Case 3075

Massive osteolysis of the hip: Gorham's disease
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Patient: 73 years, female

Clinical History:

A 73-year-old female patient presented with unremarkable history and progressive pain in her left hip. A clinical examination was done, which revealed a decreased range of motions in her hip extension, abduction and external rotation. Plain films of her hip were taken, which showed an extensive osteolysis of femoral head, neck and acetabulum, and soft tissue swelling. No collapse, fragmentation or sclerosis of the residual bone was noted.

Imaging Findings:

The patient with an unremarkable history was admitted to our hospital because of mild pain in her left hip. On clinical examination, it was found that the joint was normal and the X-ray evaluation showed only minimal joint space narrowing. Two months later, the patient returned complaining of severe pain. She had an abnormal gait, limp, a decreased range of motions in her hip extension, abduction and external rotation. Plain films of the hip showed an extensive osteolysis of femoral head, neck and acetabulum and soft tissue swelling. No collapse, fragmentation or sclerosis of the residual bone was noted. The patient was investigated and underwent, a CT scan of abdomen and the pelvis and MRI of the hips. The results of the CT scan showed an extensive osteolysis of the acetabular roof and the femoral head, the femoral neck erosion and an intra-articular growth of the soft tissue process. On MRI, both soft tissue and osseous structures of the joint showed a low signal intensity on T1-weighted sequences, a very high signal intensity on T2-weighted sequences and a strong contrast enhancement. These findings were consistent with the presence of a hypervascular infiltrating lesion, possibly of an inflammatory origin, during which time Gorham's disease was suspected. During her hospitalization, the patient developed a septic syndrome with pyrexia, leucocytosis and positive blood cultures. A physical examination done revealed the presence of an ulcerative lesion on her scalp that was considered to be the entry point. A bacterium named Staphylococcus aureus was isolated. Hip aspiration was performed but the cytology was not conclusive. The patient was managed with antibiotics for six weeks successfully. Five months following the onset of symptoms, the results of the repeated radiography showed the progression of osteolysis with tapering of the bone remnants, which is a finding considered to be the characteristic feature of Gorham's disease. Six months later, a total hip arthroplasty was performed. During two years followup, the patient had no pain ,he had a normal gait and his daily activities found to be normal.

Discussion:

Our case refers to a unilateral rapid hip destruction in an old woman. The patient's age and history, as well as the initial X-ray findings were highly suggestive of a rapid destructive arthritis, which is an uncommon form of osteoarthritis. Radiologically, there is a progressive and rapid destruction of the joint in the absence of osteophytes
as is the case with the classic form of osteoarthritis. Avascular necrosis of the femoral head was also a possible diagnosis, but the patient had no risk factors and the characteristic imaging features of the disease such as sclerotic or cystic lesions as well as the subchondral collapse were absent. The calcium pyrophosphate dehydrate deposition disease and the apatite-associated destructive arthritis due to calcium hydroxyapatite crystal deposition could be another possible diagnosis, but the examination of the tissues and the joint fluid was not pathognomonic. Arthritis of neuropathic disorders was also excluded based on the absence of neuropathy and the lack of the characteristic exuberant osteophytes and the free intra-articular bony fragments. Analgesics induced arthropathy was not considered possible because the patient used analgesics only occasionally. Finally, infectious arthritis and neoplastic osteolytic process were the major considerations in our differential diagnosis but both were excluded after a thorough investigation. Massive osteolysis of Gorham is a condition of an unknown etiology, which includes a combination of clinical, radiological and histological findings. It is characterized by the replacement of normal trabeculae by intensively developing benign vascular tissue, which resembles hemangiomatosis or lymphangiomatosis and results in a complete or partial osteolysis. It affects any bone but is mainly localized in the pelvis, the shoulder or the mandible. The mechanism of bone destruction and resorption in Gorham's disease is still unknown. The initial X-ray findings showed the presence of ill-defined subcortical or marrow lytic lesions, which have a tendency to enlarge. In the majority of cases, there is an absence of sclerosis. Later in the disease process, these lesions are expanded eccentrically with the formation of the characteristic tapering edges of the affected bones and the complete bone resorption. It should be emphasized that the patients have mild symptoms compared to the radiological appearance. Even though the disease is benign, it can expand to the adjacent bones or soft tissues. Usually the condition is self-limited, but its outcome is not predictable. In the case presented by us, the characteristic finding was the presence of excessive lytic bone lesions of the femoral head, neck and the acetabulum, causing joint space pseudowidening and not narrowing as it is expected in the above mentioned conditions. The rapidly progressive osteolysis of the femoral head and the neck leading to bone tapering, as appeared in the serial radiographs, is a characteristic feature of Gorham's disease. The aggressive type of osteolysis in our case had radiological features of neoplastic or infectious lesions. The intraosseous and intra-articular growth of soft tissue mass, as it was shown on CT and MRI studies of our patient, has been reported as suggestive of Gorham's disease diagnosis. A similar process can occur in infectious and neoplastic conditions as well, although with different patterns of extension. The extremely high signal intensity on T2-weighted sequence and the rigorous contrast enhancement of the soft tissue are indicative of its angiomatous nature. The histological examination of the specimens (seven months after the MRI and eleven months after the onset of symptoms) detected mainly fibrous tissue and minimal angiomatous tissue, which were not expected according to MRI findings. This can be explained by the time elapsed between the MRI and the histologic examination. There are two stages of osteolysis in Gorham's disease. The first is characterized by a vascular proliferation in the connective tissue, which is consistent with hemangiomatosis, and the second is characterized by the replacement of the resorbed bone by a fibrous tissue as it was in our case. One should take into consideration its limited growth potential, which leaves the cortex intact without expanding in the adjacent soft tissues. Gorham's disease should be differentiated from skeletal hemangiomatosis. Limited growth potential, intact cortex and the absence of extraosseous extension characterize the latter as opposed to Gorham's disease. Regarding the treatment of this disease, radiotherapy, anti-osteoclastic treatment (biphosphonates), or even specific hemangiomatosis treatment has been proposed. In the case presented by us, total hip arthroplasty was considered as the most appropriate option. Due to a favorable outcome, any further treatment was not required.

**Differential Diagnosis List:** Gorham's disease.

**Final Diagnosis:** Gorham's disease.

**References:**

Assoun J, Richardi G et al. CT and MRI of massive osteolysis of Gorham. J. of Computer Assisted Tomography
Description: Extensive osteolysis and soft tissue swelling are noticed. Origin:
Figure 2

Description: The characteristic tapering of the residual femoral neck is noticed. Origin:
Description: Osteolysis and the intra-articular soft-tissue process are shown. Origin:
Description: The lesions showing homogeneously high-signal intensity. Origin:
Figure 5

Description: The lesions showing a homogeneously high-signal intensity.

Origin: