Solitary Amyloidoma Related to THA

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Abstract

Full article available online at Healio.com/Orthopedics. Search: 20130624-31

Amyloidoma of soft tissues is rare, and no previously published reports describe it in relation to hip prostheses. This article presents the case of a 78-year-old woman with a medical history of myelodysplasia and mild renal failure who underwent a right-sided metal-on-polyethylene total hip arthroplasty in 2003. She presented to the outpatient clinic with a 1-year history of right anterior thigh pain. On examination, a large proximal anteromedial lump was palpable and was nontender, nonpulsatile, and firm. She had a positive Trendelenburg test but good hip range of motion. Plain radiographs showed signs of osteolysis around the hip prosthesis. Serology showed mildly raised liver function and renal function but a normal erythrocyte sedimentation rate and white blood cell count.

Magnetic resonance imaging revealed a large mixed-signal soft tissue mass directly opposing the anterior cortex of the right femur, related to the hip prosthesis, measuring 16×10×7 cm. Associated destruction of the underlying cortex existed. Histological staining of a biopsy of the mass confirmed that the mass contained AL-type amyloid seen in primary amyloidosis.

Mean patient age at diagnosis for amyloid tumors of soft tissues is 66 years. By definition, they start as solitary lesions. The types are important: AA type is related to infection and AL type is a primary process. Patients with AL amyloidomas have a poorer prognosis because they have a higher chance of malignancies. Early diagnosis can prevent long-term serious consequences of this condition.
Amyloid is an amorphous, eosinophilic material that, when stained with Congo red, exhibits a characteristic green birefringence under polarized light. Its deposition is largely systemic, but occasionally it is limited to a single organ.

Amyloid deposits can be further categorized according to the peptide AA or AL, from which it is formed. The AL type is variable segment immunoglobulin light chains produced by the monoclonal proliferation of plasma cells or B-lymphocytes. This is usually primary and systemic but can be related to conditions including lymphoma and leukemia. The AA type is acute phase protein produced in excess to chronic inflammation, including rheumatoid arthritis, inflammatory bowel disease, or chronic infections such as tuberculosis and osteomyelitis.

Primary solitary amyloidosis or amyloidoma is characterized by localized deposition of amyloidoma in which no plasma cell dyscrasia or abnormal serum proteins are detectable. It is the least common presentation and can be presented in multiple body sites, usually in axial body including the respiratory tract, urinary tract, lymph nodes, breast, gastrointestinal tract, brain, and bone. The current authors have found no previously published reports of a soft tissue amyloidoma related to total hip arthroplasty (THA).

This article presents the case of a 78-year-old patient with an amyloidoma involving her THA.

**Case Report**

A 78-year-old woman with a medical history of myelodysplasia and mild renal failure underwent a right-sided metal-on-polyethylene THA in 2003. She presented in 2011 to the authors’ outpatient clinic with a 1-year history of right anterior thigh pain. On examination, a large proximal anteromedial lump was palpable and was nontender, nonpulsatile, and firm. She had a positive Trendelenburg test but good hip range of motion. Plain radiographs showed signs of marked proximal bone loss adjacent to the hip prosthesis. Serology showed mildly raised liver function and renal function but a normal erythrocyte sedimentation rate and white blood cell count.

Magnetic resonance imaging revealed a large mixed-signal soft tissue mass directly opposing the anterior cortex of the right femur, related to the hip prosthesis, measuring 16×10×7 cm (Figure 1). Associated destruction of the underlying cortex existed.

An ultrasound biopsy showed a solid fibril hemorrhagic lesion with peripheral calcification, suggesting that this lesion could be a polyethylene granuloma or a soft tissue tumor. The patient was referred to the regional orthopedic oncology unit, which suggested that the lesion was unlikely to be malignant, although a specific diagnosis was not given, and surgical excision or debulking of the mass was recommended.

The authors performed surgical debulking of the mass, which intraoperatively appeared solid and had no gross appearance of infection. The proximal femur was excised and reconstructed with a massive proximal femoral endoprosthesis (Figure 3).

Histological examination of the tissue showed fibrocollagenous tissue, numerous polymorphs, and fibro fatty tissue consistent with acute inflammation (Figure 3). No evidence existed of particle wear from the prosthesis. Congo red staining of the
mass was positive and was apple green bi-refringent under polarized light, confirming that the mass contained amyloid.

Potassium permanganate staining confirmed that the amyloid was not the AA-type amyloid usually associated with infection, but the AL-type amyloid seen in primary amyloidosis. Serum electrophoresis for systemic amyloid was negative, although nonspecific background staining was high, which is consistent with renal impairment. Other tests were negative for systemic amyloidosis.

DISCUSSION

Solitary amyloidoma related to a THA has not been previously reported. Mean patient age at diagnosis for amyloid tumors of soft tissues is 66 years. By definition, they start as solitary lesions. Cases of amyloidoma have been related to bone, but these have largely been in the axial skeleton.

A painless, solid soft tissue mass adjacent to a hip prosthesis is more suggestive of fat necrosis, soft tissue tumor such as sarcoma, or reaction to wear particles from the prosthesis. Magnetic resonance imaging is helpful for characterizing the lesion, with areas of low signal on T2-weighted images, but tissue biopsy is necessary to achieve a diagnosis. The diagnosis can be challenging, with initial biopsy results nonspecific and consistent with inflammatory tissue. In the current case, specific histology staining was necessary to distinguish the amyloid from neoplastic plasma cells.

The cause of a solitary amyloidoma in the limbs is unknown. Tan et al reported that in a series of 12 cases of amyloidoma in the limbs, 8 patients had comorbidities, including breast and lymphoma cancer, and chronic conditions including diabetes, renal failure, and peripheral vascular disease. Five of these were AA-type amyloidoma, and only 1 had AL-type amyloidoma. The patient with AL amyloidoma had a history of trauma to the site and had multiple diseases, including hypercholesterolemia, diverticulosis, and hypothyroidism. The current patient had none of these characteristics and had no history of trauma.

Differentiating between AA- and AL-type amyloids is important because patients with AL-type amyloid have a poorer prognosis. These patients are at higher risk of developing malignancies. Krishnan et al reported that of 10 patients with AL-type amyloid, 8 developed lymphoplasmacellular malignant neoplasms. The AL-type amyloid can also progress into systemic disease. The AA-type diseases are generally cured by excision and do not reoccur.

With an aging patient population and an increasing number of patients undergoing THA, an increased variety of complications are likely in the future. It is important for clinicians to consider amyloid as a differential diagnosis in cases similar to the current case because a late diagnosis can lead to devastating results. Although amyloid is low on the list of differential diagnoses, it should be considered when other pathologies have been excluded.

REFERENCES