CASE REPORT

Presentation of *calcinosis cutis universalis* in mixed connective tissue disorder: an encounter during hip arthroplasty

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SUMMARY

A woman aged 23 years with a diagnosis of mixed connective tissue disorder presented with left groin pain extending over 6 months. Workup revealed avascular necrosis of the femoral head (Grade 3) secondary to systemic lupus erythematosus and chronic steroid intake. An uncemented total hip arthroplasty was considered as the patient was only in the third decade of life. During the preop workup, careful clinical assessment had revealed multiple subcutaneous nodules affecting the extensor musculature limited to the gluteal region, anterior and posterior aspects of the thigh. The diagnosis of *calcinosis cutis universalis* was made after a CT revealed calcified nodules in the subcutaneous, subfascial and muscular planes. A total hip arthroplasty using the posterior approach was performed with minimal trauma to the calcified nodules and thereby preventing a source of persistent drainage and reducing morbidity due to infection.

BACKGROUND

Patients with mixed connective disorders who are on long-term steroids are usually immunocompromised and are at increased risk of infection after a total hip arthroplasty. *Calcinosis cutis universalis* can interfere during a posterior approach to the hip as these nodules are found exclusively in the extensor aspect of lower limbs and in deeper planes. Any

damage to these calcified nodules can lead to persistent drainage from the wound site and a possible source of infection leading to reoperation. Accordingly, careful dissection during hip arthroplasty is vital in surgical cases.

CASE PRESENTATION

A woman aged 23 years presented with left groin pain and reduced ambulation for 6 months. The pain was moderate to severe, insidious onset, non-radiating, sharp pain, associated with night pain and while at rest. Her activities of daily were restricted due to the reduced ambulatory status. No treatment had been formally initiated. There was no history of trauma, no history of weight loss. She was diagnosed with systemic lupus erythematosus (SLE) at the age of 13 and was on a treatment regimen that included steroids and mycophenolate mofetil. A noted flare up of SLE symptoms resulted after recent childbirth, ~2 years ago.

No family history of SLE or mixed connective tissue disorder. The patient was not a smoker or an alcoholic. Physical examination revealed cushingoid facies, obesity, striae over the trunk and extremities,

Figure 1 Multiple, small, nodular swelling over the gluteal region as well as the posterior aspect of the thigh.

Figure 2 A CT of the pelvis and hips showing extensive calcifications nearly obscuring the hip joint on an anteroposterior view.

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Figure 3 An axial CT image showing the involvement of the subfascial and submuscular planes around the hip joint, also note the destroyed femoral head.

Figure 4 A reformatted 3D image showing the preferential involvement of the extensor compartment. This is a characteristic feature of *calcinosis cutis universalis*.

multiple nodules over the gluteal region (figure 1) and pedal oedema. The hip was in a flexed and abducted position, with an apparent shortening of 2 cm affecting the left lower limb. Movements around the hip were restricted due to pain. Neuro and vascular exams were normal and intact. A diagnosis of mixed connective tissue disorder was formulated after serological tests reported a positive U1RNP by line immune assay, positive antinuclear antibody and homogenous pattern on

immunofluorescopy. CT (figures 2–4) reveals a Grade 3 avascular necrosis of the femoral head and also calcinosis cutis in various planes, hence a diagnosis of *calcinosis cutis universalis* was made.

OUTCOME AND FOLLOW-UP

An uncemented total hip arthroplasty was performed and on follow-up after 2 months, the patient was able to walk and move around comfortably on a weight bearing as tolerated basis.

DISCUSSION

Calcinosis cutis is most commonly associated with dermatomyositis and is of four types: (1) dystrophic, (2) metastatic, (3) iatrogenic and (4) idiopathic as described by Virchow. The aetiology is not known. Dystrophic calcinosis is the most common mode of presentation. There are limited options in the treatment of this condition. Medical management of calcinosis cutis with calcium channel blockers and colchicine has been tried but not proven to be successful.¹ Surgical excision of the nodule is an option, but it is associated with the risk of recurrence.² Since, this is a rare condition, significant data regarding the natural history, course and management options do not exist. The association of mixed connective tissue disorder and *calcinosis cutis universalis* is sparsely reported in the literature. There has been no evidence in the available literature that highlights the importance of detecting calcinosis in such patients during a hip arthroplasty and thereby improving surgical outcome.

Learning points

- *Calcinosis cutis universalis*, though a rare entity, can occur in patients with mixed connective tissue disorder.
- Preop planning is necessary to avoid damage to these nodules and thereby preventing a source of infection.
- Clinical suspicion and observation is imperative to diagnose such cases.

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