

Pigmented villonodular synovitis of the knee joint in a 5-year-old girl treated with combined open and arthroscopic surgery: a case report

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Pigmented villonodular synovitis is an extremely rare disease in skeletally immature patients. Erosive destruction of the involved joint leads to early arthritis, and its high recurrence rate makes treatment challenging. Multiple surgical approaches exist, but it is unclear as to which among them achieves the lowest possible recurrence rate and morbidity. We report the case of a 5-year-old girl with left knee pain and swelling who was diagnosed with diffuse pigmented villonodular synovitis of the left knee based on MRI findings. Combined open and arthroscopic surgery was performed to completely remove the tumor. Postoperative histopathological examination confirmed the diagnosis of diffuse pigmented villonodular synovitis. The postoperative course was uneventful, with a gradual improvement in symptoms. There were no signs of recurrence on postoperative MRI performed at the 8-month follow-up, with neither knee pain nor limitation of range of motion. The favorable outcome in this case suggests that combined

open and arthroscopic surgery may be an effective method for treating pigmented villonodular synovitis in skeletally immature patients. *J Pediatr Orthop B* 00:000–000
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Introduction

Pigmented villonodular synovitis (PVNS) is a rare, benign, proliferative lesion of synovial tissue and is most likely to be neoplastic [1–3]. It is most prevalent in young adults between 20 and 45 years of age, with no sex preference. It predominantly involves the knee joint, followed by the hip, ankle, elbow, and shoulder joints [4,5], and is a locally aggressive lesion that is categorized into localized/nodular and diffuse forms [1,2,6]. PVNS is extremely rare in children and may pose a diagnostic challenge, whereas delay in diagnosis has the potential to lead to early joint destruction and development of arthritis [2,6].

Complete surgical resection of the diseased synovial lining remains the standard management for disease eradication in either form of PVNS. Although surgical tumor resection is the treatment of choice at present, the best method of resection is under debate [1,6]. The disease outcome and surgical result are generally assessed in terms of recurrence rate, arthritic progress, and perioperative complications.

Here, we present a case of PVNS of the knee, in which combined open and arthroscopic surgery was successfully performed in a small child.

Case report

A 5-year-old girl had a 3-month history of spontaneous, gradually progressive, left knee pain and swelling. There were no other associated symptoms or other joint involvement and her general health was good. On physical examination, there was diffuse, soft-to-firm swelling of the left knee joint without any signs of inflammation (Fig. 1). The range of motion (ROM) of the left knee was 0–110°. Her laboratory findings were within normal limits. Plain radiography of her left knee showed no obvious abnormality (Fig. 2). MRI of the left knee revealed an intra-articular and periarticular mass with synovial fluid collection (Fig. 3a–d). T2*-weighted images showed a low-intensity area in the tumor, indicating hemosiderin deposition (Fig. 3e). The soft tissue mass had low-intensity on both T1-weighted and T2-weighted images, mostly confined to the suprapatellar and anterolateral aspects of the knee (Fig. 3f and g). On the basis of clinical examination and radiological findings, the working diagnosis was diffuse PVNS (DPVNS).

Fig. 1



Clinical appearance of diffuse swelling of the left knee.

The management strategy was planned according to the size, location, age, and type of lesion. Under general anesthesia, a tourniquet was applied and an intraoperative open biopsy was performed through a small incision to obtain the pathological diagnosis, which took around 30 min. A suprapatellar incision was then made for en-bloc resection of the diffuse mass that extended from the suprapatellar pouch to the lateral pouch. Following open resection, closure of capsule and suprapatellar incision was carried out. Arthroscopic survey of the intra-articular lesion was carried out and residual tumor was resected from the anterior and posterior horns of the lateral meniscus, intercondylar notch, and also from the posteromedial aspect of the knee (Fig. 4). There was no osteochondral defect in the knee joint, and other structures were normal. Meticulous closure of the capsule and the wound was carried out, and a suction drain remained *in situ*. The total duration of surgery was 184 min including 54 min of tourniquet time.

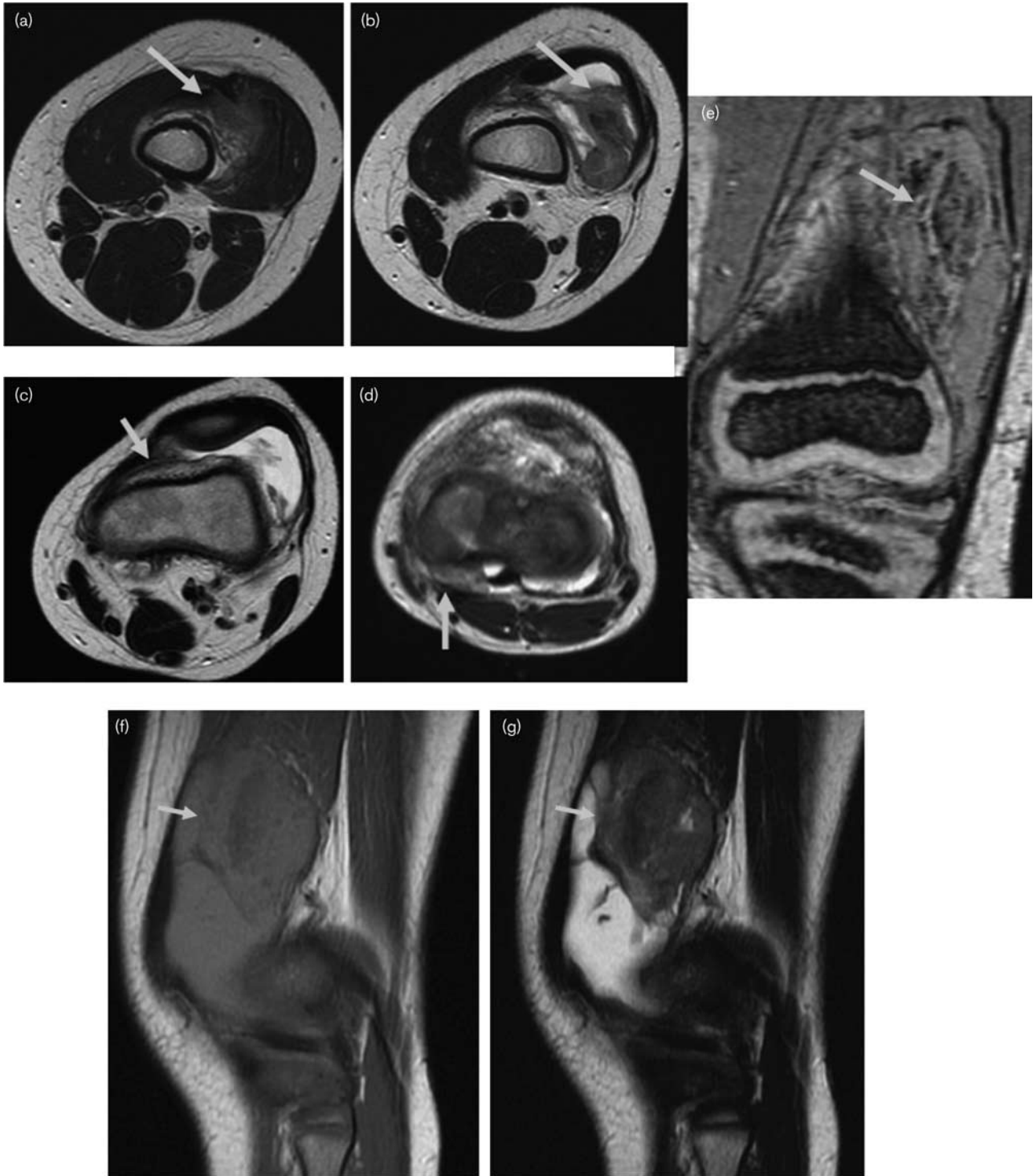
The final histopathological diagnosis was PVNS with small histiocyte-like cells and a few giant cells, which were CD68-positive, and hemosiderin deposition was identified (Fig. 5). The postoperative period was uneventful, and rehabilitation was started after drain removal on postoperative day 2. Her knee pain and limitation of ROM were disappeared at 4 months postoperatively. MRI at 8 months postoperatively showed no obvious signs of recurrence (Fig. 6) and her symptoms had continually improved. There was no obvious

Fig. 2



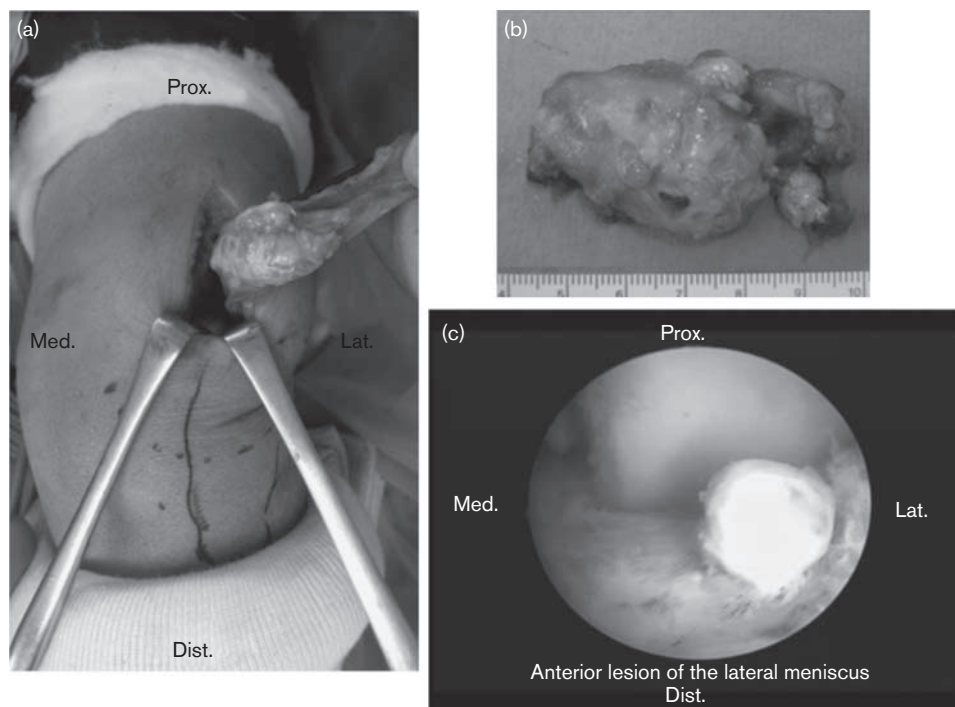
Plain radiographs of the left knee: (a) anterior posterior and (b) lateral views showing no obvious abnormality.

Fig. 3



Preoperative MRI of the left knee. Axial T2-weighted images from the proximal to distal ends: (a) superior extent of the mass with extracapsular invasion (arrow); (b) intra-articular mass in the suprapatellar aspect of the joint (arrow); (c) mass in the anterior aspect of the joint (red arrow) with significant joint effusion; and (d) mass in the posterior aspect of the joint (arrow). (e) Coronal T2*-weighted images showing a low-intensity area in the tumor (arrow), indicating hemosiderin deposit. (f) Sagittal T1-weighted and (g) T2-weighted images showing a low-intensity intra-articular mass in the anterior aspect of the joint (arrows).

Fig. 4



Intraoperative picture: (a) A diffuse tumor in the suprapatellar pouch that extends to the lateral pouch. (b) En-bloc resected tumor mass. (c) Arthroscopy showing residual tumor lesion in the anterior horn of the lateral meniscus.

recurrence in clinical examination and ultrasound at 18 months after operation.

Discussion

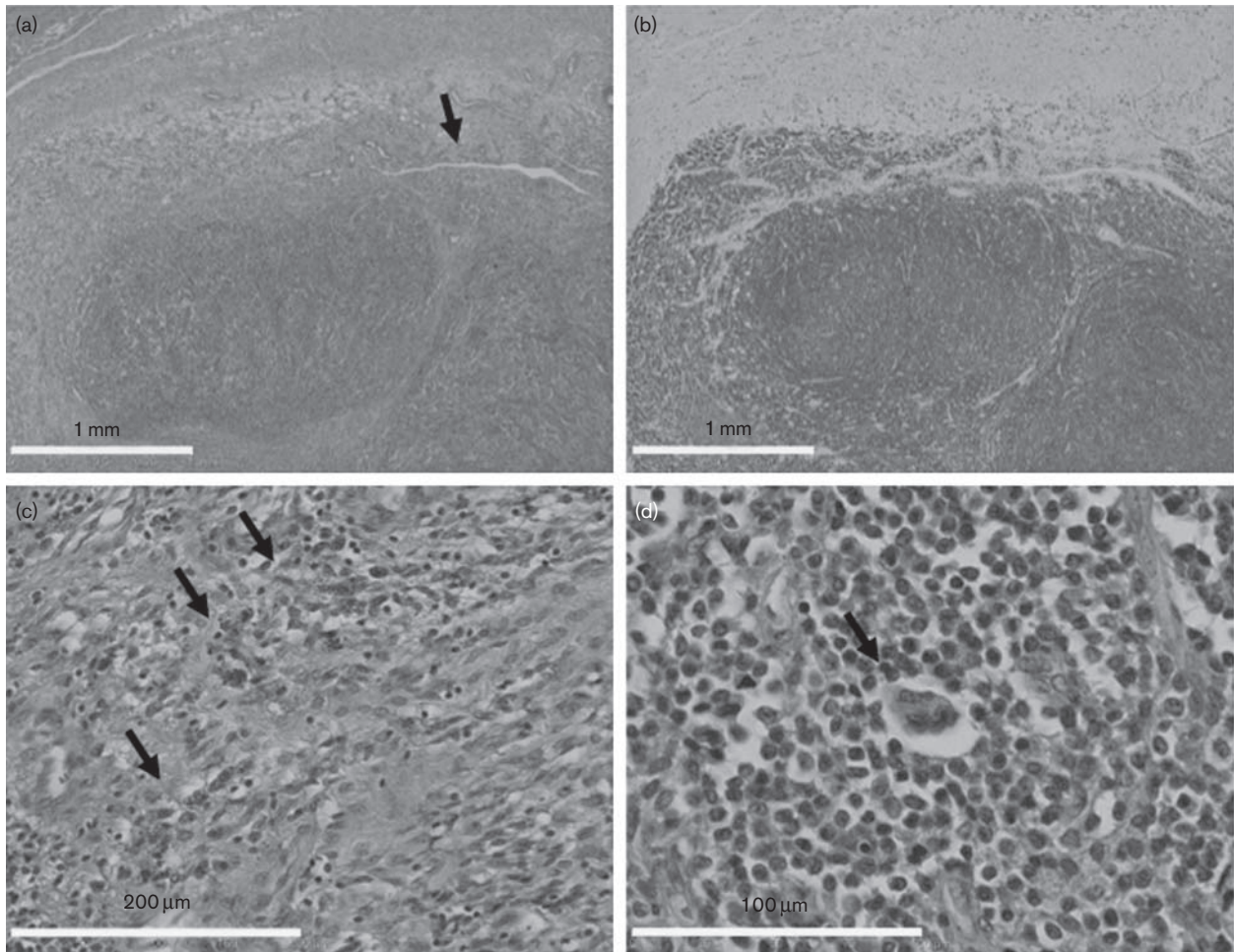
Clinical presentation of PVNS depends on the morphological form of the disease. The majority of cases are monoarticular, occurring in the third and fourth decades of life [3,5]. Neubauer *et al.* [2] reported that only one of six children was diagnosed before surgery because PVNS occurs rarely in skeletally immature patients. The radiographic findings of the disease are often nonspecific, and commonly demonstrate an ill-defined, periarticular soft tissue mass that appears more opaque than the surrounding soft tissue because of hemosiderin deposition [3]. A multinodular intra-articular lesion with patchy areas that have the characteristics of hemosiderin are strongly suggestive of PVNS, particularly when a bleeding disorder like hemophilia has been excluded [3,7]. Its capacity to erode subchondral bone, resulting in cysts and erosion leading to secondary degenerative lesion of the joint, emphasizes the importance of early diagnosis [1,6].

The treatment of choice for PVNS is surgical excision of the lesion, performed either arthroscopically or through an open arthrotomy. Localized PVNS rarely recurs because complete marginal resection is often obtained [6, 8,9]. However, treatment for DPVNS has not been clearly defined as the recurrence rate is especially high [1,

6]. Regardless of the technique used, the treatment principle for either form remains the same: careful removal of the synovial tumor [6]. Some authors have suggested radiotherapy as adjuvant treatment to reduce the high rate of recurrence. However, the use of radiotherapy for such a benign tumor is controversial because of potential damage to the epiphyseal growth plate in children and postradiation sarcoma [6,10]. Moreover, radiotherapy or isotopic synoviorthesis has not shown promising results in controlling recurrence [10,11].

DPVNS of the knee remains a clinical challenge because of the high rate of local recurrence and propensity for causing pain, swelling, and arthritic progress. Total synovectomy is accepted as standard of care, but the literature describes multiple methods of accomplishing this, including open, all arthroscopy or combined open and arthroscopic surgery [1]. Each method has its own drawbacks. One major complication of the open technique in the knee is postoperative stiffness followed by decrease in ROM [4,12]. Extracapsular extension of the disease is a well-known absolute contraindication to only arthroscopic synovectomy [1]. Hence, the gold standard for achieving the lowest possible recurrence rate and morbidity remains unclear. In a literature review, Nakahara *et al.* [4] found a 23.2% recurrence rate with open synovectomy and a 39% recurrence rate with all arthroscopic synovectomy. Colman *et al.* [1] reported that

Fig. 5



Microscopic images of diffuse pigmented villonodular synovitis. (a) A photomicrograph showing proliferative mononuclear histiocyte-like cells with cleft-like spaces (arrow). Hematoxylin-eosin staining ($\times 25$; scale bar, 1 mm). (b) CD68-positive tumor cells ($\times 25$; scale bar, 1 mm). (c) Hemosiderin deposits (arrows). Hematoxylin-eosin staining ($\times 200$; scale bar, 200 μm). (d) A few multinucleated giant cells (arrow).

the overall recurrence rate of the combined open and arthroscopic group was lower compared with the all arthroscopic or open group (9 vs. 62 vs. 64%, respectively).

In our case, as there was extracapsular invasion in the suprapatellar and anterolateral portions of the knee, we decided to use the combined open and arthroscopic approach. En-bloc resection of a large lesion was possible through the open approach. Residual lesions were identified and resected by means of arthroscopy through inspection of the knee joint. Arthroscopy was useful for the lesion such as the posteromedial aspect of the knee, which was difficult to be observed by means of only open surgery. Hence, careful resection of the tumor was possible to prevent recurrence. This combined method appears to be suitable for DPVNS, especially in small children, to avoid joint complications.

Conclusion

Clinical suspicion of PVNS should be considered in children with chronic joint swelling. DPVNS of the knee has a high overall recurrence rate regardless of treatment. Nevertheless, combined open and arthroscopic synovectomy is a comprehensive approach associated with both a low recurrence and low postoperative complication rates.

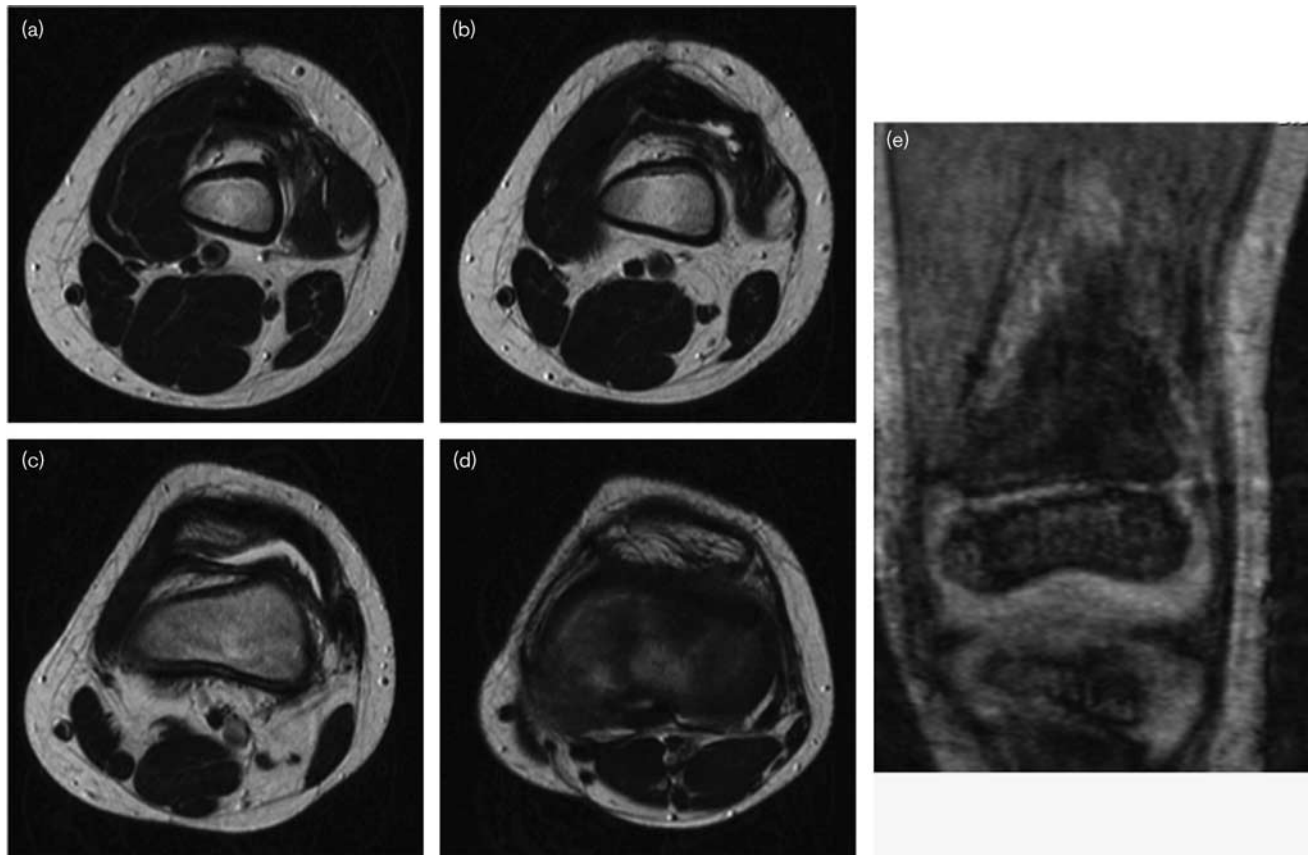
Acknowledgements

The manuscript submitted does not contain information about medical device(s)/drug(s).

Conflicts of interest

There are no conflicts of interest.

Fig. 6



Postoperative MRI of the left knee at 8-month follow-up. (a–d) Axial images from the proximal to distal ends. (e) Coronal T2*-weighted images showing no abnormal tissue or other signs of recurrence.

References

- Colman MW, Ye J, Weiss KR, Goodman MA, McGough RL. Does combined open and arthroscopic synovectomy for diffuse PVNS of the knee improve recurrence rates? *Clin Orthop Relat Res* 2013; **471**:883–890.
- Neubauer P, Weber AK, Miller NH, McCarthy EF. Pigmented villonodular synovitis in children: a report of six cases and review of the literature. *Iowa Orthop J* 2007; **27**:90–94.
- Mukhopadhyay K, Smith M, Hughes PM. Multifocal PVNS in a child—followed over 25 years. *Skeletal Radiol* 2006; **35**:539–542.
- Nakahara H, Matsuda S, Harimaya K, Sakamoto A, Matsumoto Y, Okazaki K, et al. Clinical results of open synovectomy for treatment of diffuse pigmented villonodular synovitis of the knee: case series and review of literature. *Knee* 2012; **19**:684–687.
- Dorwart RH, Genant HK, Johnston WH, Morris JM. Pigmented villonodular synovitis of synovial joints: clinical, pathologic, and radiologic features. *Am J Roentgenol* 1984; **143**:877–885.
- Baroni E, Russo BD, Masquijo JJ, Bassini O, Miscione H. Pigmented villonodular synovitis of the knee in skeletally immature patients. *J Child Orthop* 2010; **4**:123–127.
- Hughes TH, Sartoris DJ, Schweitzer ME, Resnick DL. Pigmented villonodular synovitis: MRI characteristics. *Skeletal Radiol* 1995; **24**:7–12.
- Kim SJ, Shin SJ, Choi NH, Choo ET. Arthroscopic treatment for localized pigmented villonodular synovitis of the knee. *Clin Orthop Relat Res* 2000; **379**:224–230.
- Moskovich R, Parisien JS. Localized pigmented villonodular synovitis of the knee. Arthroscopic treatment. *Clin Orthop Relat Res* 1991. 218–224.
- Blanco CE, Leon HO, Guthrie TB. Combined partial arthroscopic synovectomy and radiation therapy for diffuse pigmented villonodular synovitis of the knee. *Arthroscopy* 2001; **17**:527–531.
- Chin KR, Barr SJ, Winalski C, Zurakowski D, Brick GW. Treatment of advanced primary and recurrent diffuse pigmented villonodular synovitis of the knee. *J Bone Joint Surg Am* 2002; **84-A**:2192–2202.
- Flandry FC, Hughston JC, Jacobson KE, Barrack RL, McCann SB, Kurtz DM. Surgical treatment of diffuse pigmented villonodular synovitis of the knee. *Clin Orthop Relat Res* 1994; **300**:183–192.

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