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Case report

MRI features of pyoderma gangrenosum in a diabetic patient with ulcerative colitis: A case report and review of the literature ☆,☆☆

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ABSTRACT

Pyoderma gangrenosum (PG) is a rare noninfectious skin condition which clinical picture can overlap with that of the diabetic foot. Meticulous physical examination along with biopsy and magnetic resonance imaging (MRI) can make the distinction easier, saving the patients from undergoing a debilitating intervention.

We report a case of pathologically proven PG in the right ankle region of a 55-year old male with known uncontrolled diabetes mellitus and inflammatory bowel disease.

Radiographs revealed increased soft tissue density overlying the lateral melleolus of the right ankle. MRI showed a well-defined soft tissue mass with heterogeneously intrinsically high signal intensity on T1- and on T2-weighted images, and heterogeneous peripheral enhancement on fat-suppressed, contrast-enhanced T1-weighted images. Histologically, diffuse neutrophilic infiltrate throughout the dermis was present without micro-organisms. This is the first report of MRI findings of PG in the ankle. We also summarize the findings of previously reported cases of PG.

Introduction

PG first described in 1908, is a rare, noninfectious skin condition with an incidence of 3-10 cases per million people per year [1,2]. The name is a misnomer as it was initially mistakenly thought to be an infectious process caused by purulent streptococcal infection (hence the term pyoderma) that resulted in necrosis (hence the term gangrenosum) [3].

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Although it can affect both sex and any age group, it is more common among the young and middle-aged population (30-60 years) with a predilection for females [4].

While this disorder is characterized by heavy neutrophilic infiltration, the cause remains unknown. Initially, this condition was thought to be caused by a bacterial infection where the resultant antibodies incited an autoimmune reaction. Nevertheless, this theory has been disproved by subsequent studies in the literature suggesting that the altered neutrophilic function along with immune system and genetic tendency (genetic mutation on chromosome 15q) are the main contributors to this condition [2,5].

Patients with pyoderma gangrenosum (PG) commonly present in association with an inflammatory bowel disease (IBD, particularly UC), constituting the classical type [5–7].

We present a case of a 55-year-old male, who initially presented with symptoms of IBD and was diagnosed with UC after undergoing a colonoscopy and eventually biopsy. Upon the multiple UC flare up episodes the man kept developing bilateral ankle ulcers that were eventually proven to be PG by magnetic resonance imaging (MRI) and pathology. We describe the image characteristics of our case of PG and reviewed the MRI features of the few available related case reports in the literature.

Case report

A 55-year-old male, with known osteoarthritis, uncontrolled diabetes mellitus and polysubstance abuse presented to the emergency with the chief complain of bloody diarrhea and bilateral buttocks, legs and ankles skin lesions. A colonoscopy followed by biopsy was performed which demonstrated squamocolumnar junction mucosa showing marked acute and chronic inflammation with cryptitis and crypt abscess, and ulceration with fibrinopurulent exudates within the region of the sigmoid and rectum. The diagnosis of UC was made. MR enterography was performed and confirmed the diagnosis and extent of the disease (Images included at the end of the article).

The buttocks and legs showed multiple skin lesions, with similar appearance in the form of round ulcers with clean edges and overlying eschar, surrounded by edema. No discharge was found (Fig. 1). The patient was started on steroids and was discharged.

Patient was not compliant with steroids and during the subsequent years, the patient presented with multiple flare up episodes of UC. On the most recent visit, the patient developed erythema and tenderness in both ankles, over the region of the medial and lateral malleolus. The largest lesion was seen overlying the lateral malleoulus of the right ankle measuring approximately 5 × 6 cm (Fig. 2). A punch out biopsy of the right lateral ankle skin lesion was performed demonstrating diffuse neutrophilic infiltrate throughout the dermis (Fig. 3). Both, tissue culture and histopathological stains for micro-organisms, failed to show any infection. He was placed on infliximab which was later changed to mesalamine, as he developed antibody formation.

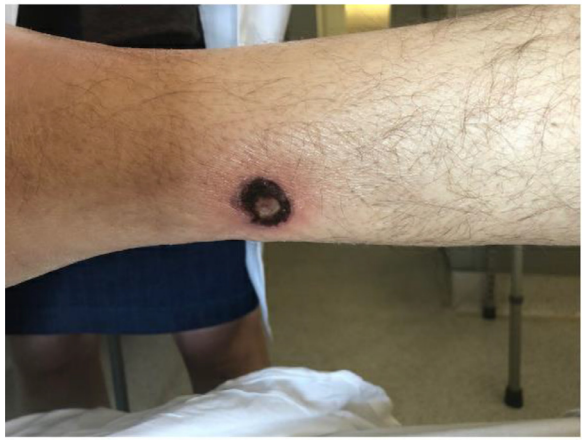

Fig. 1 – A demonstration of one of the ulcers seen within the leg.

MRI of the right ankle with contrast was performed and demonstrated a 4.3 × 2.2 × 5.6 cm (AP x TRV x CC) complex appearing, peripherally enhancing fluid collection within the subcutaneous tissues overlying the lateral aspect of the distal fibular metadiaphysis. The lesion demonstrated intrinsic T1 hyperintense signal with some dependent, layering debris within this collection. Additionally, there was a subdermal portion of this collection which demonstrated a dependent fluid-fluid level. The walls of this collection were in contact with the underlying distal fibula which demonstrated normal signal with no evidence of osteomyelitis. There was extensive subcutaneous edema along the lateral aspect of the distal leg and ankle, extending into the dorsum of the midfoot and beyond (Fig. 4). A radiograph of the right ankle was performed and showed soft tissue swelling overlying the lateral malleolus corresponding to a complex fluid collection (Fig. 5).

The diagnosis of PG was made based on the aforementioned clinical, pathologic and MRI findings. The right ankle was debrided (Fig. 6) and the patient was started on steroids and proper wound care was conducted, with the wound showing good signs of healing in subsequent encounters.

Discussion

PG is a diagnosis of exclusion. Although the clinical picture significantly overlaps with the diabetes related complications such as infection and venous ulcers, rendering it a medical dilemma, certain diagnostic elements aid in the differentiation [8].

Multiple types of PG have been described in the literature, the most common is the classical ulcerative type, as patients present with an autoimmune condition, such as IBD (majority UC), inflammatory arthritis, connective tissue disease, hepatitis C and hematological disorders (notably myeloproliferative, myelodysplastic and lymphoproliferative) [5–7,9–11].

Other forms include the bullous type, which is usually associated with hematologic disease and presenting in the arms

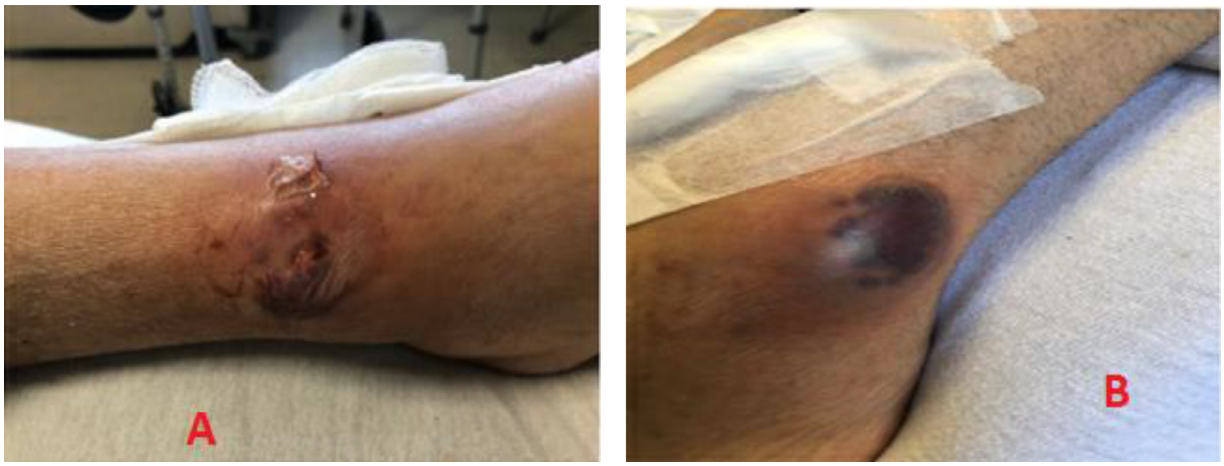

Fig. 2 – Demonstration of the multiple bilateral skin lesions (A) the right ankle lesion appears as a flucuant blister with purple discoloration., while the left medial ankle lesion (B) appears as an unfroofed blister, tender to palpation. (Color version is available online.)

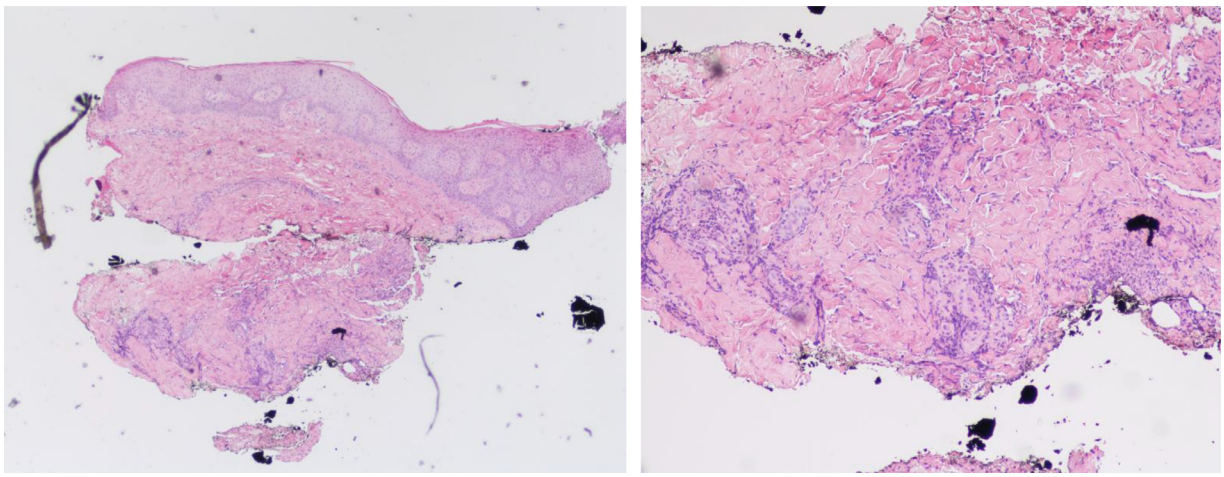

Fig. 3 – Diffuse neutrophilic infiltrates throughout the dermis. No organisms detected.

and the face, the pustular and vegetative types which mostly occur in the head and neck regions [2,5].

The reported case presented in the form of the classical type where the lesions typically start as a tender pustule in the lower extremity, preferentially within the pretibial region, with no history of prior trauma. These lesions tend to rapidly progress into a necrotic ulcer [2,5,7].

A detailed clinical history, meticulous physical examination, and skin biopsy are of a great benefit. However, thus far there are no validated, established diagnostic clinical or pathological criteria to diagnose PG. Maverakis et al have recently proposed new criteria, Delphi criteria, based on a consensus of international experts, requiring 1 major and 4 minor criteria [1].

The diagnostic criteria include 1 major and 8 minor criteria. The major criterion states that the biopsy of the ulcer should show evidence of neutrophilic infiltrate. The 8 minor criteria includes (1) exclusion of infection as a cause, (2) history of pathergy, (3) history of IBD or inflammatory arthritis, (4) history of a papule, vesicle or pustule ulcerating within 4 days if appearing, (5) peripheral erythema, undermining border and tenderness in the ulcers, (6) multiple ulcerations, at least one in the anterior lower legs, (7) cribriform scars, and (8) decreased ulcer size within 1 month of treatment with immunosuppressive medication [1,5]. This is yet to be widely adopted, but no longer renders PG as a diagnosis of exclusion which may thus provide an improved diagnostic tool.

Our case met the described Delphi criteria. The biopsy results of the ulcer in the right ankle demonstrated evidence of neutrophilic infiltrates. Infection was excluded, and history of autoimmune disease was present in the form of UC. The lesion started as a blister before breaking down to form an ulcer and finally the presence of peripheral erythema and tenderness in the ulcer.

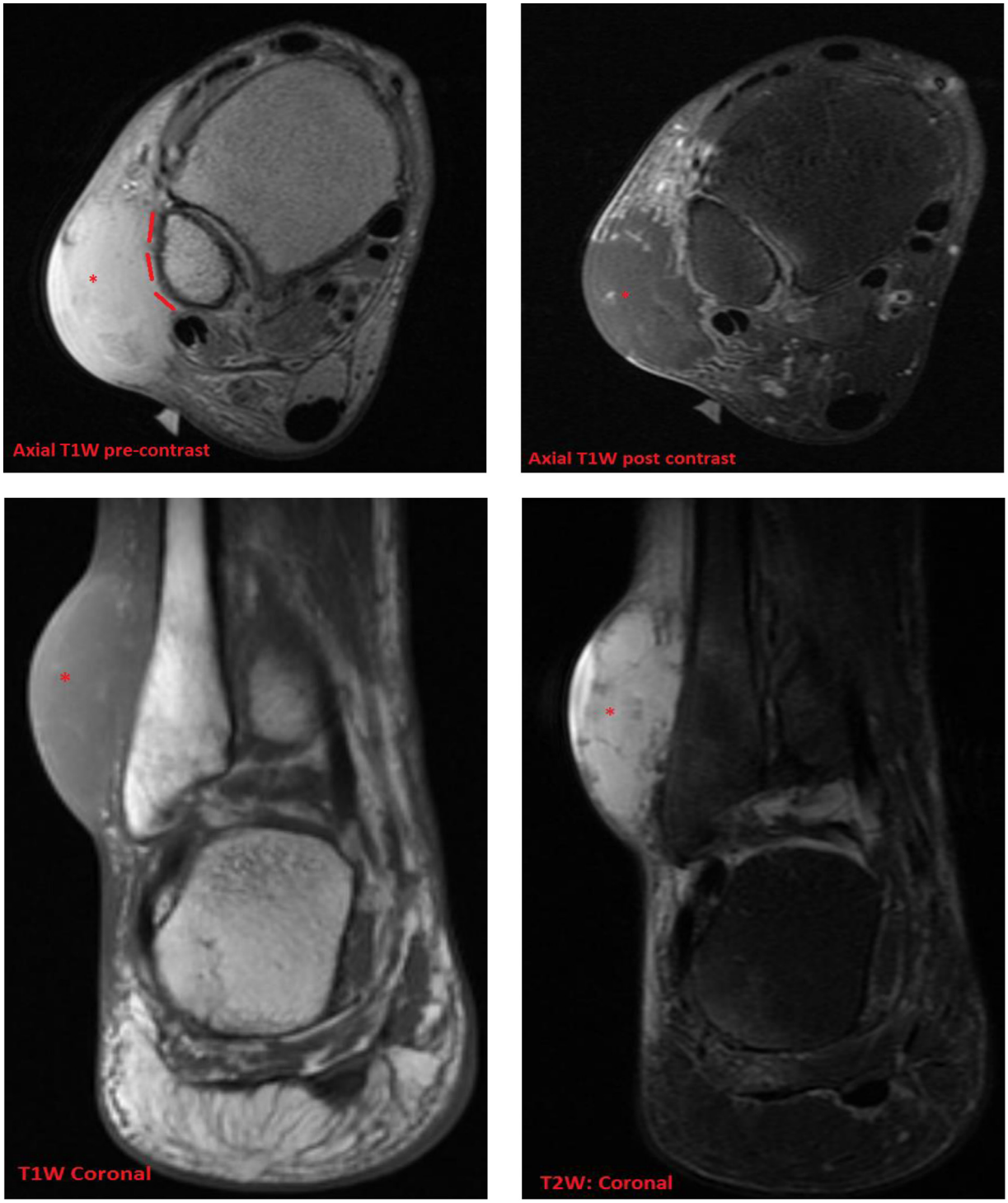

Fig 4 – A complex appearing, peripherally enhancing fluid collection (red asterisk) within the subcutaneous tissues overlying the lateral aspect of the distal fibular metadiaphysis, associated with extensive subcutaneous edema extending along the lateral part of the distal leg and ankle in conjunction with mild cortical thickening and/or chronic periostitis of the distal tibia and fibula (dashed line). (Color version is available online.)

Role of imaging

Although imaging has not been included as a criterion in the Delphi consensus for the diagnosis on PG, we believe that familiarity and awareness with this form of presentation is a must on the radiology practice, as the image findings of PG can easily be mistaken for abscess, cellulitis or even a tumor. Therefore, the acknowledgement of such an entity will help avoid erroneous diagnosis leading the patients to superfluous

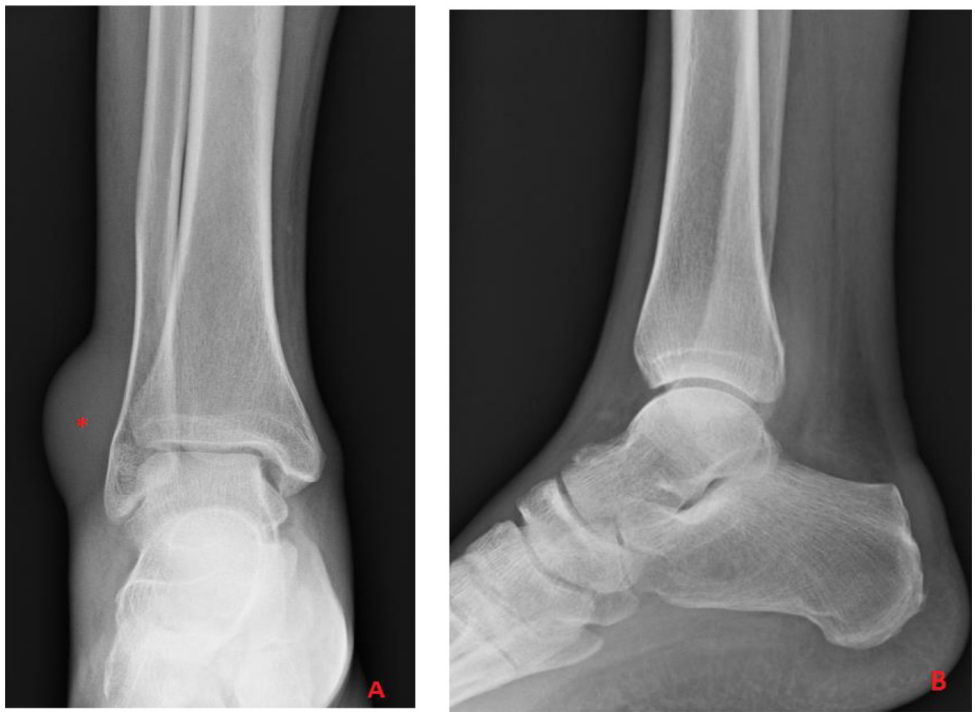

Fig 5 – Frontal (A) and lateral (B) radiographs of the right ankle showing soft tissue swelling (red asterisk) lateral to the lateral malleolus corresponding to a complex fluid collection. (Color version is available online.)

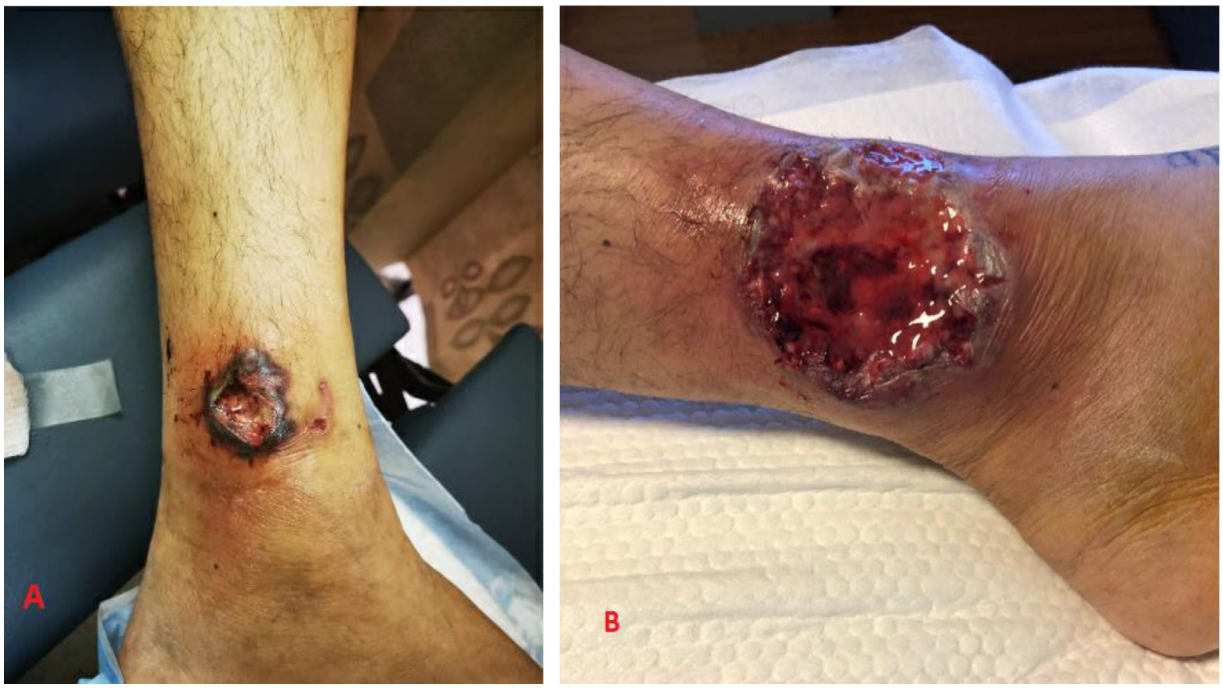

Fig 6 – Right ankle. Pre (A) and post (B) debribement.

treatments that could potentially impact patients' morbidity or even mortality.

We reviewed and summarized the few available case reports in the literature regarding the radiological findings related to this condition.

The presence of an ulcer raises concern for subjacent abscess formation or adjacent osteomyelitis (as in our case) [8]. Few case reports available in the literature demonstrated the association of osteomyelitis related to PG, which can be infectious vs sterile [9–11]. The distinction between them is

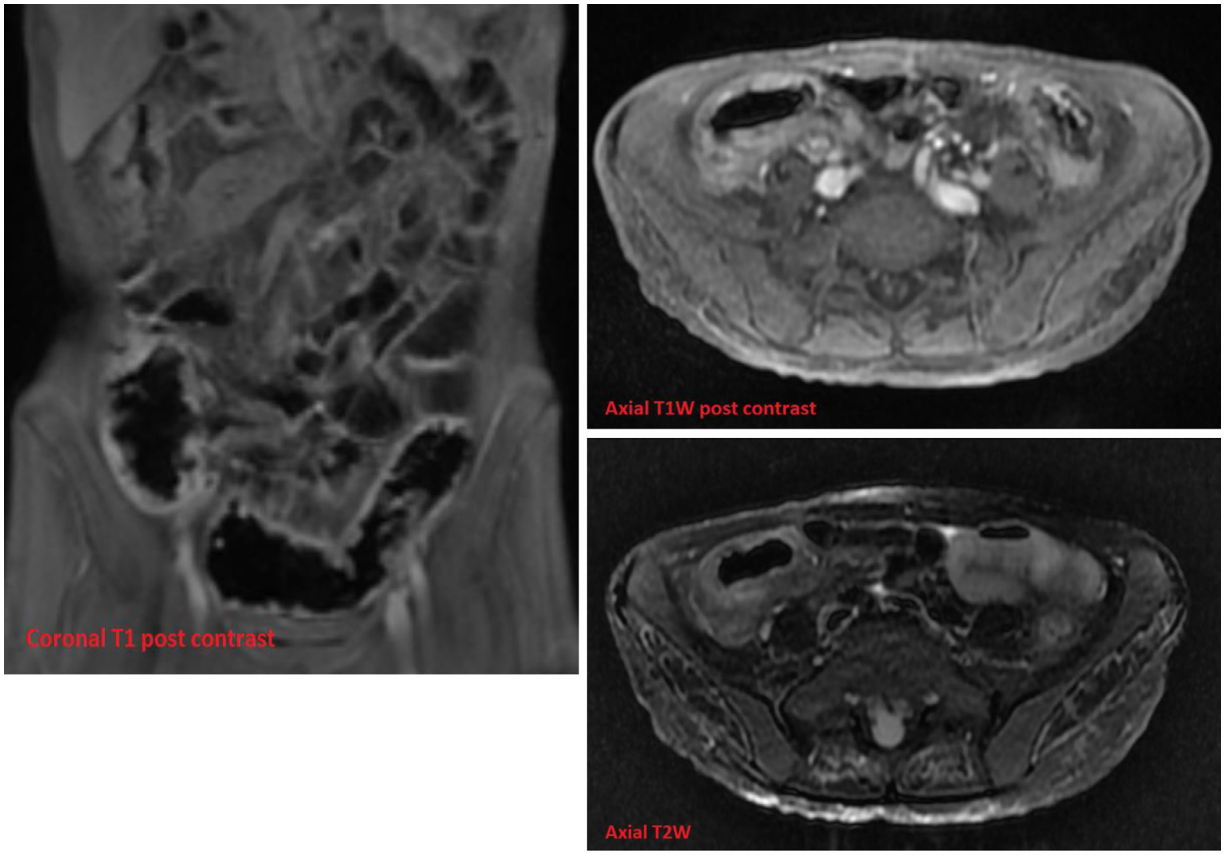

Fig. 7 – MRE showing active inflammation involving the entire colon, consistent with pancolitis and patient's known history of ulcerative colitis. Thickening and enhancement of approximately 20 cm length of jejunum, for which infectious or inflammatory enteritis is considered.

made based on the clinical and histological findings. MRI can provide value information in evaluating the presence of osteomyelitis and determining the extension of the bone marrow abnormality. The main MR sequence needed to assess osteomyelitis is T1 weighted image. Osteomyelitis will present as a confluent region of low signal intensity on T1 weighted images within the bone marrow. Fluid sensitive sequences will demonstrate high signal intensity within the affected bone marrow and administration of contrast will show associated enhancement and will help in determining the presence of an associated soft tissue abscess [14]. High signal intensity on fluid sensitive sequences in the absence of confluent low signal on T1 weighted image suggests reactive hyperemia. No confluent loss of expected marrow signal to suggest underlying osteomyelitis in our case.

In the setting of a known ulcer and soft tissue swelling, both clinical presentations of PG, MRI can also be useful to assess the presence of cellulitis, fluid collections or nodular lesions. MRI can determine the degree and extension of cellulitis which will present as skin thickening and increased signal intensity within the subcutaneous soft tissue on fluid sensitive sequences [8], assess the presence, morphology, size and location of a fluid collection and determine the presence of nodular subcutaneous lesions [3–15].

Additionally, PG can involve joints; portraying subchondral bones changes [15]. Furthermore, the changes can extend into muscles and fascial planes, ranging from myositis to myonecrosis [12]. A published case in the literature described associated muscle necrosis that appeared as areas of enhancement within the anterior tibilias muscle on Post T1 sequences [12]. Nevertheless, in the presence of subcutaneous gas PG could be misdiagnosed as necrotizing fasciitis due to the necrotic tissue [16]. The findings related to these entities can be assessed by MRI.

Conclusion

Although PG is a diagnosis of exclusion, certain diagnostic criteria have been made to reach the diagnosis, which take into consideration the clinical picture and the biopsy findings. Despite the fact that this is a clinical diagnosis, it is of utmost importance for the radiologists to be aware of this entity, and be able to assess potential associations such as osteomyelitis and differentiate from other entities such as abscesses especially in the setting of a diabetic foot, which can render the diagnosis difficult; as a way to prevent the patients from

undergoing unnecessary debilitating treatments, such as antibiotics or even amputations (Fig. 7).

Patient Consent Statement

Dear Editor, no consent was obtained nor required for the writing of this manuscript, as it is waived by our institution's IRB policy.

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