

Sinding-Larsen-Johansson syndrome

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A spectrum of entities is involved in injury to the inferior aspect of the patella and the proximal patella tendon, including Sinding-Larsen-Johansson syndrome, patellar sleeve avulsion and jumper's knee. The patellar tendon is usually only a few centimetres long, arising from the inferior patella and inserting distally into the tibial tuberosity. Sinding-Larsen-Johansson syndrome is an osteochondrosis of the inferior pole of the patella and is often bilateral. It is not osteonecrosis, epiphysitis or osteochondritis as previously described in the literature. Sinding-Larsen-Johansson is commonly seen in active adolescents aged between 10 and 14 years, as described in our patient. Our case highlights the importance of an adequate history, clinical examination combined with correct imaging in accurate, early diagnosis of Sinding-Larsen-Johansson syndrome, a rare but important course of patellofemoral pain.

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History and clinical findings

A 12-year-old boy presented to a paediatric rheumatology clinic with a 2-month history of painful knees. He had been referred from the paediatric oncology department with a previous diagnosis of acute lymphocytic leukaemia, in remission for 5 years. The pain was felt just below the patella, especially after a game of soccer, climbing stairs or walking for long distances. There was associated swelling of both knees, but no early-morning stiffness or deformities were described. Clinical examination revealed bilateral mild knee effusions with tenderness over the infrapatellar area in the region of the infrapatellar ligament. A prominence of the infrapatellar area was noted during flexion of the knee. The child was unable to assume a kneeling position due to pain over the knees. No ligamentous or meniscal instability was noted. There was no evidence of synovitis of any of the joints or evidence of an inflammatory arthropathy.

Imaging

Lateral radiographs of bilateral knees demonstrate mild beaking of the inferior pole of the patella (normal variant) (Fig. 1). There is

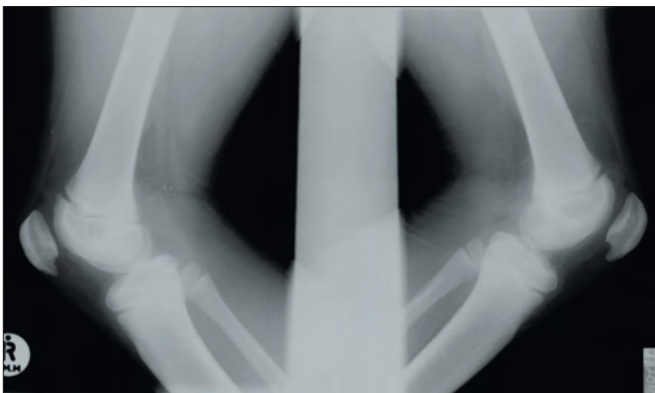


Fig. 1. Lateral radiographs of bilateral knees demonstrate mild beaking of the inferior patellar pole (normal variant). There is no significant peripatellar soft-tissue swelling, nor are there osseous fragments adjacent to the inferior pole of the patella.

no significant peripatellar soft-tissue swelling or osseous fragments adjacent to the inferior pole of the patella.

MRI of bilateral knees, sagittal T2-weighted, fat-suppression sequence, demonstrate hyperintense signal in the inferior pole of the patella, proximal and posterior part of the patellar ligament and surrounding soft tissues (Fig. 2). This correlates with hypo-intense signal in these anatomical areas on T1-weighted MRI.

Diagnosis and management

A diagnosis of Sinding-Larsen-Johansson syndrome (osteochondrosis of the inferior pole of the patella) was made. The patient was treated with naproxen 250 mg twice daily and was asked to stop



Fig. 2. Sagittal T2-weighted MRI of the left knee, fat-suppression sequence, demonstrating hyperintense signal in the inferior pole of the patella, proximal and posterior part of the patellar ligament and surrounding soft tissues, in keeping with Sinding-Larsen-Johansson syndrome.

playing soccer. At a follow-up examination 2 months later the patient's symptoms had resolved.

Discussion

The extensor mechanism of the knee comprises the quadriceps muscles and tendon, the patella, the patellar ligament and the supporting retinaculum. Injuries to the extensor mechanism are common and may be due to acute trauma, overuse injuries or chronic degenerative disease.¹

A spectrum of entities is involved in injury to the inferior aspect of the patella and the proximal patellar ligament, including Sinding-Larsen-Johansson syndrome, patellar sleeve avulsion and 'jumper's knee'.²

The patellar ligament is usually only a few centimetres long, arising from the inferior patella and inserting distally into the tibial tuberosity.² Patellar tendinopathy (jumper's knee) is a common overuse condition that classically occurs in young athletes who jump, kick and run, placing repeated stress on the patellofemoral joint. It includes a spectrum of pathology from chronic degeneration to partial tearing.¹

In paediatric patients, patellar sleeve avulsion fractures are the most common patellar fractures, although they are rare. A full circumference of cartilaginous tissue and often a bony fragment is avulsed from the lower pole of the patella. The injury most often occurs with jumping, and skateboarding is a common cause. On plain radiographs, an abnormally superiorly displaced patella and/or a small bone fragment distal to the lower pole of the patella can provide a clue to diagnosis.¹

Sinding-Larsen-Johansson syndrome is an osteochondrosis of the inferior pole of the patella and is often bilateral in nature. It is not osteonecrosis, epiphysitis or osteochondritis, as previously described in the literature.³ The syndrome is caused by traction on the patellar ligament, causing inflammation at the insertion of the proximal ligament into the inferior pole of the patella.¹

Sinding-Larsen-Johansson syndrome is commonly seen in active adolescents between 10 and 14 years of age, as described in our patient.⁴ It should be distinguished from patellar sleeve avulsion, which is due to a cartilaginous injury to the lower pole of the patella.² MRI is necessary to distinguish this entity from Sinding-Larsen-Johansson syndrome, which has a similar radiographic appearance but represents a pure osseous injury without the extensive cartilaginous involvement seen with patellar-sleeve avulsion.² Sinding-Larsen-Johansson syndrome can be distinguished from patellar tendinopathy by the presence of bone-marrow oedema in the patella.¹

The mechanism of injury is similar in all three of the entities and is believed to be due to forceful contraction of the quadriceps against resistance, particularly in adolescent male athletes.² Cerebrospastic children are also predisposed. There is no known association between Sinding-Larsen-Johansson syndrome and malignancy as described in our patient. Presentation is with point tenderness at the inferior pole of the patella, associated with focal swelling.⁴

Clinically this condition should be distinguished from Osgood-Schlatter disease, which is due to osteochondrosis of the patellar ligament insertion into the tibial tuberosity.⁵ This disease is also a common cause of knee pain in adolescents and is caused by repeated traction on the immature tibial tuberosity by the patellar ligament, which can cause inflammation, avulsion fractures and excess bone growth. It can be suggested on plain radiographs if radiographic



Sinding-Larsen-Johansson syndrome is a rare but important cause of patellofemoral pain.

soft-tissue swelling is present anterior to the tibial apophysis, as fragmentation of the tibial apophysis can be a normal variant.¹

Osteochondrosis of the tibial tuberosity (Osgood-Schlatter disease) and osteochondrosis of the patella (Sinding-Larsen-Johansson syndrome) occur at tendinous insertions into the patella at the distal and proximal levels, respectively. The dynamic stress and microtrauma due to the active function of the ligament are responsible for the onset of both these disorders. For this reason, these osteochondroses of the knee have also been called 'non-articular osteochondroses', since they occur in ossification centres that are submitted to traction, not to compression stress.⁶

Other pathologies to consider in an adolescent with knee pain, tenderness and disability of the knee joint (of the extensor mechanism) include patellar ligament rupture, quadriceps tendinopathy, quadriceps tendon rupture, patellar dislocation, chondromalacia patellae and prepatellar bursitis.¹

Imaging of Sinding-Larsen-Johansson syndrome may require a combination of radiographs, MRI and ultrasound. Lateral radiographs may reveal peripatellar soft-tissue swelling, patella-alta deformity and one or multiple tiny osseous fragments adjacent to the inferior pole of the patella.² There may be stranding of adjacent portions of Hoffa's fat pad.⁴ The exact extent of damage is frequently underestimated on radiographs and early imaging findings may be subtle or absent,⁴ as described in our patient, necessitating further evaluation with MRI.² On MRI, the inferior pole of the patella, proximal and posterior part of the patellar ligament and surrounding soft tissues are hypointense on T1-weighted MRI sequences and hyperintense on T2-weighted MRI (fat-suppression) sequences.⁴

According to De Flaviis *et al.*,⁶ ultrasound images are either equally effective as, or more effective than, radiographs, especially when evaluating the soft tissue. Ultrasound is proposed as a simple and reliable method for diagnosing knee-joint osteochondrosis, especially during the early stages of the disease. Ultrasound is also suitable for periodic follow-up in the course of the disease.⁶ On ultrasound, the lower pole of the patella appears fragmented and hypochoic with swelling of the cartilage, in particular at the insertion of the patellar ligament. Follow-up examinations demonstrate progressive consolidation of the affected bone.⁶

The age of the patient and the presence of tenderness and localised swelling at the inferior pole of the patella are characteristic of the disorder. Radiographs are often obtained for the first evaluation of the disease, but high-resolution ultrasound of the knee is an effective and reliable first-line technique for the diagnosis of this osteochondrosis. The sonographic picture yields the same diagnostic information as the radiograph, namely the development, structure, and profile of the ossification centres, but also provides a clear picture of the superficial soft tissues and of the non-ossified cartilage.⁶

Distinguishing between the spectrum of entities involving injury to the inferior aspect of the patella and the proximal patellar ligament is important with regard to patient treatment. Minimally displaced fractures as those seen with Sinding-Larsen-Johansson syndrome are often managed conservatively and non-operatively.² Initial treatment consists of relieving the pain by resting for a few days and strengthening exercises with modification of activities. Non-steroidal anti-inflammatory drugs (NSAIDs) may be necessary, and in severe cases a cast is used for maintaining immobility.⁵ Our patient's symptoms improved with NSAIDs and modification of his activities.

Displaced patellar sleeve avulsion fractures are treated with open reduction and possible internal fixation as well as extensor mechanism reconstruction.²

Conclusion

This case highlights the importance of an adequate history and clinical examination combined with correct imaging in accurate, early diagnosis of Sinding-Larsen-Johansson syndrome, a rare but important cause of patellofemoral pain.

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