

Unexpected Revelation of Hip Amyloidosis Following Pelvic Trauma: A Case Report

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Abstract

Recent advancements in dialysis therapy have significantly improved the lives of individuals with declining renal function. This is particularly important in countries like Japan, where kidney transplantation is not widely accepted or frequently performed.

By the end of 1999, 197,231 individuals were receiving dialysis in Japan, with over 21,150 having been on dialysis for a year or more. The number of long-term dialysis patients has steadily increased. The three leading causes of death among these patients are cerebrovascular disease, heart failure, and infection.

One of the most common complications affecting the quality of life for long-term dialysis patients is dialysis-related amyloidosis (DRA). DRA affects 25.6% of all Japanese dialysis patients, with the incidence rising with longer treatment durations. It impacts 35.1% of those on dialysis for 10 years, 56.8% for those treated for more than 15 years, and 74.6% for those who have been on dialysis for over 20 years.

Keywords: Amyloidosis; Hip; Beta 2-Microglobulin; MRI

Introduction

Osteoarticular amyloidosis (OA) is a serious and often underdiagnosed complication in patients undergoing long-term dialysis, particularly those with chronic kidney disease. It is characterized by the deposition of beta-2 microglobulin amyloid in bones, joints, and periarticular tissues, leading to a variety of musculoskeletal symptoms such as joint pain, stiffness, and functional impairment. As the incidence of end-stage renal disease rises and patients live longer due to advances in dialysis and transplantation, the prevalence of OA has increased. Timely diagnosis through clinical, radiological, and histological assessments is crucial for effective management and the prevention of serious complications.

Case Report

This is a 77-year-old woman with a history of end-stage chronic kidney disease, on hemodialysis for the past 7 years, who has been complaining of hip pain for the last 8 months. She presented to the emergency department following a fall from standing height, with the point of impact on her left hip. Upon examination, there was complete limitation of movement in the left hip, but no fever or other associated signs.

Biologically, inflammatory markers were slightly elevated, while the electrolyte panel and complete blood count were unremarkable. A standard X-ray was initially performed, revealing bilateral lytic lesions of the femoral neck, along with a fracture of the left femoral neck. A CT scan was then performed to obtain 3D bone reconstructions, which confirmed bilateral osteolytic lesions of the femoral neck, as well as the fracture of the left femoral neck. There was also soft tissue infiltration around the area, with pseudo-thickening of the articular synovium (Figure 1).

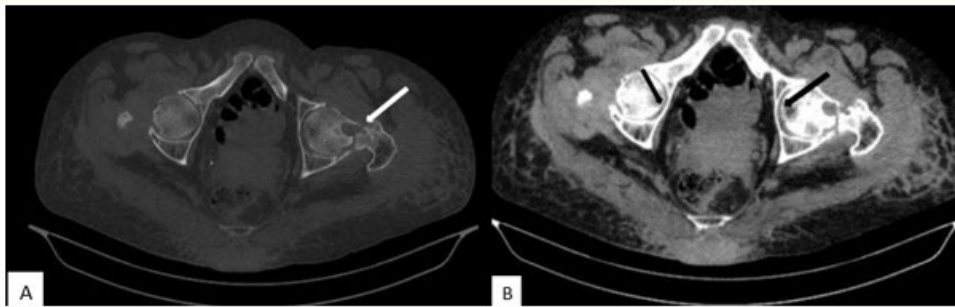


Figure 1: Axial slices of a pelvic CT scan in bone window (A) and soft tissue window (B) showing a fracture line of the left femoral neck passing through the osteolytic lesion, associated with bilateral thickening of the articular synovium.

Given the suspicion of osteoarticular amyloidosis, an MRI of the pelvis was conducted, revealing periarticular pseudo-masses with T1 hypointensity, associated with geodes in the femoral necks with heterogeneous T2 hyperintensity. These lesions appeared to communicate in places with the articular synovium, which was thickened and enhanced after gadolinium injection.

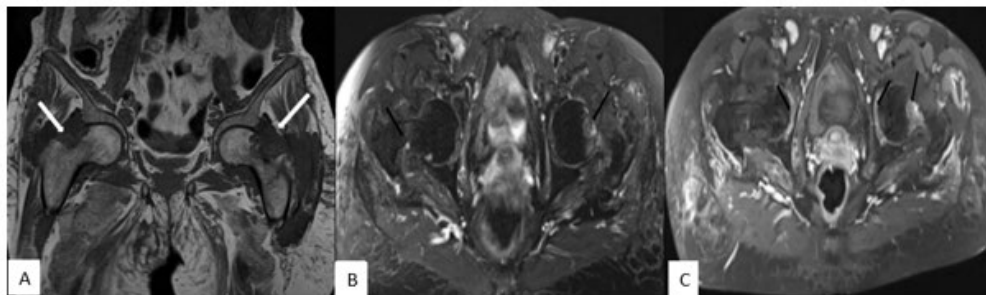


Figure 2: Pelvic MRI images in coronal T1 sequence (A), axial T2 (B), and axial T1 FAT SAT after Gadolinium injection showing bilateral periarticular pseudo-masses with T1 hypointensity, slight heterogeneous T2 hyperintensity, and mild heterogeneous enhancement after Gadolinium injection. Additionally, there are geodes in the femoral necks with heterogeneous T2 hyperintensity, along with thickening of the articular synovium, which is enhanced after Gadolinium injection, consistent with amyloid deposits.

Discussion

Dialysis-related amyloidosis is a common complication in patients with chronic kidney disease undergoing dialysis, particularly affecting the musculoskeletal system. It is caused by the accumulation of beta-2 microglobulin in the blood, typically manifesting after 6 to 9 years of dialysis, with a more consistent occurrence after 20 years of treatment. Its prevalence has increased due to the rise in life expectancy and chronic conditions such as end-stage renal disease, although the use of high-flux membranes in hemodialysis appears to delay its onset. Both men and women are affected equally [1].

Although the musculoskeletal system is the primary site of impact, histological evaluations often reveal more frequent visceral involvement than previously recognized. This visceral involvement can be asymptomatic, but it may also lead to severe complications such as intestinal infarction and heart failure [1,2].

Acquired osteoarticular amyloidosis (AOA) typically begins insidiously, with chronic joint pain, periarticular swelling, and/or carpal tunnel syndrome occurring 7 to 10 years after starting dialysis. Joint pain is the most common symptom, with a preference for the shoulder, followed by the hip, knee, and wrist.

Bone and joint abnormalities, including osteoporosis, lytic lesions, pathological fractures, osteonecrosis, periarticular soft tissue masses, cysts, and subchondral erosions, as well as joint contractures and subluxations, are caused by amyloid deposits in periarticular tissues, synovium, and bones [3]. Spinal involvement manifests as destructive spondyloarthropathy, with severe disc-vertebral lesions and possible neurological signs.

The histological demonstration of beta-2 microglobulin amyloid deposits, obtained via synovial biopsy or during surgical procedures, remains the gold standard for diagnosis. However, non-invasive diagnostic procedures are preferred in clinical practice. Ultrasound and MRI can detect periarticular soft tissue swelling, subchondral cysts, and synovial thickening. The main radiological features of beta-2 microglobulin amyloidosis include subchondral bone cysts and joint erosions. CT scans are useful for accurately assessing the size of bone defects and the risk of fractures, particularly in the femoral neck, knees, and posterior vertebral arches. Bone scintigraphy shows increased isotope uptake in affected joint areas, although this hyperfixation is not specific to dialysis-related amyloidosis and can also be observed in other synovial conditions. Scintigraphic procedures using radiolabeled P component 24 or radiolabeled beta-2 microglobulin can more specifically localize amyloid deposits [4].

A comprehensive radiological assessment, including plain radiographs and MRI, is sufficient to diagnose acquired osteoarticular amyloidosis (AOA) and avoid biopsy. The differential diagnosis includes multiple myeloma, metastatic disease, and Waldenström's macroglobulinemia, among others. It is challenging to differentiate between myeloma and AOA, as both present with similar subcortical radiolucent lesions. Metastatic lesions are often located in the spine or in the metaphyseal/diaphyseal regions of long bones. Brown tumors secondary to hyperparathyroidism may be difficult to distinguish from amyloid cysts in hemodialysis patients. Joint conditions such as rheumatoid arthritis, gout, xanthomatosis, and pigmented villonodular synovitis are also differential diagnoses to consider [2,3].

The management of osteoarticular amyloidosis is either preventive or palliative. The reduction in dialysis duration through kidney transplantation and advances in dialysis, including the use of high-permeability dialysis membranes since the 1990s, have allowed for better clearance of beta-2 microglobulin. More recently, the use of so-called ultrapure dialysis water has further improved the purification process. These measures have led to the near-elimination of dialysis-related arthropathy, though cystic lesions still persist.

Palliative measures include surgery, which involves curettage and bone grafting, though it carries a risk of infection and graft incorporation failure. Percutaneous cementoplasty is a less invasive alternative for preventing pathological fractures and has shown promising results [5].

Conclusion

In summary, acquired osteoarticular amyloidosis (AOA) is a complication that should not be overlooked in dialysis patients. It requires thorough clinical, radiological, and histological evaluation for an accurate diagnosis and appropriate management.

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