[10] M. Zeng, J. Xie, Y. Hu, Total knee arthroplasty in patients with Charcot joints, Knee Surg. Sports Traumatol. Arthrosc. 24 (8) (2016) 2672–2677.
[11] S. Safavi-Abbasi, T.B. Mapstone, J.B. Archer, et al., History of the current understanding and management of tethered spinal cord, J. Neurosurg. Spine 25 (1) (2016) 78–87.
[12] J.C. Xie, X.D. Chen, J. Yang, Diagnosis and surgical treatment of tethered cord syndrome accompanied by congenital dermal sinus tract in adults, Beijing Da Xue Xue Bao Yi Xue Ban 54 (6) (2022) 1163–1166.
[13] G.L. Prasad, A. Hegde, S. Divya, Spinal intramedullary abscess secondary to dermal sinus in children, Eur. J. Pediatr. Surg. 29 (3) (2019) 229–238.
[14] C.L. Piccolo, C.A. Mallio, F. Vaccarino, et al., Imaging of knee osteoarthritis: a review of multimodal diagnostic approach, Quant. Imag. Med. Surg. 13 (11) (2023) 7582–7595.