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A Case of Dialysis-related Amyloidosis of the Hip and Cervical Spine: Imaging Findings[1]

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Dialysis-related amyloidosis is a complication of long-term hemodialysis and it is characterized by the accumulation of β2-microglobulin in the osteoarticular structures. We describe here the imaging findings of a case of dialysis-related amyloidosis involving the hip and cervical spine in a 62-year-old woman who received long-term dialysis. We focus here on the CT and MR imaging findings of the cervical spine and we include a review of the relevant literatures.

Index words : Amyloidosis,
Spine, radiography
Spine, CT
Spine, MR
Dialysis

Dialysis-related amyloidosis is a complication of long-term hemodialysis and it is characterized by the accumulation of β2-microglobulin in the osteoarticular structures. In 1984, Kuntz et al. (1) first described destructive spondyloarthropathy in ten patients who received long-term hemodialysis. The lesions in that study were radiographically characterized by severe narrowing of the intervertebral disk and erosions and cysts of the adjacent vertebral plates without osteophytosis.

In the axial skeleton, a rapidly progressive destructive spondyloarthropathy may be difficult to distinguish from infectious spondylitis because both conditions have similar radiographic findings (2). Thus, needle biopsy is generally performed to confirm the diagnosis and exclude infectious spondylitis.

In this article, we describe the imaging findings of a case of dialysis-related amyloidosis involving the hip and cervical spine in one patient who received long-term dialysis. This case is presented to provide appropriate information about this disease and so help physicians minimize unnecessary biopsy procedures.

Case Report

A 62-year-old woman was suffering with chronic renal failure secondary to glomerulonephritis. She complained of hip and neck pain for 6 months after receiving dialysis for 12 years. There was no history of trauma. She was afebrile and displayed no clinical features of infection. Repeated measurements of the erythrocyte sedimentation rate (ESR) and C-reactive protein were within normal limits at the time of presentation.

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Repeated blood cultures were also negative.

Simple radiography of the hip demonstrated multiple cystic erosions in the femoral head and neck. There was significant loss of the disk space at C5 - C6, endplate irregularities and subchondral sclerosis without significant osteophytosis on the simple radiography of the cervical spine (Fig. 1). The CT examination of the cervical spine also demonstrated multiple erosions, irregularities and sclerosis of the vertebral endplates (Fig. 2). MRI of the hip showed multiple high-signal erosions in the femoral head and neck (Fig. 3A). The MRI spin-echo T1- and fat saturated T2-weighted images of the cervical spine showed multiple areas of abnormal low signal intensity involving the fifth and sixth vertebral bodies and the fifth disk, and the gadolinium-enhanced sagittal T1-weighted images showed mild enhancement of the adjacent marrow (Figs. 3B - 3D).

Total hip replacement was done and the specimen was sent for pathologic examination. Congo red staining of a surgically resected specimen demonstrated apple-green colored amyloid material that was observed under a polarized light microscopy (Fig. 4). The final diagnosis was made on the basis of the pathology reports.

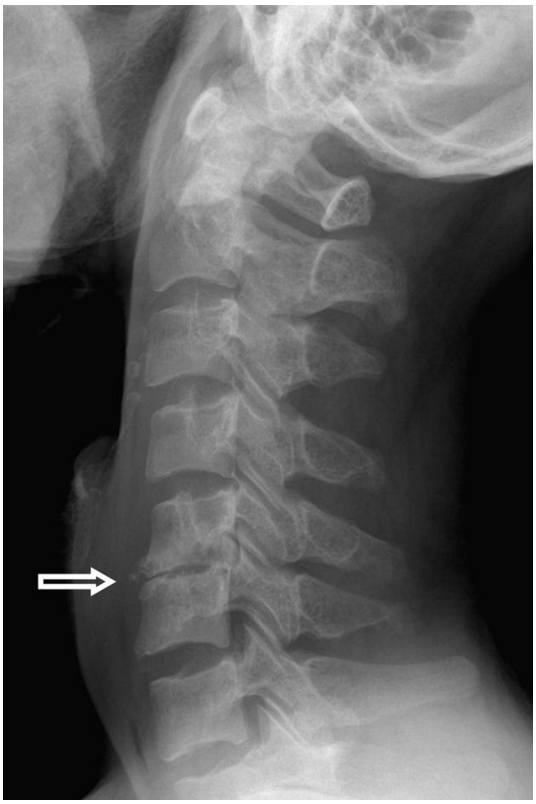


Fig. 1. A 62-year-old woman with hip and neck pain for 6 months.
Lateral radiograph of the cervical spine shows severe narrowing of the disk space at the C5-C6 level, endplate irregularities and subchondral sclerosis (arrow).

Discussion

Dialysis-related amyloidosis is a relatively uncommon complication resulting from long-term hemodialysis. The incidence of this disease increases with the number of years the patients has been on dialysis. In previous studies, the incidence was reported to be 80% to 100% for the patients who have been undergoing dialysis for over 20 years (3). In our case, the duration of dialysis was similar to that of the majority of other cases described in the literature. When this condition is observed in the spine, it affects the cervical area in most cases (approximately 70%) and less commonly in the lumbar (20%) and thoracic areas (10%) (2). Clinically, most patients present with myelopathy or radiculopathy, and the most serious complication that 's been reported was cord compression as a result of β2-microglobulin deposition in the spinal canal (4). In case of hip involvement, the patients showing large cystic erosions of the femoral neck had a high prevalence of pathologic fracture (5). Our patient was surgically treated because of the impending pathologic fracture of the right femoral neck that was secondary to the extensive cystic erosion.

The pathogenesis of spinal abnormalities is still poorly understood. However, mechanical factors seem to be important as the disease is most often detected in the highly mobile segments of the cervical and lumbar spines such as C5 - C7 and L3 - L5 (6). Lesion was de-

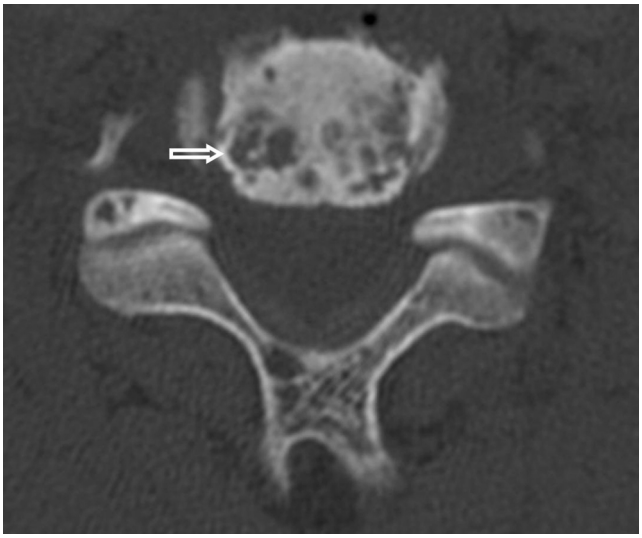


Fig. 2. The noncontrast, axial CT scan through the C5-6 level shows multiple erosions of the vertebral endplate (arrow).

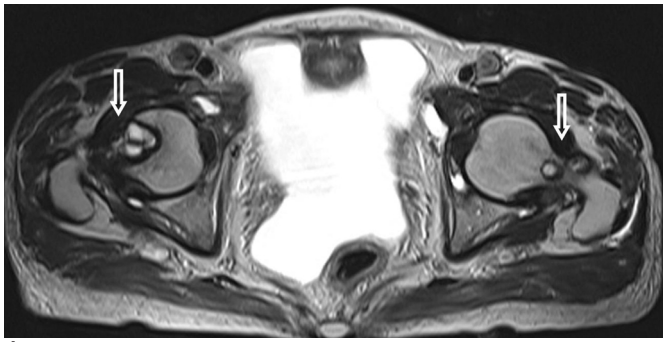
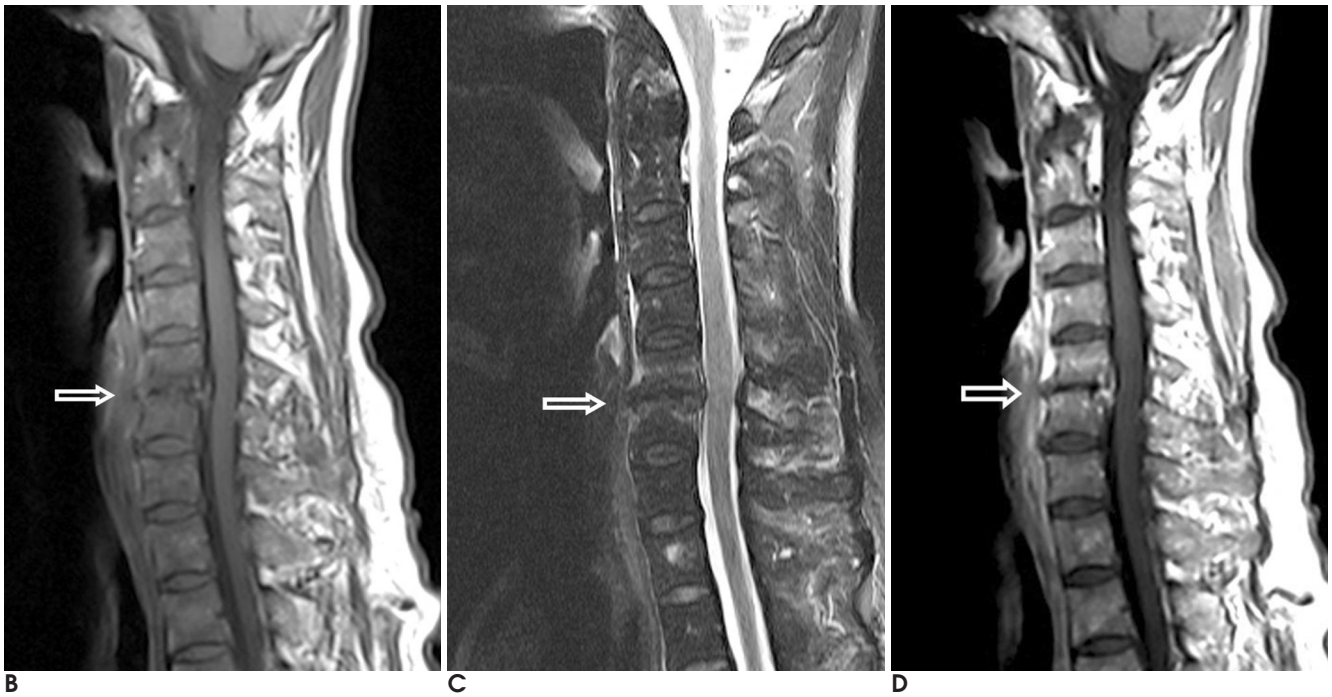


Fig. 3. A. The axial spin-echo T2-weighted (TR/TE, 3600/90) MR image of the hip shows multiple high-signal erosions in the femoral head and neck (arrows).
B, C. Sagittal spin-echo T1-weighted (TR/TE, 536/11) (**B**) and sagittal fat saturated spin-echo T2-weighted (3480/102) (**C**) MR images of the cervical spine show multiple areas of abnormal low signal intensity involving the fifth and sixth vertebral bodies and the intervening disk (arrow).
D. The corresponding gadolinium-enhanced sagittal spin-echo T1-weighted (536/11) MR image shows mild enhancement of the adjacent marrow (arrow).

A



B C D

tected at C5 - C6 in our patient. Ohashi et al (7) have reported the relationship between disk degeneration and deposition of β2-microglobulin, and they explained that β2-microglobulin has a strong affinity for collagen that has degenerated via mechanical stress.

Even though there are many radiographic and CT findings of dialysis-related spondyloarthropathy such as significant loss of disk space, endplate irregularities and subchondral sclerosis without significant osteophytosis, it's difficult to distinguish between dialysis-related spondyloarthropathy and infectious spondylitis (1, 2). However, dialysis-related spondyloarthropathy shows low signal intensity in the intervertebral disk and the adjacent vertebral endplates on both the T1- and T2-weighted spin echo MR images (8, 9). On the other hand, infectious spondylitis shows increased signal intensity on the T2-weighted spin echo images. The most important imaging feature that distinguishes dialysis-re-

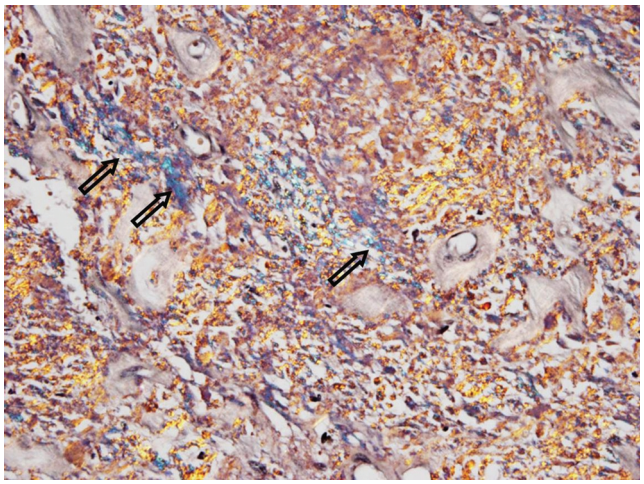


Fig. 4. Photomicrograph demonstrates deposition of apple-green colored amyloid material under polarized light microscopy (arrows) (Congo-red stain, ×400).

lated spondyloarthropathy from infectious spondylitis is the absence of a paraspinal mass despite of the rapidly progressive discovertebral destruction (8). Our patient hadn't any clinical or laboratory evidence of infection, and the MR imaging findings in the cervical spine were similar to those of the previously reported cases. Therefore, MRI allows the accurate diagnosis of dialysis-related amyloidosis and the exclusion of infectious spondylitis. In addition, gout and calcium pyrophosphate dihydrate (CPPD) crystal deposition disease may involve the intervertebral disk and adjacent endplates, and this disease produces changes that may simulate those of dialysis-related spondyloarthropathy (9). However, for our patient, the 12 years duration of dialysis, the laboratory data and the concomitant disease of the hip joint helped to exclude gout and CPPD. Thus, imaging findings combined with the patient's history and laboratory data are essential for the assessment of musculoskeletal involvement by the dialysis-related amyloidosis.

In the radiological literature, the MR imaging findings of dialysis-related amyloidosis involving the peripheral joints are well known (5, 10, 11). The radiographic findings of this entity may be difficult to distinguish from those of pigmented villonodular synovitis (PVNS) because of the similar radiographic findings on both the simple radiography and MRI. In most of cases, dialysis-related amyloidosis is a systemic, multiarticular process, but PVNS is a monoarticular process that has a predilection for the large joints of the lower extremity (5). The presence of low signal intensity in hemodialysis arthropathy on the T2-weighted spin echo images is caused by the hypocellular and fibrous nature of the amyloid-containing tissues (12). On the other hand, the low signal areas noted in PVNS on the T2-weighted spin echo images are caused by a paramagnetic susceptibility effect of hemosiderin. Thus, the gradient-echo sequences may be helpful because of the increased paramagnetic susceptibility effect produced by the hemosiderin relative to that produced by the hypocellular and fibrous nature of the amyloid-containing tissues (12). In our case, the 12 years duration of dialysis and the absence of hemosiderin helped to exclude PVNS.

We report here on the imaging findings of dialysis-related amyloidosis that involved the hip and cervical spine in a 62-year-old woman who had received long-term dialysis. In conclusion, knowledge for the imaging findings of this uncommon condition will provide the appropriate information to physicians to help prevent invasive diagnostic and treatment procedures.

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투석과 관련된 고관절과 경추의 아밀로이드증: 영상소견의 증례보고[1]

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투석과 관련된 아밀로이드증은 장기간 투석후에 발생하는 합병증으로 골관절 부위에 β2 - microglobulin 침착된다. 장기간 투석후 고관절과 경추를 침범한 투석과 관련된 아밀로이드증의 62세 환자에서 경추부위를 중심으로 영상소견을 문헌 고찰과 함께 보고한다.