Clinical Case Report

Medicine®

OPEN

A rare case of giant synovial osteochondromatosis of the thigh

A case report

Shuzhong Liu, MD[a], Xi Zhou, MD[a], An Song, MD[b], Zhen Huo, MD[c], Yipeng Wang, MD[a,*], Yong Liu, MD[a,*]

Abstract

Rationale: Giant synovial osteochondromatosis of the thigh is a highly unusual disease without standard diagnosis and curative managements so far. Our focus is to report a very rare case of giant synovial osteochondromatosis successfully operated by surgical treatment. The management of these unique cases has certain educational significance in clinical practice.

Patient concerns: A 63-year-old previously healthy man presented to our institution with a 4-year history of continuous progressive hip pain and local numbness of right side in January 2018. One month ago, the patient felt that the above symptoms were aggravated, and the right hip and proximal thigh were significantly swollen.

Diagnosis: Computed tomography and magnetic resonance imaging of the hip revealed the irregular mass in his right thigh. Postoperative pathology confirmed the diagnosis of synovial osteochondromatosis of the thigh.

Interventions: Considering the large volume of the mass and possibility of malignancy, the patient underwent surgical exploration and complete tumor resection.

Outcomes: The patient's neurological deficits and symptoms improved significantly after the surgery, and the postoperative period was uneventful at the 1-year follow-up visit. There were no complications associated with the operation during the follow-up period.

Lessons: Taken together, the lesion's clinical features, imaging results, and pathological characteristics are unique. Synovial osteochondromatosis of the thigh, although rare, should be part of the differential diagnosis when the patient presents with local pain, numbness, swelling or other symptoms. We recommend surgical treatment for the occupying lesion when the tumor has caused symptoms or neurological deficits.

Abbreviations: CT = computed tomography, MRI = magnetic resonance imaging, PET/CT = Positron Emission Tomography-Computed Tomography T1WI = T1-weighted image, T2WI = T2-weighted image, VAS = visual analogue scale.

Keywords: diagnosis, imaging characteristics, surgical treatment, synovial osteochondromatosis, thigh

1. Introduction

Synovial osteochondromatosis (SOC) is a monoarticular, synovial, proliferative disorder. It is a rare entity which presents with multiple cartilaginous nodules in synovial joints, bursae or tendon sheaths.[1–3] SOC most commonly involves knee joint with a frequency of 50% to 65%.[1,2] Other places that are involved frequently include hip, elbow, shoulder, and ankle joint.[1–3] Observed clinical symptoms were pain, swelling, numbness and limitation of joint movements at the involved area.[4,5] Secondary osteoarthritis findings such as generalized joint effusions, locking, tenderness, and snapping may also occur.[1–3] Therefore, early diagnosis and proper treatment of this

Editor: N/A.

SZL and XZ contributed equally to this work.

Written informed consent was obtained from the patient for publication of this article, a copy of which is available for review from the editors of Medicine. Because this article does not involve any human or animal trials, it did not require institutional ethical review and approval.This study was supported by National Natural Science Foundation of China (Project No. 81871746; Grant recipient: YW), and Peking Union Medical College Graduate Student Innovation Fund (2018) (Project No. 2018–1002–02–08; Grant recipient: SL). The funders had no role in study design, data collection and analysis, decision to publish, or preparation of the manuscript.The authors have no conflicts of interest to disclose.

[a] Department of Orthopedic Surgery, Peking Union Medical College Hospital, Peking Union Medical College and Chinese Academy of Medical Sciences, [b] Department of Endocrinology, Key Laboratory of Endocrinology, National Health and Family Planning Commission, [c] Department of Pathology, Peking Union Medical College Hospital, Chinese Academy of Medical Science & Peking Union Medical College, Beijing, China.

[*] Correspondence: Yong Liu, and Yipeng Wang, Department of Orthopedic Surgery, Peking Union Medical College Hospital, Peking Union Medical College and Chinese Academy of Medical Sciences, No. 1 Shuaifuyuan Wangfujing, Beijing 100730, China (e-mail: liuyong_pumch@126.com, ypwang_pumch@163.com).

How to cite this article: Liu S, Zhou X, Song A, Huo Z, Wang Y, Liu Y. A rare case of giant synovial osteochondromatosis of the thigh: A case report. Medicine 2019;98:49(e18269).

Received: 28 March 2019 / Received in final form: 26 October 2019 / Accepted: 7 November 2019

http://dx.doi.org/10.1097/MD.0000000000018269

unique disease is of great significance. Although rare, SOC should be considered in differential diagnosis of cases presenting with similar symptoms.

To the best of our knowledge, this is a less-documented case of giant synovial osteochondromatosis of the thigh in a man presenting with continuous progressive hip pain, local numbness and swelling, who underwent total resection of the space-occupying lesions. In the follow-up visit, the patient's conditions improved significantly postoperatively. After reviewing pertinent literatures, we discussed common clinical considerations in patients with giant synovial osteochondromatosis of the thigh and management considerations for these patients.

2. Case report

A 63-year-old previously healthy man presented to our institution with a 4-year history of continuous progressive hip pain and local numbness of right thigh in January 2018. Upon examining and questioning, the patient stated he has been experiencing a gradual increase in his hip pain, as well as worsening numbness and swelling of the right thigh. In the medical journal of his current illness, the pain in his right hip can reach 4–5 points using the visual analogue scale and cannot be alleviated with rest and hot compresses. One month ago, the patient felt that the above symptoms were aggravated, especially during sleeping and sitting. He denied history of injury and any other underlying diseases. No pertinent family history was identified, including hypertension and cancer.

Physical examination showed a mass sized 12×24 cm in the proximal thigh, which was hard in texture, unclear in boundary, adherence to surrounding tissues, poor mobility, and no tenderness. However, low skin temperature and varicose vein were not found on the surface of the mass. Routine laboratory studies were almost within normal range, except that the tissue polypeptide specific antigen was significantly elevated to 101.55 U/L (normal: <80 U/L). Plain radiographs showed irregular shadow of a soft tissue mass in his right thigh (Fig. 1A and B). Computed tomography (CT) showed multilocular cystic-solid mass in the right thigh root, with high suspicion of malignancy (Fig. 2A–E). Magnetic resonance imaging (MRI) of the hip revealed the irregular mass in his right thigh mimicking a parosteal sarcoma (Fig. 3A–I).

Considering the large volume of the mass and possibility of malignancy, surgical exploration and complete tumor resection were performed according to the designed surgical procedure. After successful anesthesia, the patient was placed in a supine position, with the right buttock being cushioned high. During the operation, multiple cystic masses in the deep muscles were seen, with a size of about $24 \times 15 \times 10$ cm extending from the upper middle of the thigh to the right hip (Fig. 4A–C). Each capsule contains clear yellowish and reddish liquids, which are filled with a large number of round, tough, oval-shaped, white translucent, cartilage-like granules with a diameter of about 0.5 to 2.0 cm. The chondroid granules in the capsule were cleaned completely, the wall of the capsule was separated from the surrounding tissues, and the capsule was excised completely and sent for pathological examination. The incision was closed. Intraoperative blood loss was approximately 900 mL, thus we used erythrocyte 2U. The postoperative pathology confirmed the diagnosis of synovial osteochondromatosis of the thigh (Fig. 5A–F). Pathological result was positive for Vimentin and S-100. Biopsy samples were negative for AE1/AE3 and EMA, with 5% Ki-67 positive nuclei.

One week after the operation, the patient's symptoms improved significantly compared to the preoperative status. Postoperatively, visual analogue scale score of his hip pain improved to 0–1 points compared to the preoperative status, 4–5 points. At a 1-year follow-up visit, the patient was doing well, with no local recurrence or new symptoms. There were no complications associated with the operation during the follow-up period.

3. Discussion

Synovial osteochondromatosis (SOC) is a benign disorder of nodular cartilaginous neoplastic development of the synovium that can lead to multiple loose bodies within the articular

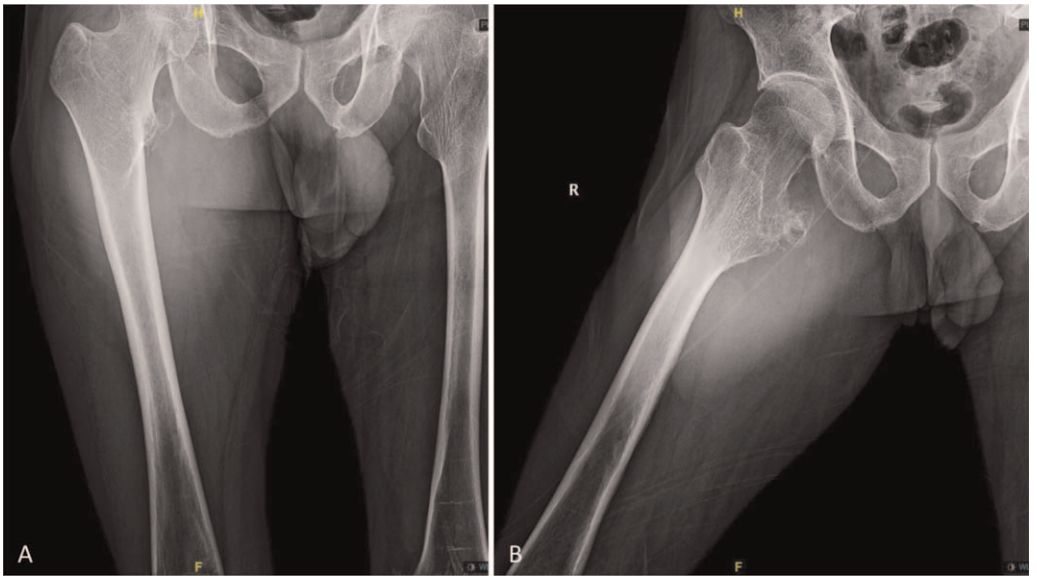

Figure 1. (A, B) Posteroanterior X-ray film of the right hip revealed irregular shadow of a soft tissue mass in the right thigh.

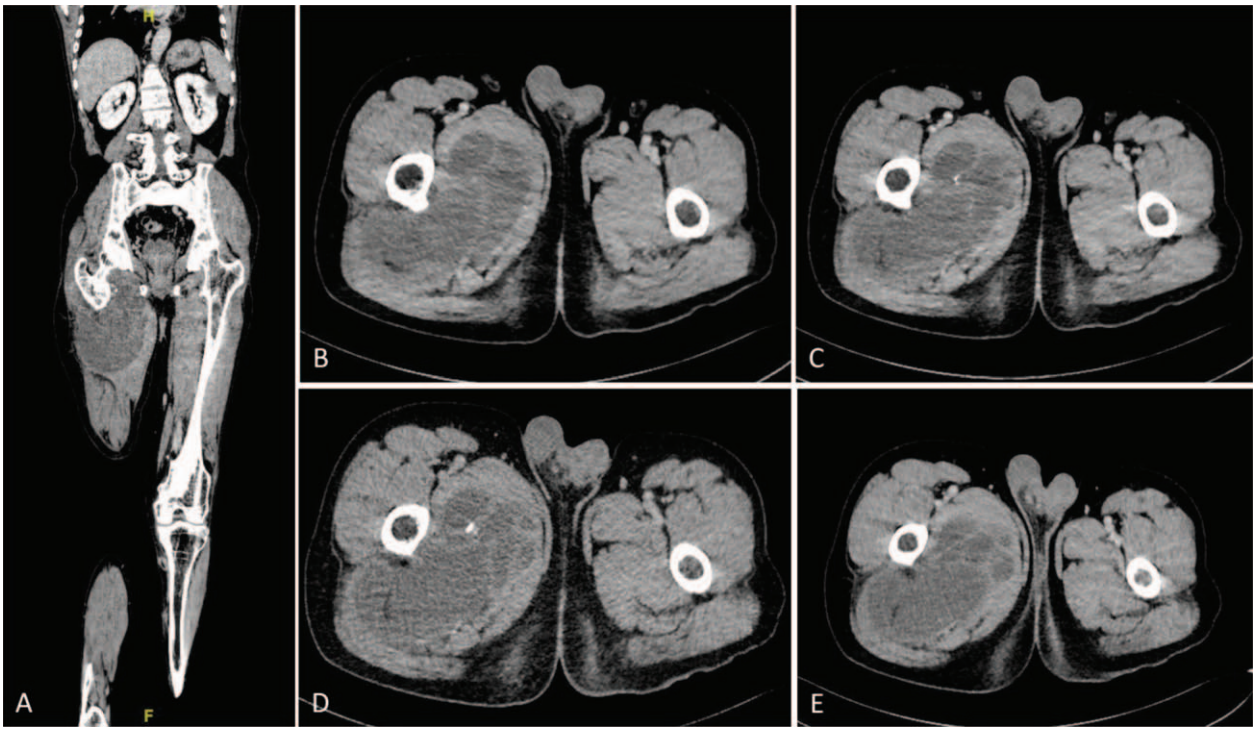

Figure 2. (A–E) Preoperative coronal and transverse CT scan revealed multilocular cystic-solid mass in the right thigh root, with high suspicion of malignancy.

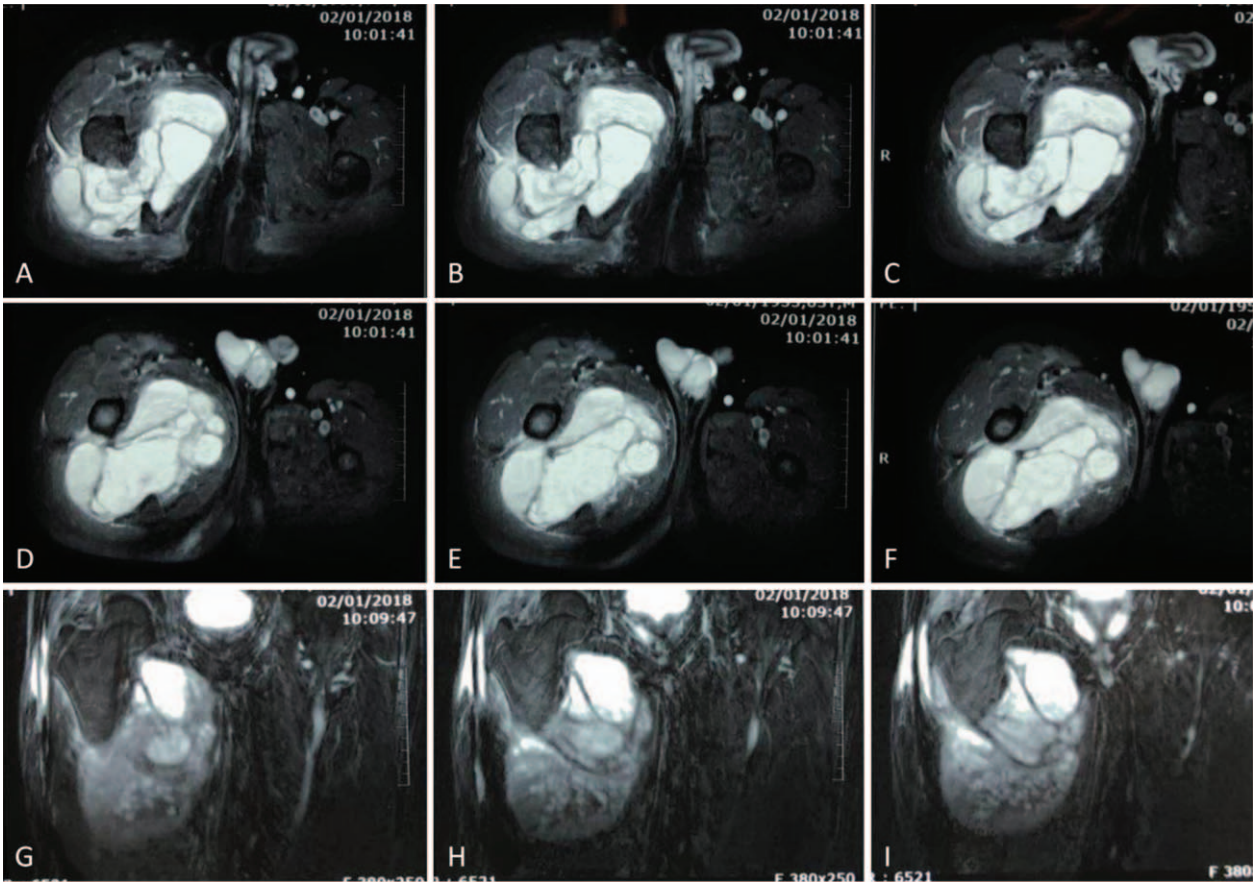

Figure 3. (A–I) Preoperative coronal and transverse MRI scan revealed the irregular mass in his right thigh.

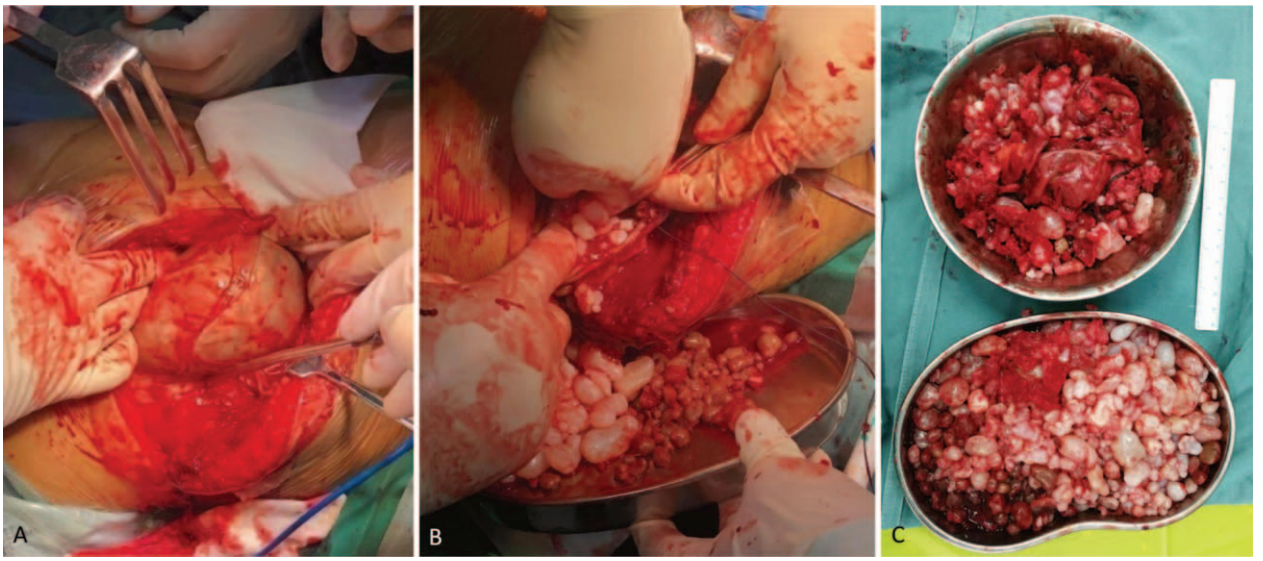

Figure 4. (A, B) Intraoperative photography depicting the exposed tumor. (C) Intraoperative photography depicting totally resected synovial osteochondromatosis.

space.[1–3] SOC can be originated from any joint, tendon sheath, or bursae that has synovial tissue, which is characterized by cartilaginous nodule formation secondary to synovial metaplasia.[1–4] Although it is generally progressive in nature, it can limit itself and regress.[5,6] This condition is usually a monoarthritic disease and affects the knee joint in more than 50% of all cases suffering from SOC.[1–3,7] To date, few reports of SOC of the thigh causing clinical symptoms have been documented so far. Therefore, the management of our reported case has certain educational significance in clinical practice. Delay in diagnosis of synovial chondromatosis may occur due to slow progression of disease course and calcification of free cartilage fragments at later stages.[8,9] Clinical manifestations of the patients with SOCs are usually due to mechanical and oppressive effects of the mass. Milgram defined 3 stages of SOC: Stage 1 (early stage) is the active intrasynovial stage during which there is no free articular body. Stage II (mid-term stage) is the transition stage from intrasynovial lesions to free bodies. In this stage, there are both

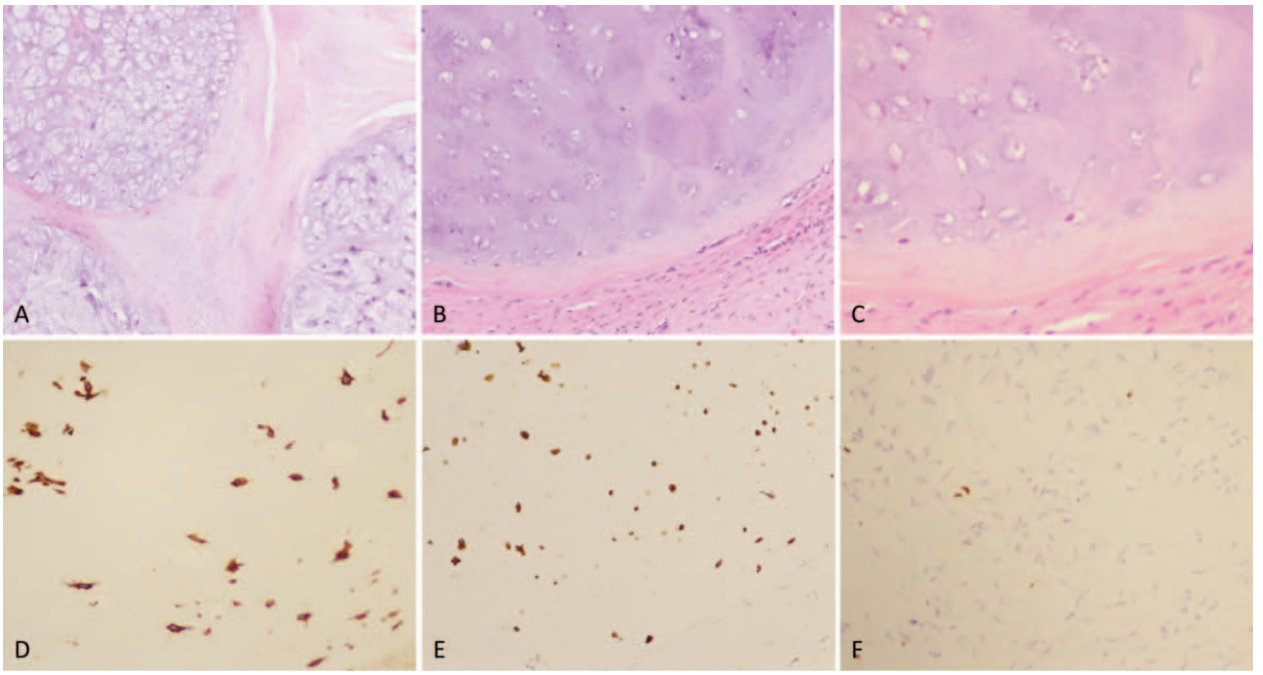

Figure 5. Pathologic histology of tumor specimens. (A–C) Microphotography showing characteristic nests of tumor cells (Zellballen) (H&E, original magnification 40×, 100×, and 200×). (D, E) Immunohistochemistry of the lesion showed Vimentin and S100 positive staining. (F) Ki-67 immunostaining shows 5% Ki-67 positive cells. Ki-67 staining is localized in the tumor nuclei.

active intrasynovial lesions and free articular bodies. In stage III (late stage), there are multiple free articular bodies in the absence of intrasynovial involvement.[1–3,10,11]

The most common radiologic findings, free bodies with varying sizes, can be seen at any place in the tumor cavity.[12,13] Calcification occurs at the last stage and may not be observed in some patients at earlier stage.[14] Intraarticular liquid-like masses, non-calcified masses or swellings which can be differentiated with MRI or CT scan.[12–15] However, difficulties of diagnosis despite CT and MRI should also be considered. In neglected cases or in cases with a long-term disease course, changes in bone or joint cartilage induced by multiple intraarticular lesions, bony erosion, or presence of local osteoporosis make accurate preoperative diagnosis much more difficult.[16] Imaging studies including CT, MRI, bone scan and PET/CT are non-specific, making it difficult to differentiate SOC from other common occupying lesions.[15–17] However, imaging studies may play a crucial role in the decision making of surgical intervention. In our case, the entire size of SOCs was $24 \times 20 \times 12$ cm, and our case had been one of the biggest SOCs ever reported in literature.

Differential diagnosis of synovial osteochondromatosis includes many other occupying diseases. SOC should be differentiated from multiple benign or malignant lesions such as synovial hemangioma, pigmented villonodular synovitis, synovial cyst, osteosarcoma and synovial sarcoma.[18–21] Malignant transformation was reported in few cases at long-term follow-up visit.[21,22] While the literature describes the knee as the most common location of the usual form of SOC, the location in the thigh is extremely a rarity.

The "gold-standard" diagnosis of SOC relies on pathological findings. The main pathological characteristic is chondroid metaplasia of the subintimal tissue of synovial joints.[1–3,23,24] Further reports and analyses of the giant form of SOC are necessary to improve our understanding of this pathological entity and its differences from the usual form to optimize proper clinical management. In our reported case, pathology results showed significant chondroid metaplasia without cellular atypia, which was consistent with synovial osteochondromatosis.

The treatment of first choice for SOC is surgical excision with an open or arthroscopical approach.[1–3,25–27] In the literature, early arthroscopical or open debridement, synoviectomy, and removal of free cartilage masses at an early stage before cartilage damage occurs have shown to be efficient treatments.[25–27] Severity of disease course and affected location should also be considered when surgeons decide whether surgery should be performed.[25,26] Additionally, in some cases that had osteochondroplasty after debridement osteoarthritis did not relapse in 2-years follow up and successful results were achieved.[25–28] Under this circumstance, surgical extent, surgical procedures, and postoperative complications are critical factors that need further investigation. According to our single-center experience, we prefer and recommend open surgery for patients with giant SOCs due to the size of the mass.

In conclusion, this is the first report of giant synovial osteochondromatosis of the thigh in a patient. Although uncommon, synovial osteochondromatosis of the thigh should be part of the differential when the patient presents with atypical symptoms. Surgical treatment is a definite therapy of first choice. Our case highlights the significance of accurate diagnosis and proper treatment for synovial osteochondromatosis. With an accurate diagnosis, proper planning, and accurate surgical manipulation, synovial osteochondromatosis can be diagnosed and managed much more effectively.

Acknowledgments

We would like to thank our colleagues at the Department of Orthopedic Surgery, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College.

Author contributions

Conceptualization: Shuzhong Liu, Xi Zhou, An Song, Yipeng Wang, Yong Liu.
Funding acquisition: Shuzhong Liu, Yipeng Wang, Yong Liu.
Investigation: Shuzhong Liu, Xi Zhou, An Song, Zhen Huo, Yong Liu.
Resources: Shuzhong Liu, Xi Zhou, Zhen Huo, Yipeng Wang, Yong Liu.
Supervision: Yipeng Wang, Yong Liu.
Writing – original draft: Shuzhong Liu, Xi Zhou, An Song, Yong Liu.
Writing – review & editing: Shuzhong Liu, An Song, Yipeng Wang, Yong Liu.

References

[1] Tuncer K, Izgi E, Cankaya B, et al. Huge bursitis and bursal synovial osteochondromatosis associated with scapular osteochondroma mimicking a giant calcific mass of the chest wall. Am J Phys Med Rehabil 2019;98:e1–3.
[2] Ropars M, Hervé A, Stock N, et al. Giant synovial chondromatosis of the metacarpophalangeal joint. Joint Bone Spine 2016;83:351.
[3] Serbest S, Tiftikçi U, Karaaslan F, et al. A neglected case of giant synovial chondromatosis in knee joint. Pan Afr Med J 2015;22:5.
[4] Jamshidi K, Barbuto R, Shirazi MR, et al. Giant solitary synovial chondromatosis mimicking chondrosarcoma: report of a rare histologic presentation and literature review. Am J Orthop (Belle Mead NJ) 2015;44:E286–90.
[5] Efrima B, Safran N, Amar E, et al. Simultaneous pigmented villonodular synovitis and synovial chondromatosis of the hip: case report. J Hip Preserv Surg 2018;5:443–7.
[6] Liu X, Wan S, Shen P, et al. Diagnostic accuracy of synovial chondromatosis of the temporomandibular joint on magnetic resonance imaging. PLoS One 2019;14:e0209739.
[7] Kwon DR, Chae S, Moon YS, et al. Carpal tunnel syndrome caused by synovial osteochondromatosis of the finger flexor tendon: a case report. Medicine (Baltimore) 2018;97:e13943.
[8] Wahab H, Hasan O, Habib A, et al. Arthroscopic removal of loose bodies in synovial chondromatosis of shoulder joint, unusual location of rare disease: a case report and literature review. Ann Med Surg (Lond) 2018;37:25–9.
[9] Wen J, Liu H, Xiao S, et al. Synovial chondromatosis of the hip joint in childhood: a case report and literature review. Medicine (Baltimore) 2018;97:e13199.
[10] Lee YK, Moon KH, Kim JW, et al. Remaining loose bodies after arthroscopic surgery including extensive capsulectomy for synovial chondromatosis of the hip. Clin Orthop Surg 2018;10:393–7.
[11] Vellone V, Bracciolini V, Ramieri V, et al. Synovial chondromatosis and calcium pyrophosphate deposition of the temporomandibular joint: challenging diagnosis. J Craniofac Surg 2018;29:e792–4.
[12] Zhu W, Wang W, Mao X, et al. Arthroscopic management of elbow synovial chondromatosis. Medicine (Baltimore) 2018;97:e12402.
[13] Kim HS, Lee W, Choi JW, et al. Temporomandibular joint synovial chondromatosis accompanying temporal bone proliferation: a case report. Imaging Sci Dent 2018;48:147–52.
[14] Aramberri M, Tiso G, Haeni DL. Arthroscopic and endoscopic technique for subcoracoid synovial chondromatosis of the shoulder through a medial transpectoral portal. Arthrosc Tech 2018;7:e279–83.

[15] Yang YP, Wang JJ, Li HY. Atypical synovial chondromatosis of the right knee: a case report. Exp Ther Med 2018;15:4503–7.
[16] Yukata K, Murase M, Hashimoto T, et al. Ulnar nerve palsy caused by synovial protrusion in synovial chondromatosis of the elbow: a case report and literature review. Shoulder Elbow 2018;10:128–32.
[17] Kunzler DR, Shazadeh Safavi P, Warren BJ, et al. Arthroscopic treatment of synovial chondromatosis in the ankle joint. Cureus 2017;9:e1983.
[18] Khandwala K, Waheed AA, Alvi MI, et al. Bursal synovial chondromatosis secondary to underlying osteochondroma in a child. Cureus 2017;9:e1944.
[19] Leite PCC, Tolentino ES, Yamashita AL, et al. Surgical treatment of synovial chondromatosis in the inferior compartment of the temporomandibular joint with articular disc involvement. J Craniofac Surg 2018;29:e199–203.
[20] Dwidmuthe S, Sharma M. A case report of primary synovial chondromatosis with bilateral genu valgum. J Orthop Case Rep 2017;7:92–5.
[21] Chen TJ, Tsai YF, Chou YH, et al. Shoulder joint synovial chondromatosis presenting as multiple axillary masses: a case report. J Clin Ultrasound 2018;46:361–3.
[22] Derek Stensby J, Fox MG, Kwon MS, et al. Primary synovial chondromatosis of the subtalar joint: case report and review of the literature. Skeletal Radiol 2018;47:391–6.
[23] Trevino M, Laks S, Kafchinski L, et al. Intermetatarsal bursa primary synovial chondromatosis: case report and review of the literature. Skeletal Radiol 2017;46:1769–73.
[24] Philip MC, Usman S. Synovial chondromatosis: a rare differential diagnosis of hip pain in a child. J Orthop Case Rep 2017;7:37–9.
[25] Ng VY, Louie P, Punt S, et al. Malignant transformation of synovial chondromatosis: a systematic review. Open Orthop J 2017;11:517–24.
[26] Acharya BM, Devkota P, Shrestha SK, et al. Bilateral symmetrical synovial chondromatosis of shoulder: a case report. Rev Bras Ortop 2017;53:647–50.
[27] Kreines A, McMillan S, Ford E, et al. Reverse total shoulder arthroplasty for the treatment of synovial chondromatosis: a case report and review of the literature. Arch Bone Jt Surg 2017;5:117–20.
[28] Houdek MT, Wyles CC, Rose PS, et al. High rate of local recurrence and complications following total knee arthroplasty in the setting of synovial chondromatosis. J Arthroplasty 2017;32:2147–50.