Case Report

Amyloidomas on the bilateral buttocks associated with osteoarthropathy of the hips：a case report focusing on CT and MRI findings

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Abstract

A tumoral presentation of amyloidosis is uncommon in soft tissue. We here report a case of bulky amyloidomas on bilateral buttocks in a patient with osteoarthropathy of the hips. The 66-year-old patient's history included 26 years of hemodialysis along with osteoarthropathy of the hips and a pathological fracture of the femoral neck.

CT showed ill-defined bulky masses with a slightly higher density compared with that of muscles in the subcutaneous regions of the bilateral buttocks. Moderate contrast enhancement of the lesions was revealed. The masses contained diffuse tiny fat depositions and psammomatous calcifications, suggestive of amyloid deposition. On MRI, the masses demonstrated markedly hypointense signals on T2-weighted image, consistent with amyloidosis. Characteristic scattered spots of high intensity, suggesting fat or microcalcifications, were noted within the masses on T1-weighted image.

Mass-like synovial thickening was observed in the bilateral hip joints with discrete marginal erosions of the femoral heads. The CT and MRI features of the hips were compatible with typical amyloid osteoarthropathy.

（Key words：amyloidoma, β_2-microglobulin, buttocks, osteoarthropathy, CT, MRI）

Introduction

Joint and tenosynovial deposits of β_2-microglobulin（β_2-M）amyloid are often observed in patients undergoing long-term hemodialysis[1,2]. Amyloid deposition in soft tissues is rare, but a few cases of tumoral amyloidoma involving the buttocks have been reported[3-7]. We experienced a patient with extensive, bulky amyloidomas in the subcutaneous regions of the bilateral buttocks, associated with osteoarthropathy of the hip joints. Here, we describe the aforementioned case, focusing mainly on the CT and MRI manifestations.

Case Report

A 66-year-old man presented with masses in the bilateral gluteal region, which had gradually increased in size over several years. His medical history included end-stage chronic renal failure, which had been controlled with hemodialysis for 26 years, and osteoarthropathy of the hip joints. Four years before admission, he experienced a femoral neck fracture；since then, he was unable to walk unassisted and was confined to a bed or wheelchair all day. Physical examination revealed elastic, hard subcutaneous masses that were palpable beneath the gluteal skin. He did not complain of any pain or tenderness.

CT showed extensive ill-defined, bulky solid masses in the subcutaneous region of the bilateral buttocks（Fig.1）. The lesions demonstrated a slightly higher density compared with the muscles. In addition, abnormal subcutaneous soft-tissue density behind the coccyx was observed. On contrast-enhanced CT, the masses revealed moderate enhancement. Diffuse tiny fat deposition and psammomatous calcifications were noted within them. CT at the femoral head level showed mass-like synovial thickening in the bilateral hip joints, accompanied by marginal erosions and bone

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Received：27 April 2018, Accepted：31 August 2018

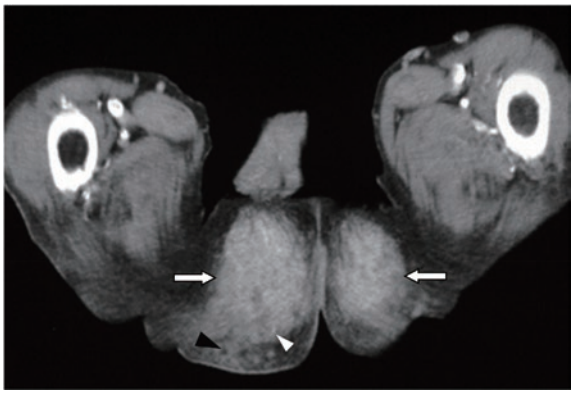

Figure 1 Contrast-enhanced CT at the level of the buttocks.
Ill-defined bulky solid masses were noted in subcutaneous regions of the bilateral buttocks (arrow). Theses lesions exhibited slightly higher density than the muscles and moderate contrast enhancement. Diffuse tiny fat depositions (black arrow head). and psammomatous calcifications (white arrow head) were observed within the masses. The muscles of both patients' thighs were atrophic.

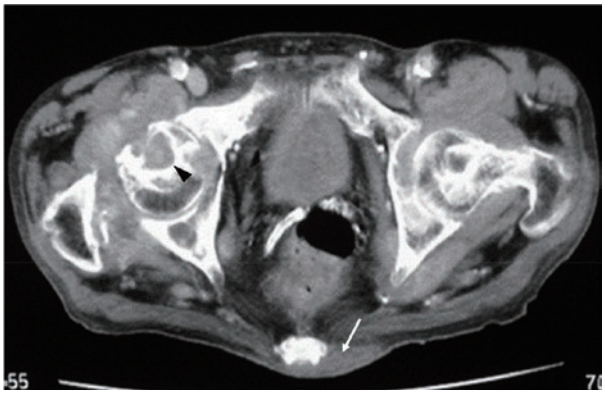

Figure 2 Contrast-enhanced CT at the level of the femoral heads
Lobulated soft-tissue swelling was depicted in and around the hip joints bilaterally, accompanied by marginal bone erosions and destruction in the right femoral head (arrow head). There was abnormal soft-tissue density in the subcutaneous region behind the coccyx (arrow).

destruction at the femoral heads (Fig.2). The gluteal soft-tissue masses were not continuous with the hip joint lesions.

On MRI, the masses on the buttocks demonstrated heterogeneous signal intensities ; these were markedly hypointense relative to muscles on T2-weighted image and isointense on T1-weighted image (Fig.3). Scattered psammomatous spotty high signals, suggestive of fat or calcifications, were also observed within the masses on T1-weighted image. The signal intensity of lobulated synovial lesions in the bilateral hip joints were identical to that of the gluteal masses (Fig.4).

Amyloid deposition was suggested on the basis of the image findings, but subcutaneous tumoral amyloidoma was considered to be rare. A percutaneous needle biopsy of the gluteal lesion was performed to exclude malignant tumor, and confirmed the diagnosis of β_2-M amyloid deposition (Fig.5).

Discussion

Dialysis-related amyloidosis is a disabling disease characterized by soft-tissue deposition of β_2-M amyloid in patients undergoing long-term hemodialysis [1, 2]. This disorder predominantly involves the musculoskeletal system and clinically manifests as conditions such as erosive and destructive osteoarthropathies, destructive spondyloarthropathy, or carpal tunnel syndrome. Skin involvement is not uncommon in dialysis-related amyloidosis ; however, massive subcutaneous deposits of β_2-M amyloid are rare [3-7]. Subcutaneous amyloidoma occurs in the buttocks most commonly, as follows in order of the lower leg, shoulder, popliteal fossa and inguinal region, in a

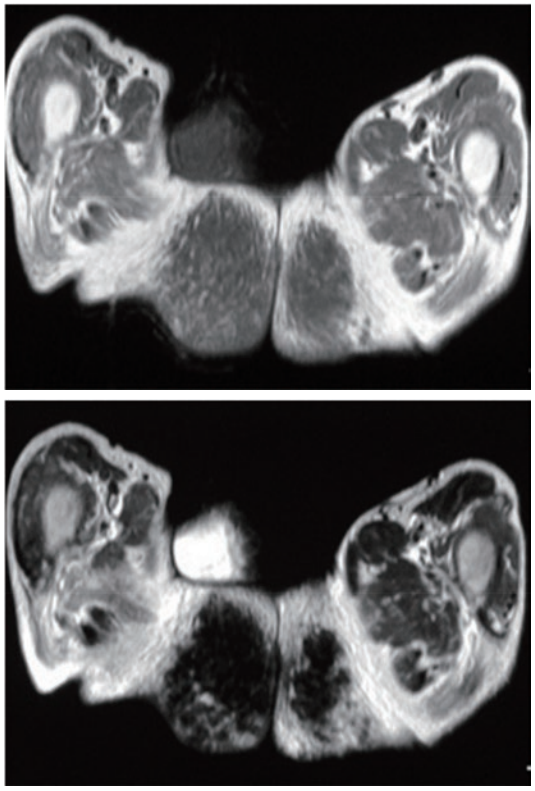

Figure 3 Axial MR images at the level of the buttocks.
A : T1-weighted image. Bulky masses were noted in the subcutaneous regions of the buttocks bilaterally. The signal intensities of the lesions were slightly lower than the muscles. Some spots of high signal intensity could be observed within the right-side mass.
B : T2-weighted image. The masses were markedly hypointense relative to the muscles with heterogeneous intensities.

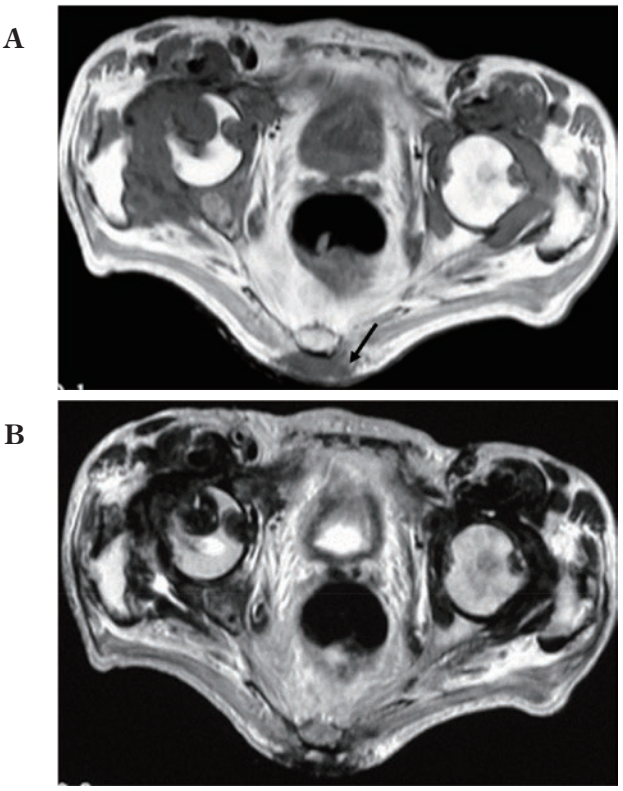

Figure 4 Axial MR images at the level of the femoral heads.

A : T1-weighted image. Well-demarcated lobulated synovial thickening, showing low signal intensity, was noted in the bilateral hip joints. Marginal bone erosions and destruction were depicted in the femoral heads. Abnormal soft-tissue signal was observed behind the coccyx (arrow).

B : T2-weighted image. The signal intensity of the lesions is markedly hypointense compared to the muscles, which was identical to that of the gluteal masses.

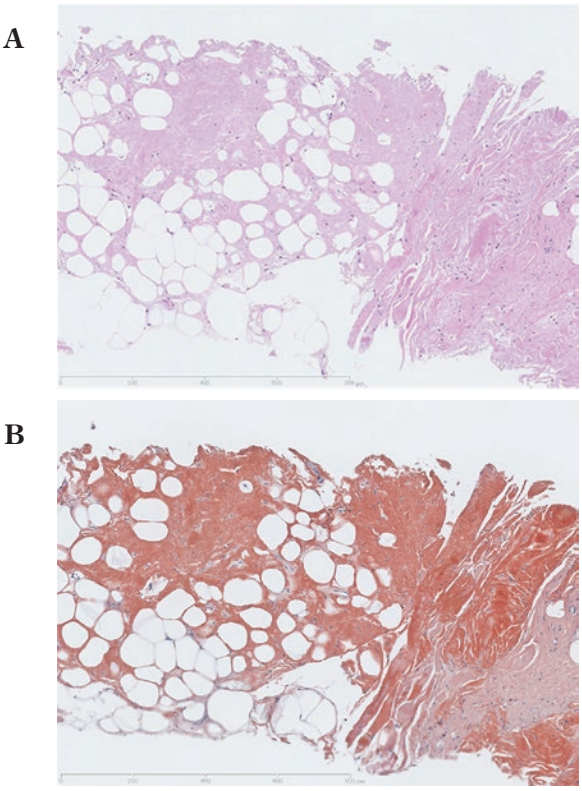

Figure 5 Pathological findings.

A : HE staining. B : DFS staining.

HE staining shows a light pink material suggesting amyloid deposits. DFS staining shows orange amyloid deposits.

small number of cases[4].

The pathogenesis of dialysis-related amyloidosis has been reported to be associated with the duration of renal failure, the patent's age, the duration of hemodialysis, and bioincompatibility of the dialysis membranes[2]. Typical clinical features include age greater than 40, years of undergoing more than 10 years of hemodialysis, suggesting that this unusual complication occurs late in the dialysis-related process[2]. The development of subcutaneous amyloid deposition in the bilateral buttocks and behind the coccyx suggests that chronic pressure injury experienced by a debilitated patient may have been a contributing factor of this condition.

There have been recent review articles discussing the imaging findings of amyloidosis [8-10], but these articles did not consider subcutaneous amyloidoma. If thorough clinical information is not available, the definitive diagnosis of amyloidoma may be difficult through image evaluation alone. The concurrence of synovial osteoarthropathy of the hips was decisive evidence for the diagnosis of β_2-M amyloidosis.

In our case, soft-tissue masses demonstrating a slightly higher density than muscles were considered to be distinctive CT findings. The symmetrical bilateral locations and the diffuse tiny fat deposition and psammomatous calcifications within the lesion suggested disorders related to the deposition of some sort of material. Although amyloid material generally does not show contrast enhancement, most reported cases of localized amyloidosis have been enhanced [11, 12]. The degree of the enhancement of amyloidoma is considered to vary according to the amount of inflammatory tissue around the amyloid material, which may have played a role in the enhancement.

On MRI, amyloid deposition shows a heterogeneously low signal on both T1- and T2-weighted images [13, 14]. Especially, markedly hypointense on T2-weighted image is a specific feature of amyloidoma and is useful in differentiating it from other pathological conditions. The short T2 relaxation

time is probably due to the hypocellular and fibrous nature of amyloid-containing tissues [13]. Scattered psammomatous spotty high signals within the masses, suggestive of fat or microcalcifications, may be distinctive findings on T1-weighted image.

The differential diagnosis includes fibromatosis and pathological conditions characterized by hemosiderin deposition as a result of the tendency of lesions to bleed. Fibromatosis can be distinguished by its common location and the lack of contrast enhancement. Amyloid deposition does not exhibit a paramagnetic effect on gradient-echo sequences, which can be helpful in the differential diagnosis of hemosiderin deposition [15].

Amyloidomas should be considered when diagnosing bilateral gluteal soft-tissue masses in patients undergoing long-term hemodialysis. Both CT and MRI are useful imaging modalities for evaluating the extent of amyloidoma and assessing tissues according to the characteristic manifestations of the disease.

Conclusion

This report presents a case of bilateral gluteal amyloidomas associated with osteoarthropathy of the hips, focusing on the CT and MRI manifestations. Diffuse tiny fat depositions and psammomatous calcifications suggested amyloid deposition on CT. On MRI, markedly hypointense lesions were noted on T2-weighted image and scattered spots of high signal intensity were characteristic findings on T1-weighted image.

Declaration of interest

Conflict of interest disclosure：None

Acknowledgements

The authors would like to thank Enago（www.enago.jp）for English language review.

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アミロイド骨関節症を伴った臀部アミロイドーマの１例：CT, MRI 所見を中心に

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要　　約

皮下の軟部組織に腫瘤を呈するアミロイドーシスは稀である。26年間の血液透析歴があり、股関節の骨関節症を伴う両側殿部のアミロイドーマを来した症例を経験したので、CTおよびMRI所見を中心に報告した。

CTでは両側殿部の皮下に境界不明瞭な軟部腫瘤を認め、筋肉よりもやや高い吸収値を示し、中等度の造影効果が見られた。腫瘤内部にびまん性の微細な脂肪及び砂粒状の石灰化が認められ、アミロイドの沈着が示唆された。MRIではT2強調像で著明な低信号を呈し、T1強調像では脂肪または微細石灰化を示唆するびまん性の点状の高信号が腫瘤内に認められた。CTおよびMRI所見はともにアミロイドーマに特徴的な画像所見と考えられた。

患者は両側股関節に骨関節症と大腿骨頚部に病的骨折の病歴を有していた。両側股関節には腫瘤様の滑膜肥厚と大腿骨頭部に境界明瞭な骨侵蝕像が認められ、アミロイド骨関節症を示唆する所見を呈していた。

（キーワード：アミロイドーマ、β_2-ミクログロブリン、殿部、骨関節症、CT、MRI）

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受付：2018年４月27日，受理：2018年８月31日