A 59-year-old woman presented to the orthopaedics outpatient clinic of our hospital due to complaints of swelling and pain anteriorly of the left knee since three years. Movement of the knee was not impaired. The patient's mother had morbus Paget, the family history was otherwise unremarkable.

**Imaging Findings:**

A CT and gadolinium enhanced MRI were performed. CT showed a diffusely enlarged patella, with coarse bone trabeculae and cortical thickening. Centrally in the patella there were several small sclerotic and lytic focal areas. (Figures 1 and 2). On MR images the thickened cortex was iso-intense on T1, had a high signal intensity on T2-weighted images, and enhanced strongly after gadolinium. Centrally, the lesion had mixed signal intensities on MRI. The patellar lesion did not extend into the surrounding soft tissue. Retropatellar chondropathy was present. (Figures 3-6)

**Discussion:**

**BACKGROUND**

Paget disease (PD) is a bone metabolic disorder with a disturbed osteoblast and osteoclast equilibrium that changes bone architecture.[1] The disease was first described in 1877 by Sir James Paget, and it was called “osteitis deformans” at the time. The epidemiology of PD appears to be changing rapidly: several groups in different parts of the world reported a marked reduction in the prevalence and incidence of PD, as well as in the severity of disease seen by clinicians. These findings seem most likely to be caused by changes in exposure to (partly still unknown) environmental factors that have a role in the development of PD.[1] However, paramyxovirus has been put forward as a possible infectious cause.[2] Genetic factors are also important: mutations in SQSTM1 occur in 25-50% of familial PD.[1] The disease incidence increases with age, and the male to female ratio is 2:1. Three phases are described in PD: the first being an osteolytic phase (“hot” phase), the second a mixed lytic and blastic phase, and the third a sclerotic phase. Typically, bone expansion, cortical thickening and trabecular bone thickening are present in the second phase. In 1% of patients, a malignant transformation to osteosarcoma or giant cell tumour can take place. Treatment with biphosphonates can be applied to decrease the osteoclast activity and bone resorption.[2] The most affected bones in PD are the skull, spine, pelvis and femur. Usually the disease is polyostotic. When only one bone is affected, this type is called monostotic PD. PD of the patella is very rare. It has been previously
described in a review by Ploumis et al. [3] To our knowledge, there have been 4 case-reports previously published on the topic.[4-7]

OUTCOME
After the MRI and CT had been done, a CT-guided biopsy was performed to confirm the radiological suspicion of PD (Figure 7). Pathology showed coarsening of the bone trabeculae, an increased number of osteocytes and focally increased osteoblastic activity. The diagnosis of a “Paget-like reaction” with a differential diagnosis including PD, posttraumatic changes or changes due to an inflammatory reaction was made. The patient had experienced a trauma several years ago. To clarify the diagnosis, a total body bone scintigraphy was performed which showed intense increased uptake of the radiopharmacon (99 Technetium) by the left patella without other suspect lesions, (Figure 8) consistent with PD.

Differential Diagnosis List: Morbus Paget of the patella, Chronic osteomyelitis, Reactive post-traumatic changes, Metastasis

Final Diagnosis: Morbus Paget of the patella

References:
Description: Note the diffusely enlarged patella, with coarse bone trabeculae and cortical thickening.
Origin: Radiology, LUMC.
Description: Small sclerotic and lytic focal areas are seen centrally in the patella. Origin: Radiology, LUMC.
Description: Strong enhancement of the patellar cortex is seen, with mixed signal intensities centrally in the patella. Origin: Radiology, LUMC.
**Description:** Besides the strong enhancement of the patellar cortex, there is no surrounding tissue enhancement. **Origin:** Radiology, LUMC.
Description: An iso-intense to high signal of the patellar cortex is seen. Origin: Radiology, LUMC.
Figure 6

Description: The biopsy needle is shown, entering the left patella from medially. Origin: Radiology, LUMC.
**Description:** Focal high uptake in the left patella is present, without any other high uptake areas.

**Origin:** Radiology, LUMC.
Description: An iso-intense signal of the patellar cortex is seen on T1-weighted images. Origin: Radiology, LUMC.