

**CASE REPORT**

**Open Access**

# Desmoid tumor of posterior cruciate ligament of the knee: a case report

Wang Ling<sup>1†</sup>, Song Kedong<sup>2†</sup>, Wang Hong<sup>3\*</sup>, Zhang Weiguo<sup>3</sup> and Lv Decheng<sup>3</sup>

## Abstract

**Background:** Desmoid tumor is a rare type of cancer that develops in the tissues that form tendons and ligaments. These tumors, also called aggressive fibromatosis, are considered benign with no metastatic potential. They may invade nearby tissues and organs, however, and can be difficult to control. Desmoid tumor in the posterior cruciate ligament (PCL) of the knee has never been described in the literature.

**Case presentation:** A 49-year-old man presented with a 2-month history of posteromedial knee dull pain and decreased range of motion of the knee. He was diagnosed desmoid tumor of posterior cruciate ligament of the knee by intraoperative biopsy, and underwent successful PCL resection and reconstruction by Four-strand semitendinosus and gracilis tendon autograft arthroscopically, and fortunately five years after operation, there were no clues as to recurrence of the tumor examined by Magnetic Resonance Imaging (MRI).

**Conclusion:** Desmoid tumor is characterized by infiltrative growth and a tendency towards recurrence, as this tumor entity is rare, data giving evidence based recommendations for the optimal treatment algorithm for this disease is lacking. At present there is no definite and effective method of treatment. However, early detection of the tumor play an important role, MRI is now the most important method for the detection of tumor extent, which facilitates the treatment choice as well as the prediction of prognosis. In our case, we followed-up the patient five years postoperatively by MRI and got a good result.

**Keywords:** Desmoid tumor, Posterior cruciate ligament, Knee, Arthroscopical reconstruction, Surgery

## Background

Desmoid tumor, also known as aggressive fibromatosis, is histologically benign fibrous lesion with aggressive infiltration of adjacent tissues and high local recurrence rate but without metastatic potential. It is a relatively rare neoplasm accounting for 3% of soft tissue tumors and 0.03% of all neoplasms. Main locations of desmoid tumor occurrence are the abdominal wall, proximal extremities, and the mesenteric intestine in patients with familial adenomatous polyposis (FAP) [1]. Desmoid tumor covers a broad spectrum of fibromatoses, which arise from the connective tissue, fascia or aponeurosis of a muscle [2]. The exact pathogenesis of desmoid tumor is unknown, however, genetic abnormalities (Familial adenomatous polyposis and Gardner's syndrome), sex hormones and trauma have

been considered as causative factors [1]. In our case the patient reported to have undergone extensive walking before the tumor occurred. This leads to the assumption, that the growth of the tumor could have been induced by this repeated minor trauma to the ligament, as is reported by Stollwerck et al. [3]. The incidence of desmoid tumor is reported to be 2.4 to 4.3 cases/1 million/year, whilst the majority of desmoid tumors are found intraabdominally and only 7–15% of which are found in the extra-abdominal area.

## Case presentation

A 49-year-old man presented with a 2-month history of posteromedial knee dull pain and decreased range of motion of the knee. The patient also complained of subjective swelling of the involved lower leg. The general physical examination was unremarkable. Examination of the knee showed no effusion, there was no palpable inguinal or popliteal lymphadenopathy, neither warmth nor erythema was detected. There was no ligamentous

\* Correspondence: whwl@hotmail.com

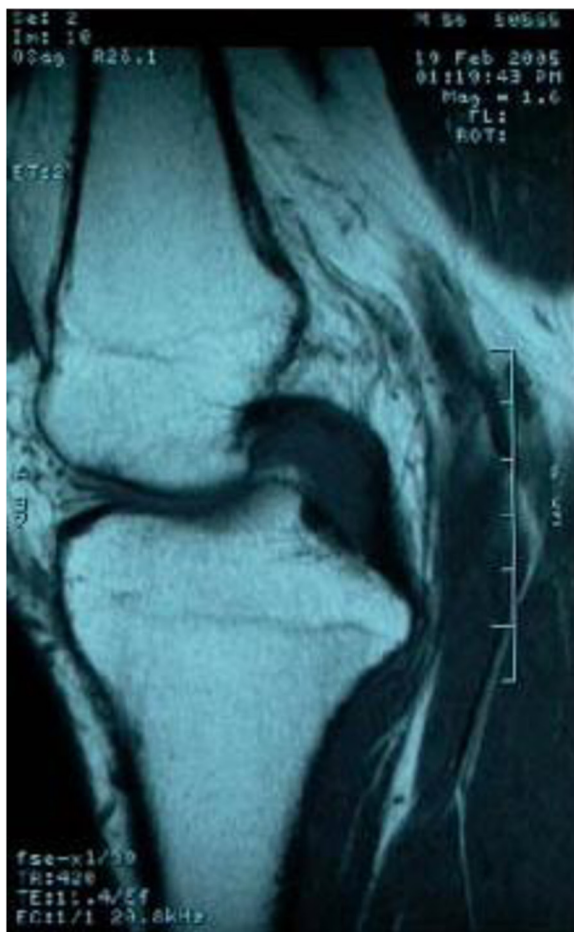
<sup>†</sup>Equal contributors

<sup>3</sup>Department of Orthopaedics, First Affiliated Hospital of Dalian Medical University, Dalian, People's Republic of China

Full list of author information is available at the end of the article

instability or joint line tenderness. The only and most significant positive sign was that the range of motion of the right knee was limited to 0°-90°. The rest of his musculoskeletal examination was normal. The laboratory workup was unremarkable. No recent trauma or major injury had occurred, but the patient walked 4-5 hours per day to work for about 6 months, which might have some business with it according to the patient himself.

Plain radiographs of the right knee were unremarkable except for mild degenerative changes. Computed tomographic (CT) examination was not carried out because of financial concern. T1-weighted MRI (Figure 1) showed slight hypo-intensity lesion similar to muscle and peripheral hypo-intensity ring. Fat-suppressed T2-weighted MRI (Figure 2) showed hyper-intensity lesion with distinct border localized in posterior cruciate ligament. Contrast MR image (Figure 3) showed slightly heterogeneous moderate enhancement and hypo-intensity ringlike non-enhancement.



**Figure 1** Sagittal, coronal and axial T1-weighted MR image showed slight hypo-intensity lesion similar to muscle and peripheral hypo-intensity ring.



**Figure 2** Sagittal fat-suppressed T2-weighted MR image showed hyper-intensity lesion with distinct border localized in posterior cruciate ligament.

Firstly, diagnostic arthroscopy was performed. The posterior cruciate ligament appeared wider and grayer than normal on gross examination, and it was tough and inelastic (Figure 4). Intraoperative frozen histology showed a uniform proliferation of spindle cells with a moderate amount of collagen fibers led to a diagnosis of desmoid tumor in the PCL (Figure 5). Then the PCL was completely resected by clipper bit by bit, and the remaining part of the starting and ending points of the tendon were cleared off by plane iron and low temperature radiofrequency ablation. After that it was reconstructed by Four-strand semitendinosus and gracilis tendon autograft arthroscopically directly after the intraoperative histology and resection. Endobuttons were used to fix the ends of the transplanted ligament. No adjuvant treatment was given postoperatively.

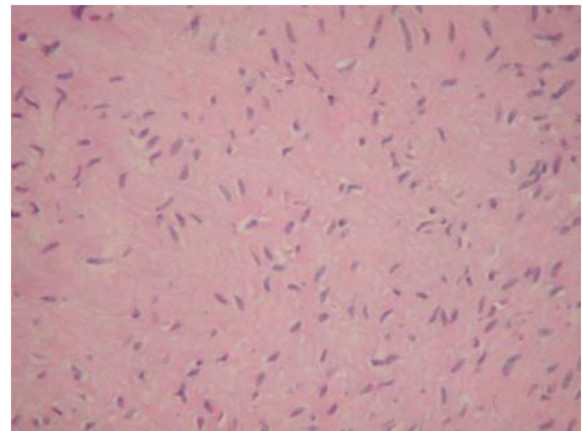
Since then, the patient has remained asymptomatic without any pain around the knee, and after a six-month rehabilitation process, the range of motion of the involved knee reached 0°-130°. Now five years after operation, there



**Figure 3** Contrast MR image showed slightly heterogeneous moderate enhancement and hypo-intensity ringlike non-enhancement.



**Figure 4** The PCL appeared wider and grayer than normal on gross examination, and it was tough and inelastic.



**Figure 5** Histopathological evaluation revealed uniform proliferation of spindle cells, with a moderate amount of collagen fibers, suggesting desmoid tumor.

were no clues as to recurrence of the tumor examined by MRI (Figure 6).

#### Discussion

Desmoid tumor is characterized by infiltrative growth and a tendency towards recurrence; however, unlike sarcoma,



**Figure 6** MRI of left knee 5 years after operation showing the satisfactory morphology of the reconstructed PCL and no signs of recurrence.

it never metastasizes. To avoid local recurrence, an early diagnosis is required for this type of tumor, which is difficult as most of these patients are asymptomatic like other kinds of rare tumors within the knee including Intra-articular ganglia, Extra-osseous osteochondroma, pigmented villonodular synovitis and even sarcoma inside the knee [4-8]. At the same time, it is also important to preoperatively detect the exact localization and extent of the lesion in this tumor, and MRI is now the most important method for detection of the tumor, diagnosis, decision making and follow-up. In our case, MRI showed that the pathological changes were limited within PCL, after a complete resection and one stage functional reconstruction, we followed-up the patient five years postoperatively by MRI and there were no clues as to recurrence of the tumor. Here, we think the early stage diagnosis and complete resection contribute mainly to the good results.

Desmoid tumor of the abdominal wall mostly occurs in women during pregnancy or soon after delivery, whereas extra-abdominal desmoid tumor is more frequent from puberty to 50 years of age, with both men and women equally affected. Extra abdominally desmoid tumor may occur in a variety of anatomical locations, including the muscles of the shoulder, the chest wall and back, thigh, and head and neck. However, solitary occurrence in PCL of the knee has never being reported. Here, we present a case of desmoid tumor in the cruciate ligament of knee joint. Resection and functional reconstruction was successfully performed, and the patient has been tumor-free for five years after surgery.

Unexplained etiology and the various locations make the treatment of desmoid tumor extremely difficult. As this tumor entity is rare, data giving evidence based recommendations for the optimal treatment algorithm for this disease is lacking [9]. At present optimal treatment of desmoids remains controversial, and there is no definite and effective method of treatment. However, complete resection or a wide surgical excision is the first choice of therapy for this type of tumors, as was advocated nearly 100 years ago [1], although Some authors affirmed that there were no significant difference between negative margins and microscopic positive margin regarding recurrence rate [10-12]. For patients with desmoid tumors that are not amenable to surgery, radiotherapy or adjuvant therapy using non steroidal anti-inflammatory agents, tamoxifen, interferon, anti-neoplastic agents, either alone or in combination have been reported to be useful in some cases, but unfortunately, the exact benefit offered by them is lack of evidence due to non-sufficient patient numbers and missing prospective randomized studies, although The French sarcoma group recently reported encouraging results in patient treated with chemotherapy [13]. We did not apply any above ad-junctive treatment of choice and fortunately, the patient

has been tumor-free for five years after surgery. For this patient we think early detection of the tumor because of the proper application of MRI in the beginning and complete resection is the key role of the good results, but still longer term curative effect remains to be followed-up.

## Conclusion

Early detection of desmoid tumor and complete resection play an important role in its prognosis, and MRI is a good choice for its early detection.

## Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

## Competing interests

The authors declare that they have no competing interests. No financial support has been received.

## Authors' contributions

WL and SK have been involved in drafting the manuscript and revising it critically for important intellectual content; ZW, LD have made substantial contributions to conception and design, acquisition of data, analysis and interpretation of data; WH has taken part in the whole procedure. All authors read and approved the final manuscript.

## Acknowledgements

This work was supported by the National Natural Science Foundation of China (No. 81271719), Research Fund for the Doctoral Program of Higher Education of China (20112105120005 and 20112105120006), Fok Ying Tung Education Foundation (132027), the Fundamental Research Funds for the Central Universities (DUT11SM09/DUT12JB09) and SRF for ROCS, SEM.

## Author details

<sup>1</sup>Department of Oncology, First Affiliated Hospital of Dalian Medical University, Dalian, People's Republic of China. <sup>2</sup>Dalian R&D Center for Stem Cell and Tissue Engineering, Dalian University of Technology, Dalian 116024, China. <sup>3</sup>Department of Orthopaedics, First Affiliated Hospital of Dalian Medical University, Dalian, People's Republic of China.

Received: 23 July 2012 Accepted: 19 February 2013

Published: 23 February 2013

## References

1. Ferenc T, Sygut J, Kopczynski J, Mayer M, Latos-Bielenska A, Dziki A, Kulig A: Aggressive fibromatosis (desmoid tumors): definition, occurrence, pathology, diagnostic problems, clinical behavior, genetic background. *Pol J Pathol: official journal of the Polish Society of Pathologists* 2006, **57**(1):5-15.
2. Goy BW, Lee SP, Eilber F, Dorey F, Eckardt J, Fu YS, Juillard GJ, Selch MT: The role of adjuvant radiotherapy in the treatment of resectable desmoid tumors. *Int J Radiat Oncol* 1997, **39**(3):659-665.
3. Kumar V, Khanna S, Khanna AK, Khanna R: Desmoid tumors: experience of 32 cases and review of the literature. *Indian J Cancer* 2009, **46**(1):34-39.
4. Herzog J, Miles J, Skinner JA: Primary sarcoma of the anterior cruciate ligament - a case report. *Knee* 2012, **19**(1):69-71.
5. Tachibana Y, Ninomiya T, Goto T, Yamazaki K, Ninomiya S: Intra-articular ganglia arising from the posterior joint capsule of the knee. *Arthroscopy: the journal of arthroscopic & related surgery: official publication of the Arthroscopy Association of North America and the International Arthroscopy Association* 2004, **20**(Suppl 2):54-59.
6. Oliva F, Marconi A, Fratoni S, Maffulli N: Extra-osseous osteochondroma-like soft tissue mass of the patello-femoral space. *BMC Musculoskeletal Disord* 2006, **7**:57.
7. Kim RS, Lee JY, Lee KY: Localized pigmented villonodular synovitis attached to the posterior cruciate ligament of the knee. *Arthroscopy: the journal of arthroscopic & related surgery: official publication of the Arthroscopy*

*Association of North America and the International Arthroscopy Association* 2003, **19**(6):E32–E35.

8. Oliva F, Frizziero A: **One step open synovectomy without adjuvant therapy for diffuse pigmented villonodular synovitis of the knee in a soccer player.** *Muscles, Ligaments And Tendons Journal* 2011, **1**(1):36–39.
9. Bouffleur AI: **Fibrosarcoma of abdominal wall: Desmoid fibroma beginning in Poupart's ligament and extending around the iliac vessels into the pelvis—removal with resection of the abdominal wall and extensive dissection of the iliac vessels—repair of defect in abdominal wall by plastic operation.** *J Am Med Assoc* 1899, **33**:820–823.
10. de Bree E, Keus R, Melissas J, Tsiftsis D, van Coevorden F: **Desmoid tumors: need for an individualized approach.** *Expert Rev Anticancer Ther* 2009, **9**(4):525–535.
11. Salas S, Dufresne A, Bui B, Blay JY, Terrier P, Ranchere-Vince D, Bonvalot S, Stoeckle E, Guillou L, Le Cesne A, et al: **Prognostic factors influencing progression-free survival determined from a series of sporadic desmoid tumors: a wait-and-see policy according to tumor presentation.** *J Clin Oncol: official journal of the American Society of Clinical Oncology* 2011, **29**(26):3553–3558.
12. Huang K, Fu H, Shi YQ, Zhou Y, Du CY: **Prognostic factors for extra-abdominal and abdominal wall desmoids: a 20-year experience at a single institution.** *J Surg Oncol* 2009, **100**(7):563–569.
13. Garbay D, Le Cesne A, Penel N, Chevreau C, Marec-Berard P, Blay JY, Debled M, Isambert N, Thyss A, Bompas E, et al: **Chemotherapy in patients with desmoid tumors: a study from the French Sarcoma Group (FSG).** *Ann Oncol: official journal of the European Society for Medical Oncology / ESMO* 2012, **23**(1):182–186.

doi:10.1186/1471-2474-14-69

**Cite this article as:** Ling et al.: Desmoid tumor of posterior cruciate ligament of the knee: a case report. *BMC Musculoskeletal Disorders* 2013 **14**:69.

**Submit your next manuscript to BioMed Central and take full advantage of:**

- Convenient online submission
- Thorough peer review
- No space constraints or color figure charges
- Immediate publication on acceptance
- Inclusion in PubMed, CAS, Scopus and Google Scholar
- Research which is freely available for redistribution

Submit your manuscript at  
www.biomedcentral.com/submit

