CASE REPORT

Total patellectomy with extensor mechanism reconstruction following pathological fracture due to patellar Ewing's sarcoma

Eyad Alqasim,¹ Abdulla Aljowder,² Naji Alammari,³ Amani A Joudeh⁴

¹Orthopaedic Surgery, King Hamad University Hospital, Muharraq, Bahrain

²Orthopaedic Surgery, Bahrain Defence Force Royal Medical Services, Riffa, Bahrain

³Orthopaedic Surgery, King Fahad Specialist Hospital, Dammam, Saudi Arabia

⁴Pathology and Laboratory Medicine, King Fahad Specialist Hospital, Dammam, Saudi Arabia

Correspondence to

Dr Eyad Alqasim,
dreyadalqasim@hotmail.com

Accepted 23 January 2018

SUMMARY

A young patient presented with a progressive right knee pain and swelling that was worse on ambulation. Following appropriate investigations, he was confirmed having Ewing's sarcoma of the right patella. Two cycles of chemotherapy were initiated before he sustained a fall and a resultant pathological patellar fracture. He finally underwent a total right patellectomy and extensor mechanism reconstruction using Achilles tendon allograft in an attempt to eradicate the disease and salvage his knee extensor mechanism. At 1-year follow-up, no disease recurrence was observed, and the patient had a full range of motion but with slight weakness of the extensor mechanism.

BACKGROUND

Patients with anterior knee pain due to a tumour may suffer from delay in diagnosis due to, in our opinion, rarity of patellar tumours, overlooking the radiographic changes and maintaining a low index of suspicion. Such delay may have devastating results especially in patients below 40 as primary tumours of the patella are more common than metastatic tumours in this age group making their diagnosis even harder.

It is very rare to see Ewing's sarcoma affecting the patella. For this reason, it is very important to properly investigate any suspicious lesions seen on the initial X-ray radiographs.

Ewing's sarcoma may be treated successfully by chemotherapy. However, tumours in such locations as the patella, where the tumour may be resected, may also benefit from a combination of chemotherapy and surgical resection.

CASE PRESENTATION

This is a case of a 24-year-old man who was referred to our outpatient clinic for progressive right knee pain and swelling for about 1 year. The pain was noted mostly on ambulation and the swelling progressively increased in size. He also had intermittent fever without any other associated infection. He did not recall any history of trauma to the right knee. Physical examination revealed localised tenderness together with diffuse, hard and non-mobile swelling in the right patella, and no neurovascular deficits. The range of motion was full and not associated with any pain. Weight-bearing was also normal but painful.

INVESTIGATIONS

Initial X-ray films revealed a right patellar lytic lesion with streaks of sclerosis and cortical destruction of the inferior pole (figures 1–3). MRI with contrast was also done demonstrating increased signal intensity in the middle and lower parts of the patellar substance with inferior pole cortical discontinuity suggesting lower pole cortical breach, but without any evidence of soft tissue involvement (figures 4 and 5). These findings correlated with a contained heterogenic lesion that was measured 4.2×2.3×3 cm in size. A whole body scan was performed showing isolated increased uptake in the right patella with no other body part involvement (figure 6). Finally, CT of the lungs and abdomen confirmed no metastasis. A suspected preliminary diagnosis of giant cell tumour versus chondroblastoma and osteosarcoma was initially considered and, accordingly, the patient was planned for an open bone biopsy.

Histopathological examination demonstrated infiltration by sheets of relatively small round blue cells. The infiltrate was destructive and occupied the spaces between the patellar bone trabeculae (figure 7). The cells had monomorphic round nuclei, indistinct nucleoli, slight nuclear indentations and a rim of clear-to-eosinophilic cytoplasm (figure 8). Furthermore, well-controlled immunohistochemical stains were performed using antibodies against the following antigens; CD99, CD56, desmin, Myo-D1, pancytokeratin, vimentin, CD45, S-100, epithelial membrane antigen (EMA) and B-cell lymphoma 2 (Bcl-2). The neoplastic cells showed diffuse and strong positivity for CD99 and vimentin and somewhat weaker positivity for pancytokeratin and Bcl-2 (figure 8). Additional cytogenetic analysis revealed rearrangement involving the EWSR1 gene at 22q12 (figure 9). A definitive diagnosis of Ewing's sarcoma of the right patella was, therefore, finally made.

DIFFERENTIAL DIAGNOSIS

Giant cell tumour, chondroblastoma, osteosarcoma.

TREATMENT

The case was discussed in the tumour board, and a decision to refer the patient to the medical oncology team for chemotherapy sessions was taken. Two cycles of etoposide-vincristine-adriamycin (doxorubicin)-ifosfamide-actinomycin D

To cite: Alqasim E, Aljowder A, Alammari N, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-222853

Figure 1 X-ray right knee—anteroposterior view. Poorly demarcated patella with multiple lucencies and some streaks of sclerosis.

(EVAIA) chemotherapy regimen over 2 weeks were started with no post-therapy complications.

However, 2 weeks later, the patient presented again to the emergency room after falling directly on his right knee, sustaining an avulsion fracture of the lower pole of the right patella ([figure 10](#)). No further extensive investigations such as MRI, bone scan or CT were performed as such investigations were carried out recently; and one would not expect to see any radiological evidence of contamination or seeding in the acute post-trauma stage. Considering the patient's history of Ewing's sarcoma together with the newly superimposed patellar fracture, a final decision to perform a total patellectomy was taken.

Figure 2 X-ray right knee—lateral view. Lytic lesion extending from the superior to the inferior poles of the patella. There is blurring of the inferior pole cortex which suggests cortical breach by the lesion.

Figure 3 X-ray right knee—sunrise view. Sharp and well-demarcated superior pole cortex indicating sparring of the superior pole of the patella. The extent of the lysis may also be appreciated.

Intraoperatively, the knee was approached through an anterior incision. The surrounding soft tissues were grossly examined and found to be healthy with no evidence of necrosis, fibrosis, calcification or haemorrhage. The patellar fragments were also excised completely and examined. Apart from the slight irregularities of the inferior pole and some scattered areas of weakened consistency, no other gross pathological findings were observed ([figure 11](#)). However, the extensor mechanism was noticed to be deficient and short; therefore, an Achilles tendon allograft was used for its reconstruction ([figures 12 and 13](#)). The patellar specimen was finally sent for histological examination, further confirming the diagnosis ([figure 14](#)).

OUTCOME AND FOLLOW-UP

The right knee was initially immobilised in a knee brace in extension for 2 weeks postoperatively. Thereafter, passive range of

Figure 4 Coronal MRI right knee. Increased signal intensity of the right patella with permeative moth-eaten appearance.

Figure 5 Sagittal MRI right knee. Increased signal intensity of the patella with heterogenic lesion (large arrow). The lower pole of the patella cannot be clearly demarcated indicating destruction of the lower pole (small arrow).

motion started at 2 weeks. At 4 weeks, gradual active-assisted and active range of motion were started. Chemotherapy was resumed postoperatively and continued for 12 months. Routine follow-ups were carried out initially at 2 weeks and then at 1, 3, 6, 9 and 12 months postoperatively. The patient is scheduled regular follow-ups every 3 months up to date with X-rays and MRIs performed regularly. Recent clinical and radiological examinations revealed completely eradicated disease with no evidence of recurrence. The knee range of motion extends from 130° of flexion to an expected lagging 15° of extension.

DISCUSSION

The cell origin of Ewing's sarcoma remains unknown. However, neural crest and mesenchymal stem cell origins have been

Figure 6 Whole body scan. There is isolated increased uptake in the right patella confirming no other involvement of the skeleton.

Figure 7 Pathological specimen. Diffuse infiltration by sheets of proliferating round uniform cells with entrapped patellar bone trabeculae.

suggested.¹ Ewing's sarcoma is a tumour primarily of childhood and adolescence. Its incidence is highest among patients between the ages of 10 and 19 years with an incidence rate of 9–10 cases per million per year in USA.¹ It is the second most common primary malignant tumour in children and adolescents following osteosarcoma with a slightly higher male predominance, that is, male-to-female ratio of 1.5:1.¹

Most common tumours affecting the patella in patients less than 40 include giant cell tumour, chondroblastoma, aneurysmal bone cyst, osteoid osteoma and solitary bone cyst; while in patients over 40, metastatic tumours are the most common.² To our knowledge, there were only two previously published cases describing Ewing's sarcoma involving the patella.

Ewing's sarcoma affects the diaphyseal or metadiaphyseal regions.¹ The most common location affected is the pelvis, followed by the femur and tibia.³ The tumour spreads by local infiltration into the medullary canal, cortex and the surrounding soft tissues, and by haematogenous spread to the lungs (57%) and bones (34%).⁴

Clinical features include progressive pain and swelling. Fever may also be present, a situation that has to be differentiated from

Figure 8 High-power histological specimen. Neoplastic cells with round nuclei, small indistinct nucleoli, clear-to-eosinophilic cytoplasm with indistinct cellular borders. The neoplastic cells are strongly and diffusely positive for CD99.

Figure 9 EWSR1 dual-colour fluorescence in situ hybridisation using formalin-fixed paraffin-embedded tissue. Cells with significant rearrangement involving the EWSR1 gene region (arrows)

osteomyelitis. The authors, undeniably, agree that these symptoms are not very well specific for the disease; and completely agree with Gorelik *et al* that Ewing's sarcoma constitutes an uncommon aetiology of anterior knee pain.³ However, the presence of progressive swelling together with unexplained fever, combined with proper radiological interpretation, should definitely raise the suspicion of some pathology.

Initial diagnosis starts with X-rays which reveal destructive bony lesion that may either be purely lytic or with variable amounts of reactive new bone formation. Other features include the so-called 'onion-peel effect' that results from the tendency to produce new bone lifting the periosteum into multiple layers.⁴ Extension into the surrounding soft tissue producing a 'sunray' effect and proximal and distal marginal periosteal reaction producing the Codman triangle may also be seen. MRI may reveal extraosseous component of the tumour, and bone scan may be used to demonstrate any increased uptake.⁴ In our patient, soft tissue extension was, indeed, initially suspected

Figure 10 Lateral view X-ray of right knee. Avulsion fracture of the lower pole of the patella.

Figure 11 Total patellectomy.

owing to the inferior pole cortical breach that was observed on the initial X-ray films. The lesion, however, was confirmed to be contained by the subsequent MRI examination. Finally, as with other tumours, the diagnosis may be confirmed with histopathological studies.

Treatment involves a multimodality approach to completely eradicate the disease. Generally, the mainstay of treatment involves chemotherapy.⁵ Although Ewing's sarcoma is very radiosensitive, radiotherapy is used less frequently due to the risk of secondary malignancies and the adverse effects on bone.⁵ The introduction of multiagent chemotherapy has increased the 5-year survival rate to 65%.¹ Treatment regimen for resectable Ewing's sarcoma involves neoadjuvant chemotherapy followed by a limb-salvage procedure and postoperative adjuvant chemotherapy.⁶ Valsalan and Zacharia⁷ well described this approach in their paper. However, they also described a novel technique whereby the extensor mechanism was reconstructed using a split tendo-Achilles autograft.

Local control of the tumour remains controversial.^{5,6} If the tumour is resectable with wide margins, no preoperative radiotherapy is used.⁶ If, however, the certainty of wide margin excision cannot be determined, preoperative radiotherapy should be added.⁶ If the margins are found to be inadequate postoperatively, radiotherapy may also be added thereafter.⁶

In our patient, we initially felt it would be sufficient to only subject our patient to chemotherapy alone to achieve a complete eradication of the tumour yet preserving the extensor mechanism and full knee stability and function. This was due to, among other reasons, the fact that the disease was contained without any dissemination to the adjacent bony or soft tissues, and any initial patellectomy would raise the challenge of reconstructing the extensor mechanism. Other considerations were also given to the young age and the possibility to allow this patient to have a full function of his knee.

Figure 12 Achilles tendon grafting of the extensor mechanism.

Figure 13 Reconstruction of the extensor mechanism by an Achilles tendon graft.

Following case discussion in the tumour board, however, the decision to follow the usual recommendations by initiating treatment with a 12-week preoperative chemotherapy followed by total patellectomy was taken. However, the premature complication of the superimposed pathological patellar fracture has completely changed the initial treatment plan. Any thoughts to fixation of the fractured patella would, indeed, be inappropriate as the fixation itself would neither be rigid nor stable. At that stage, we regarded the tumour resectable; therefore, a total patellectomy was performed and a long-term chemotherapy was continued thereafter.

Figure 14 The excised right patella.

One may argue that the tumour may have rendered the patella very weak, increasing its susceptibility to pathological fractures; therefore, a complete patellectomy should have been the first choice of treatment. The authors have initially given thoughts to the possibility of such complication. However, the general recommended standard of care in most patients involves a combination of neoadjuvant chemotherapy and a limb-salvage procedure, especially in expendable bones.⁵ This is because Ewing's sarcoma is very chemosensitive; and neoadjuvant chemotherapy helps reduce the size of the primary tumour, thereby facilitating easier resection and reducing the risk of recurrences.⁸ Other benefits of neoadjuvant chemotherapy include the possibility to evaluate the efficacy of the chemotherapeutic agents before any limb-salvage procedure is undertaken.⁸

Learning points

- ▶ Anterior knee pain may represent a more serious pathology. Therefore, appropriate utilisation of patient history and physical examination is a crucial step.
- ▶ The presence of constitutional symptoms, such as fever, should increase the index of suspicion of a more serious pathology.
- ▶ Radiological features should never be overlooked.
- ▶ Various regimens exist for the treatment of Ewing's sarcoma. Treatment plans should be tailored to patient-specific and tumour-specific factors in order to provide the best outcome.

Contributors EA raised the idea of writing the article. He identified the case and was involved in the clinical and operative management of the patient. He made substantial contribution in the research, writing, analysis, interpretation and final approval of the work. He is also the guarantor of the paper. AA was involved in the research of the work. NA made substantial contribution in the interpretation of the work. AAJ provided extensive pathological analysis of the specimens which she later reviewed many times and summarised to fit in the article appropriately. She also provided high quality images of the slides and made substantial contribution in the research of the work.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2018. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Choi EY, Gardner JM, Lucas DR, *et al.* Ewing sarcoma. *Semin Diagn Pathol* 2014;31:39–47.
- 2 Singh J, James SL, Kroon HM, *et al.* Tumour and tumour-like lesions of the patella—a multicentre experience. *Eur Radiol* 2009;19:701–12.
- 3 Gorelik N, Dickson BC, Wunder JS, *et al.* Ewing's sarcoma of the patella. *Skeletal Radiol* 2013;42:729–33.
- 4 Desai KI, Nadkarni TD, Goel A, *et al.* Primary Ewing's sarcoma of the cranium. *Neurosurgery* 2000;46:62–9.
- 5 Jain S, Kapoor G. Chemotherapy in Ewing's sarcoma. *Indian J Orthop* 2010;44:369–77.
- 6 Iwamoto Y. Diagnosis and treatment of Ewing's sarcoma. *Jpn J Clin Oncol* 2007;37:79–89.
- 7 Valsalan RM, Zacharia B. Ewing's sarcoma of patella: A rare entity treated with a novel technique of extensor mechanism reconstruction using tendoachilles auto graft. *World J Orthop* 2015;6:744–9.
- 8 Goto T, Okuma T, Nakada I, *et al.* [Preoperative adjuvant therapy for primary malignant bone tumors]. *Gan To Kagaku Ryoho* 2007;34:1750–4.

Copyright 2018 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow