CASE REPORT

Limited systemic sclerosis initially presenting with mesenteric panniculitis

Mariangeli Arroyo-Ávila, Luis M Vilá

Division of Rheumatology,
Department of Medicine,
University of Puerto Rico
Medical Sciences Campus,
San Juan, Puerto Rico

Correspondence to

Dr Luis M Vilá,
luis.vila2@upr.edu

Accepted 1 October 2014

SUMMARY

Mesenteric panniculitis pertains to a group of uncommon disorders named sclerosing mesenteritis that present with different levels of inflammation and fibrosis of the small bowel mesentery. It is associated with abdominal surgeries, trauma, malignancies, infections and connective tissue diseases. To the best of our knowledge, no cases of sclerosing mesenteritis have been reported in patients with systemic sclerosis. We present a case of a 61-year-old woman who had incidental CT findings of mesenteric panniculitis. Diagnosis was confirmed by biopsy that showed fat necrosis. On further review she had a 1-year history of Raynaud's phenomenon. Physical examination showed sclerodactyly. She had elevated antinuclear antibodies and skin biopsy was consistent with scleroderma. She was diagnosed with limited systemic sclerosis and was treated with D-penicillamine. After 6 years of follow-up, the mesenteric panniculitis and systemic sclerosis both remained stable. This case highlights the importance of considering rheumatic diseases in the differential diagnosis of sclerosing mesenteritis.

BACKGROUND

Mesenteric panniculitis is part of a spectrum of rare disorders that present with varying degrees of fibrosis, inflammation and necrosis of the small bowel mesentery. These disorders are grouped together into the term sclerosing mesenteritis (SM).^{1,2} SM has been found in 0.6% of abdominal CT scans performed in patients with different abdominal conditions.³ It is associated with abdominal aortic aneurysm, abdominal surgeries, trauma, malignancies, infections and, less frequently, with autoimmune diseases.^{2,4} Rheumatoid arthritis (RA), Sjögren's syndrome, systemic lupus erythematosus and ankylosing spondylitis have been described in association with SM.¹⁻⁶ To the best of our knowledge, there are no reports in the literature of SM occurring in patients with systemic sclerosis (SSc). We report a case of a woman with mesenteric panniculitis as the initial presentation of limited SSc.

CASE PRESENTATION

A 61-year-old Hispanic woman underwent an evaluation for worsening symptoms of gastro-oesophageal reflux associated with severe chest pain in 2008. A chest CT scan revealed no pulmonary pathology but extensive mesenteric fat stranding consistent with mesenteric panniculitis. An abdominal CT scan confirmed these findings (figure 1). She had chronic arterial hypertension and gastro-

Figure 1 Initial abdominopelvic CT scan showed extensive mesenteric fat stranding with multiple nodes, consistent with mesenteric panniculitis.

oesophageal reflux disease treated with irbesartan, hydrochlorothiazide and esomeprazole. She had no history of tobacco or alcohol use. The patient's surgical history was significant for total abdominal hysterectomy and bilateral salpingo-oophorectomy 7 years prior to initial presentation. Her family history was remarkable for head and neck cancer in her maternal grandfather. Mammograms, pap smears and colonoscopy within the last year were normal. Whole body F-18 fluoro-deoxy-glucose positron emission tomography CT scan did not show any significant metabolic activity and hence no findings suggestive of the presence of lymphoma. A CT-guided aspiration and biopsy of the mesenteric mass showed findings consistent with fat necrosis, without evidence of malignant cells. Acid-fast bacilli and fungal organisms were not observed on special stains.

Extensive laboratory workup revealed positive antinuclear antibodies (ANA) for which the patient was referred for rheumatologic evaluation. On further review, she had a 1-year history of arthralgias, myalgias and Raynaud's phenomenon. She had no fever, chills, weight loss, tender or swollen joints, skin rashes, oral ulcers, abdominal pain, nausea, vomiting or shortness of breath. On physical examination she had a body mass index of 28.2 kg/m². She was alert and oriented and in no distress. She was noted to have sclerodactyly but no skin lesions or digital ulcers. The remainder of her physical examination was unremarkable.

INVESTIGATIONS

Complete blood count, serum electrolytes, renal and liver function tests, coagulation profile tests and erythrocyte sedimentation rate were normal.

CrossMark

To cite: Arroyo-Ávila M, Vilá LM. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-206961

She had positive ANA at 1:640 with centromere pattern. Anticentromere antibodies were markedly elevated (titre >1:2560). Antiribonucleoprotein antibodies and rheumatoid factor were also detected. Anti-dsDNA, anti-Smith, anti-Ro, anti-La, anti-Scl-70, anti-Jo-1, anti-proteinase-3 and antimyeloperoxidase antibodies were negative. C3 and C4 serum complements were normal. A skin biopsy of her left arm revealed a thickened dermis with thick collagen bundles and sparse perivascular lymphocytic infiltrate, consistent with scleroderma.

OUTCOME AND FOLLOW-UP

The patient was diagnosed with limited SSc and was started on low-dose D-penicillamine (250 mg daily). She had no adverse events to this medication. After 6 years of follow-up she had no progression of her disease. Apart from one episode of digital ischaemia, which resolved with clopidogrel, she has had no complications related to SSc. She did not develop pulmonary, cardiac or renal involvement. Furthermore, her mesenteric panniculitis remained asymptomatic and without radiographic changes, as demonstrated on follow-up abdominal CT scans (figure 2).

DISCUSSION

We describe a patient who on CT scan evaluation for chest pain was incidentally found with SM. Further evaluation disclosed clinical features of limited SSc. Even though the most common symptoms associated with SM are abdominal pain, bloating, distention and diarrhoea, patients may present asymptotically, as in our case.² In fact, a study evaluating the prevalence of SM in patients undergoing abdominal CT scans for various clinical indications found that only 8% of patients with CT findings suggestive of SM had symptoms attributed to the entity itself and, thus, the majority of the scans were performed for unrelated conditions.³ In our case, the patient's symptoms of severe reflux were likely a manifestation of SSc rather than SM itself.

Although no cases of SM have been reported in patients with SSc, this entity has been described in patients with other autoimmune connective tissue diseases.¹⁻⁶ Akram *et al*² published a review of 92 cases of SM seen at Mayo Clinic and found that 4 patients (4.3%) had associated rheumatological conditions, including 2 with Sjögren's syndrome, 1 with RA and 1 with sarcoidosis. However, details were not provided regarding the clinical manifestations of these patients nor regarding how the diagnoses were performed. Two reports described SM in two patients with a history of RA who presented with abdominal pain.^{4 6} Both patients had resolution of symptoms after the treatment with increased doses of prednisone and continuation

of their methotrexate therapy. However, SM findings on follow-up abdominal CT scans persisted. In both patients SM occurred during the course of RA. We found no reports of SM occurring before or concomitantly with a connective tissue disease.

Other forms of panniculitides, or adipose tissue inflammation, have been described in patients with SSc. Most lesions present with indurated, subcutaneous lesions in the lower extremities.⁷ Panniculitides are less commonly seen in the arms, thighs, calves and gluteus.^{8 9} One case report described a woman presenting with panniculitis of the arm who was subsequently diagnosed with diffuse SSc.⁹ In fact, the association between panniculitis and SSc is not surprising as they share similar pathophysiological mechanisms. Although the exact mechanism of adipose tissue inflammation is unknown, it is thought to occur from injury to the vascular endothelium, activation of fibroblasts and, hence, imbalance between production and destruction of collagen.⁷ Some of these processes are also seen in SSc.

There is no consensus on the most effective therapy for SM, but several medications have been reported to be successful such as prednisone, colchicine, azathioprine, methotrexate, thalidomide and tamoxifen.^{2 4-6} Similarly, treatment for panniculitides associated with SSc has not been studied, but therapy is usually targeted to the systemic illness itself.⁷ D-penicillamine has been used for many years to treat SSc and other connective tissue diseases because it prevents the cross-linking of collagen fibres and has immunosuppressive effects.¹⁰ Nevertheless, data on its therapeutic effectiveness has been conflicting. However, in our patient, low-dose D-penicillamine treatment resulted in clinical stability of the SSc as well as SM.

In summary, our patient presented SM as an initial manifestation of limited SSc. Although the exact aetiology of SM and, hence, mesenteric panniculitis, remains unknown it seems to share pathophysiological mechanisms with SSc. This report together with other cases highlights the importance of considering autoimmune rheumatic diseases in the differential diagnosis of a patient presenting with SM.

Learning points

- Sclerosis mesenteritis is a rare condition that present with different levels of inflammation and fibrosis of the small bowel mesentery.
- It is associated with abdominal aortic aneurysm, abdominal surgeries, trauma, malignancies, infections and autoimmune diseases.
- We present a patient with limited systemic sclerosis who initially presented with mesenteric panniculitis.

Figure 2 Abdominopelvic CT scan 6 years after diagnosis of sclerosing mesenteritis showed stable appearance of mesenteric fat stranding and mesenteric nodes.

Funding This work was in part supported by an unrestricted educational grant from AbbVie.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Emory TS, Monihan JM, Carr NJ, *et al*. Sclerosing mesenteritis, mesenteric panniculitis and mesenteric lipodystrophy: a single entity? *Am J Surg Pathol* 1997;21:392-8.
- 2 Akram S, Pardi DS, Schaffner JA, *et al*. Sclerosing mesenteritis: clinical features, treatment, and outcome in ninety-two patients. *Clin Gastroenterol Hepatol* 2007;5:589-96.

- 3 Daskalogiannaki M, Voloudaki A, Prassopoulos P, *et al.* CT evaluation of mesenteric panniculitis: prevalence and associated diseases. *AJR Am J Roentgenol* 2000;174:427–31.
- 4 Makdsi F, Brit M. A case of sclerosing mesenteritis with rheumatoid arthritis. *South Med J* 2010;103:96–7.
- 5 Sugihara T, Koike R, Nosaka Y, *et al.* Case of subcutaneous and mesenteric acute panniculitis with Sjögren's syndrome. *Nihon Rinsho Meneki Gakkai Kaishi* 2002;25:277–84.
- 6 Vlachos K, Archontovasilis F, Falidas E, *et al.* Sclerosing Mesenteritis: diverse clinical presentations and dissimilar treatment options. A case series and review of the literature. *Int Arch Med* 2011;4:17.
- 7 Braunstein I, Werth VP. Update on management of connective tissue panniculitides. *Dermatol Ther* 2012;25:173–82.
- 8 Almeida MS, Lima SC, Carvalho LL, *et al.* Panniculitis-an unusual cutaneous manifestation of systemic sclerosis. *J Cutan Pathol* 2010;37:1170–3.
- 9 Binesh VG, Krishnan J, Asokan N, *et al.* Systemic sclerosis presenting as lobular panniculitis. *Indian Dermatol Online J* 2012;3:222–4.
- 10 Clements PJ, Furst DE, Wong WK, *et al.* High-dose versus low-dose D-penicillamine in early diffuse systemic sclerosis: analysis of a two-year, double-blind, randomized, controlled clinical trial. *Arthritis Rheum* 1999;42:1194–203.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow