Journal of Clinical Case Reports

Case Report Open Access

Atypical Diabetic Charcot Knee with Synovial Sarcoma CT Impression: A Case Report and Review of Current Literature

Bangura A*

Department of Research, Trinity School of Medicine, Ratho Mill, Saint Vincent and the Grenadines, USA

Abstract

Introduction: Diabetes mellitus has a broad range of complications, including diabetic Charcot neuroarthropathy of the knee. This complication is a destructive joint disorder by way of nerve damage and subsequent unfelt microtrauma. Another condition that can cause severe, but painless osteoarthropathy of the knee is synovial sarcoma. This type of tumour is a soft tissue neoplasm that predominantly presents within, or near large joints of the lower extremities. Synovial sarcoma can cause bone invasion, bone erosion, or both. These bone changes can then contribute to joint destruction. The overlap between the clinical features of Charcot knee and synovial sarcoma can be significant. Therefore, differentiating the two can be complicated. There have been reported cases of incidental findings of synovial sarcoma during treatment of knee arthropathy. Both synovial sarcoma and Charcot arthropathy require time-sensitive management, and a timely diagnosis may help reduce the risk of a poor prognosis.

Case presentation: We present a case of diabetic Charcot neuroarthropathy of the knee. This case was complicated by CT findings that were suspicious for synovial sarcoma. Furthermore, we reviewed current literature for Charcot knee and synovial sarcoma. This literature review included additional evaluations that should be considered when differentiating between these conditions and other joint destructive disorders. This case was handled by the Department of Orthopedics, Milton Cato Memorial Hospital, Kingstown, Saint Vincent and the Grenadines.

A 52-year-old black woman with an eighteen-year history of uncontrolled type II diabetes mellitus presented with nine months of chronic and progressive right knee swelling. On primary survey, the knee was moderately swollen with crepitus. Fluid from the knee joint was aspirated with no signs of pathology. Osteoarthritis remained the working diagnosis. A corticosteroid injection was administered, and the patient was given a knee brace and crutches with non-weight bearing instructions. Furthermore, plain film X-ray imaging was performed which revealed degenerative joint changes to the knee. On secondary survey, the knee became severely swollen. On physical examination, the right knee joint presented with significant effusion, increased warmth, and crepitus. There was mild tenderness only with full knee flexion. Both anterior and posterior draw tests were positive. In addition, the knee joint was capable of hyperabduction and hyperadduction of the lower leg, opening the joint medially and laterally respectively. Plain film X-ray imaging revealed a markedly edematous knee joint with extensive erosion to the femur and tibia. Periarticular debris and fragmentation were also noted. Given the patient's history of diabetes mellitus, diabetic Charcot neuroarthropathy was included in the differential diagnosis. A CT scan was ordered, and the impression indicated suspicion for synovial sarcoma. As a result of the CT findings, synovial sarcoma was added to the differential diagnosis. Due to the extensive destruction of the knee joint surgical intervention was indicated. The patient was scheduled for an arthrodesis with the intention of mass biopsy. During surgery, the knee joint was opened with direct visualization of the synovium. There was no mass or features of malignancy to warrant biopsy. Post-surgery, the patient was placed in a cast for ten weeks. The patient was diagnosed with diabetic Charcot neuroarthropathy of the knee, and the symptoms markedly resolved four months post operation.

Discussion: This literature illustrates prompt assessments to consider when diagnosing diabetic Charcot neuroarthropathy and synovial sarcoma of the knee. These assessments lead to quicker diagnosis, ideal treatment, and optimal patient outcomes.

Keywords: Diagnosis; Neuroarthropathy; Orthopedics; Synovial sarcoma

Introduction

Charcot neuroarthropathy of the knee

Charcot neuroarthropathy of the foot is more well-known than Charcot neuroarthropathy of the knee. However, it is possible that Charcot neuroarthropathy of the knee is just as prevalent, but poorly recognized. Although Charcot knee has a similar clinical course including painless swelling, reddening, instability, massive destruction of bone and joint, and without trauma, many patients are not properly diagnosed [1-7]. There are multiple theories about the contributing factors of Charcot neuroarthropathy, but two of the most common theories are the neurotraumatic theory and neurovascular theory. These two theories generally present simultaneously. The neurotraumatic theory is simply the loss of sensory feedback from the joint leading to repetitive microtrauma and destruction of the bone. The neurovascular theory is more complex. Its pathophysiology consists of combined autonomic, motor, and sensory neuropathy. The autonomic neuropathy contributes to bone resorption and osteopenia by unregulated and increased blood flow to the joint. Any microtrauma of the joint will include an unregulated inflammatory response, and inflammatory polypeptide such as receptor activator of nuclear factor kappa-B ligand (RANKL) will be overexpressed. RANKL increases osteoclast maturation

***Corresponding author:** Bangura A, Department of Research, Trinity School of Medicine, Ratho Mill, Saint Vincent and the Grenadines, USA, Tel: + 3017875673; E-mail: abdulaibangura@gmail.com

Received June 19, 2019; **Accepted** June 28, 2019; **Published** July 05, 2019

Citation: Bangura A (2019) Atypical Diabetic Charcot Knee with Synovial Sarcoma CT Impression: A Case Report and Review of Current Literature. J Clin Case Rep 9: 1258.

leading to greater bone resorption and subsequent osteopenia [8]. The motor neuropathy reduces joint support and can exacerbate joint instability. The sensory neuropathy is the causative factor for reduced proprioception, protective reflexes of the joint, and sensation. These three contributing factors of sensory neuropathy can induce more undetected microtrauma of the joint [9]. According to the Eichenholtz Classification of Charcot Arthropathy, our case would be considered to be in stage I, which includes osteopenia, periarticular debris and fragmentation, joint subluxation or dislocation, and/or ligamentous laxity. Stage II consists of a decrease in clinicals signs and integration of bone after immobilization. Stage III consists of bone mineralization and stabilization [10]. The non-operative management for Charcot knee consists of non-weight bearing and/or casting to prevent further bone destruction [11]. The goal is to progress the condition to Stage III with the least amount of bone loss [7]. However, in severe cases with massive bone destruction, operative management is indicated. Since the 1990s, there has been a transition from an arthrodesis to arthroplasty as the gold standard surgical operation. However, arthrodesis surgical procedures are still used today, and the patient should be treated on a case-by-case basis due to the different precipitating factors and sequelaes of the disease [12].

Synovial sarcoma

Synovial sarcoma is a slow growing soft tissue neoplasm that primarily presents intraarticularly to the large joints of the lower extremities. Its origin of cells remains unclear. Although called synovial sarcoma, the neoplasm does not derive from synovial cells. Synovial sarcoma is the most common intra-articular malignancy [13]. In more than 90% of cases, synovial sarcoma is associated with the chromosomal translocation t(X; 18) involving the SYT gene from chromosome 18 fusing with either SSX1 or the SSX2 gene on chromosome X. The SSX1 gene is more associated with the biphasic variant of synovial sarcoma, while the SSX2 gene involvement is more associated with the monophasic variant [14]. This chromosomal translocation can be tested by RT-PCR or FISH [15]. The tumor generally presents in adolescents or young adults [16]. On the contrary to other soft tissue sarcomas, synovial sarcoma presents with non-typical clinical presentations and symptoms can vary significantly. There could be variability in pain including a painless clinical presentation, and there have been definitive cases of synovial sarcoma diagnosis without identification of a mass [17]. These are just a few examples of the variability in clinical presentation that makes synovial sarcoma difficult to diagnose. Furthermore, the presentation of joint destruction from bone invasion and/or erosion can lead to a misdiagnosis [2]. The ideal radiology test for visualizing a synovial sarcoma is an MRI due to the presence of the "triple-sign," a heterogenous mass consisting of high, intermediate, and low intensities. Furthermore, there could be the presence of a mass with calcifications [18]. The most definitive diagnosis is a biopsy which will reveal one of the following three histological findings: a biphasic component of spindle and epithelioid cells, a biphasic component of spindle and glandular cells, or a monophasic component of only spindle cells [19]. Upon immunohistochemical staining, synovial sarcoma is found to stain positively for various antigens including keratin, epithelial membrane antigen (EMA), cluster of differentiation 99 (CD-99), S-100 protein, and transducin-like enhancer of split 1 (TLE1). This same tumour has been found to stain negative for CD-34. 16 The French Federation Nationale des Centres de Lutte Contre le Cancer scheme (FNCLCC scheme) is the most widely accepted grading scale for all sarcomas. The score of this scale is composed of three parameters including degree of differentiation, mitotic activity, and necrosis [20]. Synovial sarcoma management consists of tumour excision followed by chemotherapy. It has a 5-year survival rate of approximately 55% [15].

Case Report

Nine months prior to hospital admission

A 52-year-old black woman with an eighteen-year history of uncontrolled type II diabetes mellitus presented to Milton Cato Hospital with a complaint of a right knee swelling. The exact onset of symptoms was undetermined. She denied any history of trauma or injury. However, the knee swelling was accompanied with feelings of "clicks/pops" with ambulation. Upon physical examination, the right knee joint was moderately swollen with crepitus. There was mild tenderness and no signs of ligament laxity or hypermobility. Fluid from the knee joint was aspirated which reduced the knee swelling. The synovial fluid was clear and without cloudiness, purulence, or signs of infection. After aspiration, a corticosteroid injection was administered. She was placed on crutches and in a knee brace with a recommendation of non-weight bearing. Furthermore, plain film X-ray imaging was performed and revealed degenerative joint changes to the knee.

Eight months prior to hospital admission

The patient's knee swelling gradually worsened and was greater than the previous visit. However, the amount of the swelling was still considered to be moderate. She admitted to minimal sharp pain only when the knee joint is immobilized for long periods of time. The patient was more concerned with the progressive swelling of the knee. She reported skin irritation from the knee brace, so the patient was transitioned into a soft knee brace. Before leaving, fluid was again aspirated from her knee joint without concern for infection. Additionally, a second corticosteroid injection was given. The treatment plan of non-weight bearing with crutches and a knee brace was continued.

Seven months prior and leading up to hospital admission

The patient continued to be seen on a monthly basis. Her knee swelling waxed and waned and she continued to receive serial fluid aspirations. Throughout her management, a series of plain film X-ray images were taken and continued to show progressive degenerative changes of the right knee joint.

Day of admission

The patient presented with severe swelling of the right knee joint and reported instability of the knee joint. On physical examination, the right knee joint presented with significant effusion, increased warmth, and crepitus. There was mild tenderness only with full knee flexion. Both anterior and posterior draw tests were positive. In addition, the knee joint was capable of hyperabduction and hyperadduction of the lower leg, opening the joint medially and laterally respectively (Figures 1-4). Plain film X-ray imaging revealed a markedly edematous knee joint with extensive erosion to the femur and tibia. Periarticular debris and fragmentation were also noted (Figures 5-10). The patient's laboratory results were reviewed (Table 1). The patient's blood sugar level continued to be elevated with new evidence of decreased kidney function and anemia. A CT scan was ordered and reviewed. It read: "the presence of a mass, adjacent to the joint space, irregular contour with gross calcification inside, which affects the soft tissue and causes erosion and irregularity of the cortical bone of the distal femur and proximal tibia, measuring 8 × 4 cm." The CT impression was "suspicion of synovial sarcoma."

Figure 1: Lateral view of the right knee joint.

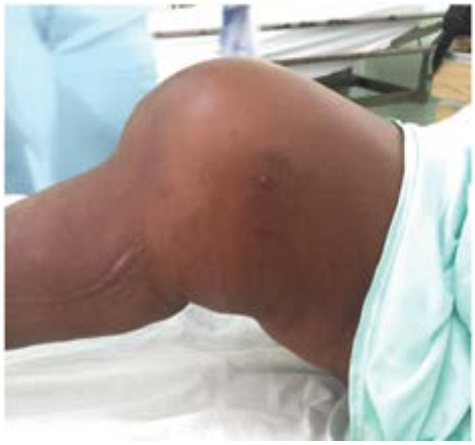

Figure 2: Medial view of the right knee joint.

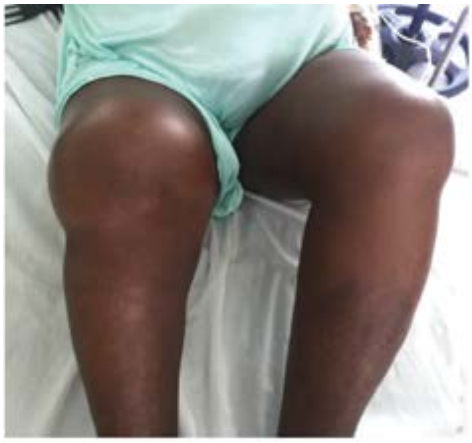

Figure 3: Anterior views of the right knee joint in comparison with left knee joint with knee flexion.

Medication list

Insulin 70/30 BID, Bendroflumethiazide (Bezide) 25 mg BID.

Past history

- Childhood illnesses: Varicella virus (chicken pox).

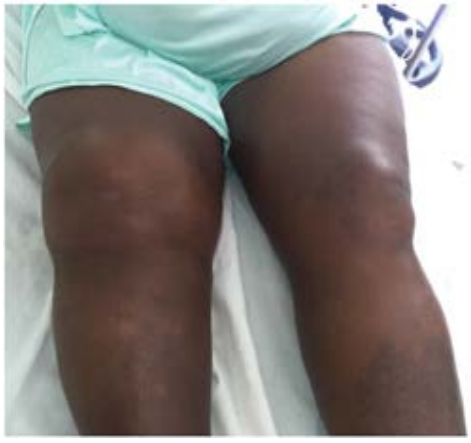

Figure 4: Anterior views of the right knee joint in comparison with left knee joint with knee extension.

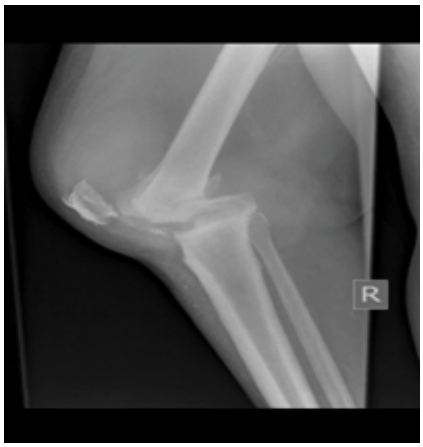

Figure 5: Lateral view x-rays of the patient right knee joint.

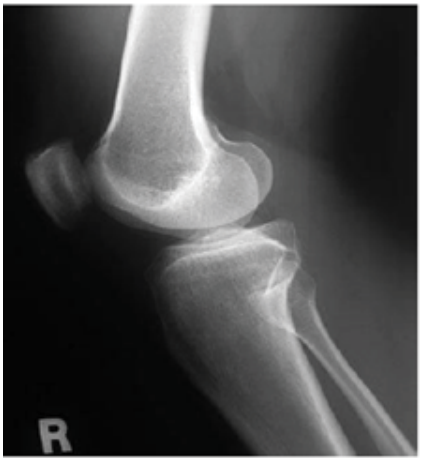

Figure 6: Lateral view x-rays of a benign right knee joint.

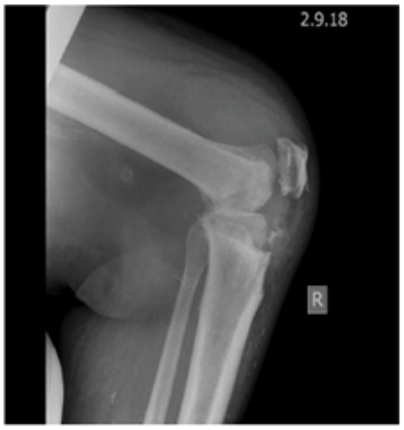

Figure 7: Lateral view with 90-degree flexion x-ray of the patient right knee joint).

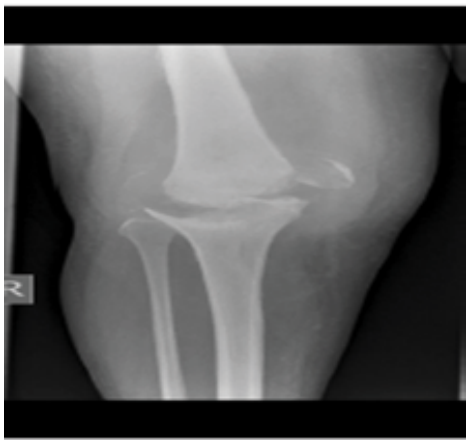

Figure 8: Medial oblique view x-rays of the patient right knee joint.

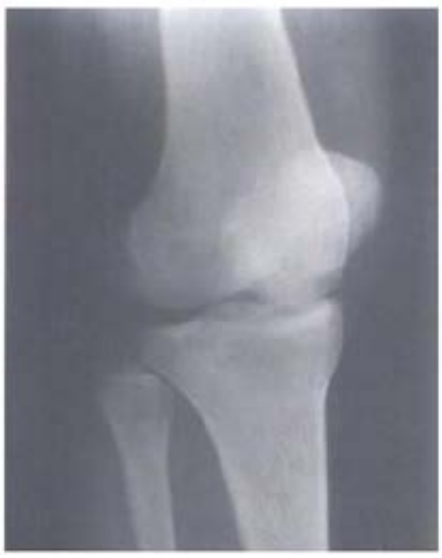

Figure 9: Medial oblique view x-rays of a benign right knee joint.

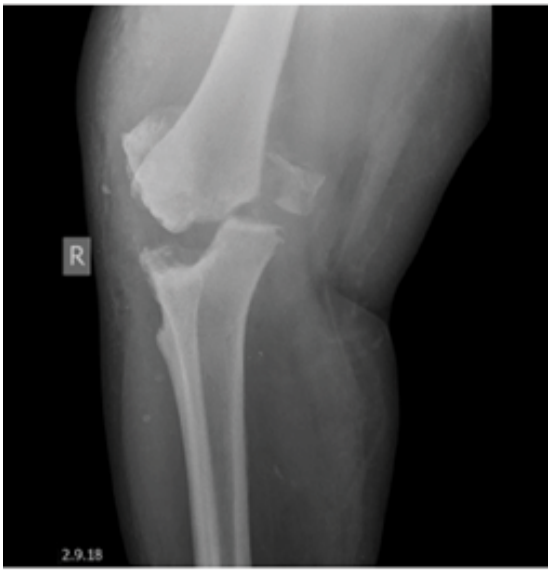

Figure 10: Additional angle exposing the extensive bone erosion and fragmentation of the right knee joint.

- Past Medical History: Diabetes mellitus II, Hypertension, Asthma.
- Allergies: NKDA
- Past Surgical History: Total hysterectomy (2005), Non-traumatic amputation of the 3-5th digits of the left foot (2013).
- Hospitalizations: Diabetes mellitus (2000), (2005), and (2013).
- Psychiatric: None.

Family history

- Mother: Heart defect, Frequent fainting.
- Cervical Carcinoma: Deceased, head injury from fainting accident, age 72.
- Maternal Uncle: Prostate Cancer age 70.
- Maternal Uncles and Aunts: DM II, HTN.
- Sister: DM II, HTN.
- Father: Deceased, traumatic accident, age 65.
- Children: No medical conditions.

Review of systems

- General: No fever.
- Skin: Pigment changes, no pallor, no cyanosis, mucous membranes pink and moist.
- Cardiovascular: No chest pain, leg swelling.
- Respiratory: Normal rise and fall of the chest, no wheezing, no cough.
- Gastrointestinal: No abdominal pain, no nausea, no vomiting, no diarrheal.
- Urinary: Polyuria, nocturia, no dysuria, no urinary incontinence.
- Musculoskeletal: Right knee swelling, right knee pain.

Test	Patient	Normal Range	Post Blood Transfusion
BMP			
Sodium	133 mEq/L		
Potassium	2.9 mEq/L		
Chloride	99 mEq/L		
CO_2	22 mEq/L	23-29 mEq/L	
BUN	13.9 mEq/L	2.2-8 mEq/L	
Creatinine	382 umol/L	44-60 umol/L	
CKD-EPI eGFR	13 L	>60 L	
CBC			
WBC	7.66 × 10^3/uL		
RBC	3.02 × 10^6/uL	4.1-5.4 × 10^3/uL	
Haemoglobin	8.7 g/dL	12-16 g/dL	9.5 g/dL
Haematocrit	26.00%	36-45%	
MCV	86.1 fL		
MCH	28.8 pg		
MCHC	32.5 g/dL		
Platelet Count	412 × 10^3/uL		
RDW-SD	50.70%		
RDW-CV	16.20%		
MPV	10.2 fL		
Random Blood Glucose Test			
BGL	269 mg/dL	<200 mg/dL	

Table 1: Laboratory results from Milton Cato Hospital during day of admission.

- Endocrinological: Polydipsia
- Neurological: Paresthesia.

Vitals

- Temperature: 98.8-degree Fahrenheit.
- Pulse: 90 bpm.
- Respiration: 24 breaths per minute.
- Blood Pressure: 160/88 mmHg.

Physical examination

- General: No acute distress.
- Skin: Diffuse skin pigmentation of the lower extremities.
- HEENT: Mallampoti score III, no retrognathia.
- Neck: Normal neck ROM.
- Respiratory: Clear to auscultation bilaterally.
- Musculoskeletal: Right knee joint with significant effusion, increased warmth, crepitus, and mild tenderness with full knee flexion, positive anterior and posterior draw, knee joint capable of hyperabduction and hyperadduction of the lower leg opening the joint medially and laterally respectively, absence of digits 3-5 of the left foot.
- Cardiovascular: Normal S1/S2, no arrhythmia.
- Neurological: RLE motor strength 2/5, LLE motor strength 4/5, BUE motor strength 5/5, Decreased peripheral pin prick sensation.

Electrocardiogram

Heart Rate 86 bpm, Normal sinus rhythm, QRS 80 ms, QT/QTcBaz 382 ms/418 ms, PR 144 ms, P 98 ms, RR/PP 720 ms/722 ms, P/QRS/T 53 ms/34 ms/51 ms.

Management

A blood transfusion was performed in an effort to treat the anemia. Due to the extensive destruction of the knee joint surgical intervention was indicated. The patient was scheduled for an arthrodesis with the intention of mass biopsy. During surgery, the knee joint was opened with direct visualization of the synovium. There was no mass or features of malignancy to warrant biopsy. However, there was evidence of chronically inflamed tissue with infiltration of bony fragments. An arthrodesis was performed. Post-surgery, the patient was placed in a cast for ten weeks. She continued to be evaluated for post operation management. The surgical wound healed appropriately and the patient's swelling markedly resolved four months post operation.

Diagnosis

Charcot neuroarthropathy of the knee.

Discussion

Diabetes mellitus has a broad range of complications, including diabetic Charcot neuroarthropathy of the knee. This complication is a destructive joint disorder by way of nerve damage and subsequent unfelt microtrauma. Charcot neuroarthropathy commonly presents at the foot of diabetic patients [8]. However, in rare cases, Charcot neuroarthropathy can present at the knee. Less than 1% of diabetics are likely to experience diabetic Charcot neuroarthropathy of the knee [21]. The complication, also known as Charcot knee, is underreported and has only been published in a limited number of literatures. Another condition that can cause severe, but painless osteoarthropathy of the knee is synovial sarcoma. This type of tumour is a soft tissue neoplasm that predominantly presents within, or near large joints of the lower extremities [1]. Synovial sarcoma can cause bone invasion, bone erosion, or both. These bone changes can then contribute to joint destruction [2]. The overlap between the clinical features of Charcot knee and synovial sarcoma can be significant. Therefore, differentiating the two can be complicated.

There have been reported cases of incidental findings of synovial sarcoma during treatment of knee Arthropathy [3-6]. Some of these cases include neuropathic osteoarthropathy identified at other locations as well. For instance, there has been an unusual case of Charcot arthropathy which presented synonymously to a synovial sarcoma at the shoulder joint. This particular patient's condition was not induced by diabetes, but instead syringomyelia. Neuropathy from syringomyelia provided a similar presentation as with long-standing diabetes.

There is a possibility that the severity of this case's presentation of Charcot knee, consisting of chronic inflammation and bone fragmentation appeared as a calcified mass on CT scan. These results could have raised the suspicion for synovial sarcoma. Charcot arthropathy and synovial sarcoma can present with clinical presentations indistinguishable to the presentations of more common conditions, such as chronic arthritis, increasing the likelihood of miser delayed diagnosis. Both synovial sarcoma and Charcot arthropathy require time-sensitive management, and a timely diagnosis may help reduce the risk of a poor prognosis.

A clinical presentation of non-painful knee swelling without history of trauma or injury should raise suspicion for Charcot arthropathy, especially if the patient is diabetic. Charcot arthropathy can present in either one of the three stages of Eichenholtz Classification of Charcot Arthropathy. The goal is to protect the patient from massive bone destruction during the earlier stages and until the patient reaches stage III which consists of bone mineralization and stabilization. In order to minimize bone destruction, microtrauma has to be decreased which can be accomplished by reducing weight bearing and providing joint stability. An earlier diagnosis leads to a better prognosis.

Conclusion

Synovial sarcomas have variable presentations including non-painful knee swelling and should be considered if the swelling is presented without history of trauma or injury. This neoplasm has been identified incidentally during the treatment of many other joint diseases. It is most prevalent in adolescents and young adults. The ideal radiology test for visualizing a synovial sarcoma is an MRI due to the presence of the "triple-sign," a heterogenous mass consisting of high, intermediate, and low intensities. Furthermore, a mass with calcifications can be present. The most definitive diagnosis is a biopsy which will reveal one of the following three histological findings: a biphasic component of spindle and epithelioid cells, a biphasic component of spindle and glandular cells, or a monophasic component of only spindle cells. Immunohistochemical staining can contribute to the diagnosis, as synovial sarcomas have been found to stain positive for various antigens including keratin, EMA, CD-99, S-100 protein, and TLE. Also, the same tumour stains negative for CD-34. The goal is to identify the neoplasm, perform surgical resection, and prevent further bone invasion or erosion. Again, an earlier diagnosis leads to a better prognosis. In conclusion, when evaluating knee osteoarthropathy without joint trauma or injury, Charcot knee and synovial sarcoma should be considered, especially for diabetics, adolescents, and young adults.

References

1. Enzinger FM, Weiss SW, Goldblum JR (2014) Enzinger and weisss soft tissue tumors, Philadelphia, PA. Elsevier Saunders.
2. Zulkarnaen M, Pan K, Shanmugam P, Ibrahim Z, Chan W (2012) Intraosseous synovial sarcoma of the proximal femur: Case report. Malays Orthop J 6: 49-52.
3. Savvidou OD, Chloros GD, Koutsouradis P, Megaloikonomos PD, Skarpidi E, et al. (2017) Synovial sarcoma complicating total knee arthroplasty. Clin Orthop Surg 9: 547-552.
4. Jamshidi K, Yahyazadeh H, Bagherifard A (2015) Unusual presentation of synovial sarcoma as meniscal cystsl: A case report. Arch Bone Joint Surg 3: 296-299.
5. Mclain R, Buckwalter J, Platz CE (1990) Synovial sarcoma of the knee. J Bone Joint Surg 72: 1092-1094.
6. Illgner U, Netten JV, Droste C, Postema K, Meiners T, et al. (2014) Diabetic charcot euroarthropathy of the knee: Conservative treatment options as alternatives to surgery. Diabetes Care 37.
7. Kucera T, Shaikh HH, Sponer P (2016) Charcot neuropathic arthropathy of the foot: A literature review and single-center experience. J Diabetes Res 1-10.
8. Babazadeh S, Stoney JD, Lim K, Choong PF (2010) Arthroplasty of a charcot knee. Orthopedic Rev 2: 65-69.
9. Rosenbaum AJ, Dipreta JA (2014) Classifications in brief: Eichenholtz classification of charcot arthropathy. Clinical Orthop Relat Res 473: 1168-1171.
10. Jacquemin C, Petitdidier N, Davidowicz K, Guignard S, Ajzenberg C, et al. (2015) Diabetic neuroarthropathy: Report of a rare case of a charcot knee. Joint Bone Spine 82: 474-475.
11. Patel A, Saini AK, Edmonds ME, Kavarthapu V (2018) Diabetic neuropathic arthropathy of the knee: Two case reports and a review of the literature. Case Rep Orthop 1-8.
12. Friedman MV, Kyriakos M, Matava MJ, Mcdonald DJ, Jennings JW, et al. (2013) Intra-articular synovial sarcoma. Skel Radiol 42: 859-867.
13. Alava E, Marcilla D, Biscuola M (2019) Round cell tumors. Pract Soft Tissue Pathol Diagnost App 233-248.
14. Folpe AL (2010) Tumors of miscellaneous type or uncertain lineage. Bone Soft Tissue Pathol 276-305.
15. Bergh P, Meis-Kindblom JM, Gherlinzoni F, Berlin R, Bacchini P, et al. (1999) Synovial sarcoma. Cancer 85: 2596-2607.
16. Chotel F, Unnithan A, Chandrasekar CR, Parot R, Jeys L, et al. (2008) Variability in the presentation of synovial sarcoma in children. J Bone Joint Surg 90: 1090-1096.
17. Heide HJ, Veth RP, Pruszczynski M, Wobbes T, Hoese QG, et al. (1998) Synovial sarcoma: Oncological and functional results. Eur J Surg Oncol (EJSO) 24: 114-119.
18. Parvizi J, Kim GK (2010) Synovial Sarcoma. High Yield Orthop 455-456.
19. Murphey MD, Gibson MS, Jennings BT, Crespo-Rodríguez AM, Fanburg-Smith J, et al. (2006) Imaging of synovial sarcoma with radiologic-pathologic correlation. RadioGraphics 26: 1543-1565.
20. Daniel A, Alegre C, Judas F, Fonseca F (2018) Diabetic charcot neuroarthropathy of the knee: A rare clinical entity. Rheumatol Adv Prac 2.
21. Gunay C, Atalar E, Ataoglu B (2014) An unusual presentation of charcot arthropathy caused by syringomyelia mimicking a soft tissue tumor. Case Rep Orthop 1-4.