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CASE REPORT
Reconstructive

Surgical Treatment of Ischial Ulcers Associated with Deposition of β2-Microglobulin in Two Cases of Dialysis-related Amyloidosis

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Summary: The accumulation of β2-microglobulin due to long-term hemodialysis is known as dialysis-related amyloidosis, a rare phenomenon that manifests as a subcutaneous mass. Subcutaneous β2-microglobulin amyloidomas are predominantly located on the buttocks. Owing to the load-bearing properties of this location and proximity to the anus, amyloidomas on the buttocks may be prone to pressure ulcers and infection. This report presents two cases of long-term hemodialysis patients who required surgical treatment for infected ulcers caused by buttock amyloidomas. In the first case, treatment failed after the amyloidoma was excised and covered with a single-stage skin flap. In the second case, successful treatment was accomplished by reducing the volume of the amyloidoma, followed by a pause to allow for granulation growth and a two-stage skin graft. Amyloids of this nature are known to be cytotoxic; thus, a robust wound preparation technique should be used until the excision site is fully covered with granulation tissue before wound closure is initiated at the time of surgery. In addition, buttock amyloidomas often extend subcutaneously through the hip joint, and repeated infections may lead to more severe outcomes, such as hip joint infections. The number of dialysis-related amyloidosis patients has been increasing in recent years; thus, we report these case studies to improve patient outcomes in similar cases. *(Plast Reconstr Surg Glob Open 2023; 11:e5039; doi: 10.1097/GOX.0000000000005039; Published online 7 June 2023.)*

In patients with dialysis-related amyloidosis (DRA), β2-microglobulin (β2-M) deposition is common in regions of the osteoarticular system, such as the carpal tunnel and spine. In rare cases, deposits have been reported in the cardiovascular system, abdominal organs, and subcutaneous areas.[1,2]

Amyloidoma is a solitary mass of amyloid protein that can be found in different organ systems.[3] Subcutaneous β2-M amyloidomas are predominantly located on the buttocks.[2] Local mechanical stress may promote the deposition of β2-M amyloids.[4] Because the hip joints and pelvic region are loading zones in both sitting and supine positions, β2-M amyloid deposits in the hip joints may increase and extend into posterior subcutaneous regions.[5]

Treatment of DRA involves both surgical treatment, depending on the site of deposition, and medical treatment using dialysis techniques that decrease β2-M in the blood.[6] Asymptomatic subcutaneous amyloidoma of the buttocks is often observed but may require surgical treatment if pressure ulcers develop. We present two cases of pressure ulcers that developed over the subcutaneous amyloidomas of the buttocks that we surgically treated.

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CASE PRESENTATION

Case 1

A 73-year-old man, who had been undergoing hemodialysis for 37 years for chronic glomerulonephritis, presented to our hospital with symptoms of cervical

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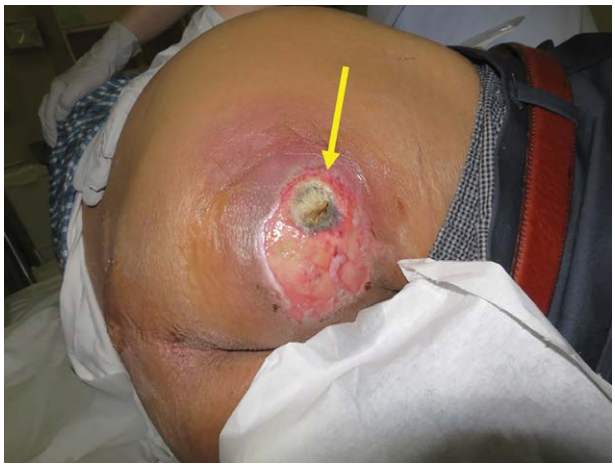

Fig. 1. Case 1: the right buttock at the initial examination. Skin erosion and exposed amyloidoma with white necrotic tissue are observed on the right ischial spine (yellow arrow).

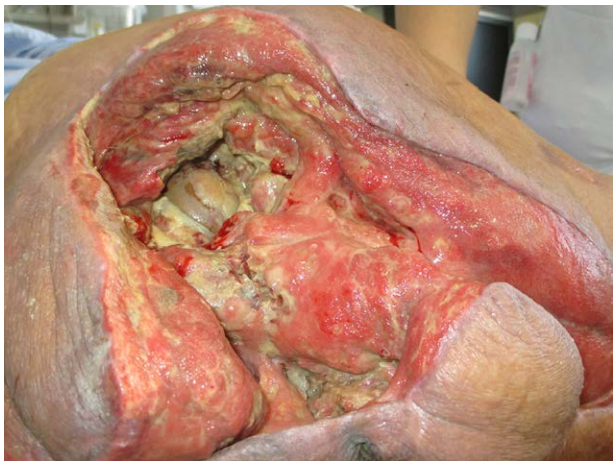

Fig. 2. Case 1: 3 months after the second surgical treatment. Dorsal view of the right buttock. The wound is opened along the skin incision of the two flaps. The hip capsule is open, exposing the femur. Amyloid deposits are observed around the hip joint.

myelopathy, polyarthralgia, amyloid arthritis of the left hip joint (which had previously been replaced), postoperative bilateral carpal tunnel syndrome, and amyloidomas in both hips. A clinical diagnosis of DRA was made.

He experienced decreased mobility for 3 months due to cervical myelopathy and developed an infected pressure ulcer with an exposed amyloidoma in the right buttock (Fig. 1). Magnetic resonance imaging showed a 63×100×66 mm subcutaneous mass around the right ischial spine directly below the pressure ulcer (see Supplemental Digital Content 1, arrows). Both T1- and T2-weighted images showed lower signals than did those in nearby muscle regions, characteristic of amyloid deposition.[5] (**See figure, Supplemental Digital Content 1,** which shows a 63×100×66 mm subcutaneous mass around the right ischial spine directly below the pressure ulcer. **http://links.lww.com/PRSGO/C595**.)

The patient was treated conservatively with antibiotics and topical ointment for 2 months. After the infection subsided, debridement and wound closure were performed. Necrotic tissue was completely removed, but the amyloidoma was attached to the hip joint and could not be completely removed. The defect was reconstructed with a posterior thigh fasciocutaneous flap. Histologically, the specimen showed eosinophilic deposits that were positive for Congo red staining, indicating amyloid deposition.

In the postoperative period, the wound gradually opened from the tip of the flap and exposed the amyloidoma, which was infected. The patient was treated with antibiotics, debridement of the infected tissue, and wound management for 3 months, including 1 month of negative pressure wound therapy. Infection was controlled, but adequate granulation on the amyloidoma was not achieved.

The wound was closed again with a one-time debridement and reconstruction. The amyloidoma adherent to the hip, including the noninfected portion, was resected as much as possible, and a rotational flap procedure with a gluteus maximus flap was performed. The flap again got detached, and it was found that the infection had spread to the hip joint (Fig. 2). Three months after the last surgery, the patient died of multi-organ failure due to sepsis.

Case 2

A 66-year-old woman, who had been undergoing hemodialysis for 38 years for chronic glomerulonephritis and had previously undergone left femoral head replacement for amyloid arthritis, presented to our hospital with symptoms typical of DRA (fractures of the cervical and lumbar spine and carpal tunnel syndrome).

She had been experiencing pain in her buttock for 1 month and skin ulcerations on her right buttock for 3 weeks before the visit. She presented with erythema, swelling, and erosion of the right buttock (Fig. 3). Computed tomography revealed a 100×95×75 mm subcutaneous mass in the right ischial spine (see Supplemental Digital Content 2, yellow arrow). Deposits around the femur were

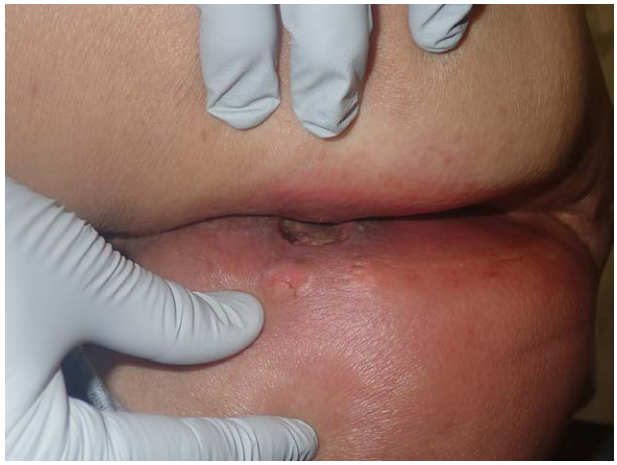

Fig. 3. Case 2: a photograph of the wound at the initial examination. The right hip is positioned at the bottom of the photograph. Manual attempts to open the gluteal cleft are limited. A skin ulcer is observed on the right buttock. The entire right buttock is erythematous and swollen.

present, with slightly higher absorption than that of those around the surrounding muscle tissue, characteristic of β2-M deposits. Elevated periadventitial fatty tissue and subcutaneous gas were also observed (see Supplemental Digital Content 2, white arrow), suggesting the presence of infection. (**See figure, Supplemental Digital Content 2,** which shows a 100×95×75 mm subcutaneous mass in the right ischial spine. **http://links.lww.com/PRSGO/C596**.)

The patient was admitted to the hospital with a diagnosis of necrotizing soft tissue infection and underwent debridement. The wound containing the exposed amyloidoma was opened, and the patient was treated with antibiotics and topical ointment. Two weeks later, a colostomy was performed to prevent fecal contamination in conjunction with a second debridement. Biopsies were taken and stained with Congo red, confirming the deposition of eosinophilic unstructured material in the region. The immunostaining results were positive for β2-M. The patient was treated with gradual daily detachment of necrotic tissue and the amyloidoma, the volume of the amyloidoma gradually decreased, resulting in the extension of granulation tissue over the amyloidoma from the surrounding normal tissue. Two months after the initial visit, the amyloidoma was completely covered with good granulation tissue, and we performed split-thickness skin grafting (Fig. 4). There was no ulcer recurrence over the next 2 years, after which the patient died of unrelated causes.

DISCUSSION

Although several reports on subcutaneous amyloidomas of the buttocks exist, few have focused on surgical treatment practices.[2,3,5,7]

In the first case, the amyloidoma was treated as a typical autologous oligemic tissue of the bone, joint, or tendon. One-stage reconstruction was performed in conjunction with debridement without waiting for granulation on the amyloidoma. However, the flap did not grow over the amyloidoma and was repeatedly infected. Amyloidomas in the buttocks may be formed by subcutaneous extensions of amyloid deposits in the hip joints.[5] Deep debridement and repeated infections may result in infections of the hip joints, which can have severe effects on patient outcomes.

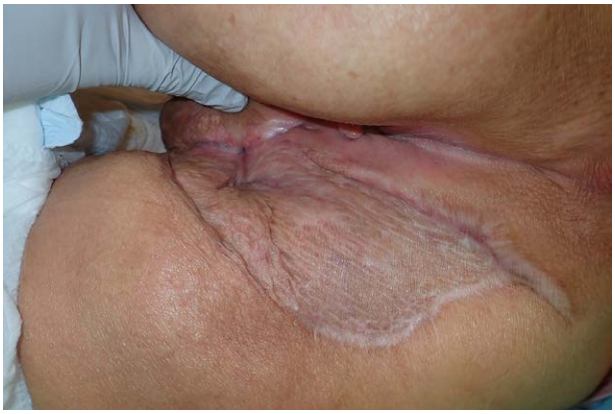

Fig. 4. Case 2: 6 months after surgical treatment by a split-thickness skin graft. No ulcer recurrence is visible.

β2-M amyloids have been shown to induce cell death in rabbit cells.[8] Unlike other autologous oligemic tissues, amyloidomas may create a hostile environment for flap growth.

In the second case, after maximum excision of the amyloidoma, adequate granulation was achieved, and the skin graft was successful. This case demonstrated that growing granulation tissue on an amyloidoma from the surrounding normal tissue is possible by surgical reduction of its volume. Although complete removal of all pathologic tissues is usually a prerequisite for surgical closure, such a palliative treatment may be an option.

The prevalence of DRA in maintenance dialysis patients is approximately 20%.[9] Epidemiological trends show that the percentage of dialysis patients experiencing DRA is decreasing owing to improvements in dialysis; however, the total number of dialysis patients continues to increase, resulting in the number of DRA patients more than tripling between 1998 and 2010.[10] As the number of DRA patients with amyloidomas requiring surgical treatment is expected to increase, and we are unaware of similar case studies, we have discussed these two cases.

CONCLUSION

If an ulcer develops on a gluteal amyloidoma, we recommend that the volume of the amyloidoma be surgically reduced and the amyloidoma be covered with granulation tissue.

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DISCLOSURE

The authors have no financial interests to declare in relation to the content of this article.

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We followed the Declaration of Helsinki and our institutional regulations.

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