

Systemic multifocal fibrosclerosis

S S Johal, S Manjunath, C Allen, D B Trash

Summary

We describe a case of hydronephrosis as a result of retroperitoneal fibrosis in a patient who had previous sclerosing lobulitis of the breast. To the best of our knowledge this is the first reported association between these two conditions in the english literature. We presume these conditions are linked and unify them under the general heading of systemic multifocal fibrosclerosis.

Keywords: sclerosing lobulitis; retroperitoneal fibrosis; systemic multifocal fibrosclerosis

Systemic multifocal fibrosclerosis (SMF) is a condition of unknown aetiology characterised by fibrous lesions occurring at various sites. Clinical variants include retroperitoneal fibrosis, Riedel's thyroiditis, sclerosing cholangitis, mediastinal fibrosis, orbital pseudotumours,¹ insulin-dependent diabetes mellitus (IDDM), Sjogren's syndrome² and maxillary sinus disease.³ Sclerosing lobulitis has already been linked with some of the above conditions.² Due to the similarities between this condition and retroperitoneal fibrosis, we consider sclerosing lobulitis also to be a variant of SMF.

Case report

A 61-year-old woman was admitted with an episode of chest pain reminiscent of her angina which lasted 30 minutes. Subsequent serial electrocardiograms and cardiac enzymes were normal. She also described a constant dull ache in the right loin for the past two months. Examination of the abdomen revealed an enlarged right kidney, the bladder was not palpable. Rectal and vaginal examination were normal. Significant laboratory tests on admission revealed normochromic normocytic anaemia with haemoglobin 8.4 g/dl, erythrocyte sedimentation rate (ESR) 83 mm/h, C-reactive protein (CRP) 74 mg/l, with a normal white cell and platelet count. Blood creatinine and urea were normal. Urine analysis showed no blood, protein or malignant cells on cytological analysis and was sterile on culture. Ultrasound of the abdomen revealed hydronephrosis of the right kidney and a fibroid uterus but the ureter could not be visualised. Computed tomography (CT) identified a dilated right ureter with obstruction at its distal end as a result of a pelvic mass, the left side being relatively normal. A CT-guided biopsy of the pelvic mass was inadequate for histological analysis. At laparotomy the lower end of the right ureter was found to



Figure 1 Histology of pelvic mass showing fibrosis extending into the fat and around nerve fibres. There are both cellular areas with fibroblastic proliferation and keloid-like glassy collagen fibres. There is a chronic inflammatory cell infiltrate with no evidence of malignancy (H&E, original magnification $\times 25$)



Figure 2 Section of breast tissue showing lobular atrophy and sclerosis with interlobular and perilobular lymphocytic infiltrate. There are areas of both fibroblastic proliferation and hypocellular areas of keloid-like glassy collagen fibres (H&E, original magnification $\times 10$)

be involved in fibrosis and adhesions but no specific mass was identified. The ureter was dissected out and stented. Subsequent histology confirmed the presence of retroperitoneal fibrosis (figure 1).

Two years prior to these symptoms she had undergone a lumpectomy from her right breast. The histology of the resected specimen was reported as sclerosing lobulitis of the breast (figure 2).

She was commenced on 40 mg per day of prednisolone which was tapered off over 2 months. Subsequent investigations revealed a normal blood count, ESR, CRP, resolving hydronephrosis and no recurrence of the pelvic mass.

**Walsall Manor
Hospital, Walsall, West
Midlands, WS2 9PS,
UK**
**Department of
Medicine**
S S Johal
S Manjunath
D B Trash
**Department of
Histopathology**
C Allen

Correspondence to
SS Johal, 69 Liskeard Road,
Park Hall, Walsall, West
Midlands WS5 3ES, UK

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Discussion

Sclerosing lobulitis was first described by Soler and Khardori in 1984 in a group of patients with Hashimoto's thyroiditis, IDDM and cheiroarthropathy with histological similarity between breast tissue and thyroiditis.⁴ Fibrous mastopathy, diabetic mastopathy, and sclerosing mastitis are terms synonymous with sclerosing lobulitis. It is an uncommon condition associated with a raised ESR, CRP and normochromic normocytic anaemia. The histology is very similar to that of retroperitoneal fibrosis. Both conditions have a chronic inflammatory cell infiltrate with predominance of B lymphocytes and expression of HLA-DR antigen. Fibroblastic proliferation leads to hypocellular areas of keloid-like glassy collagen fibres. Autoimmune pathogenesis has been suggested.²⁻⁵ To the best of our knowledge, this is the first reported association between retroperitoneal fibrosis and sclerosing lobulitis of the breast. We feel all these conditions are clinical variants of SMF.

This case also illustrates how retroperitoneal fibrosis can present as an isolated pelvic mass and the requirement often of laparotomy to establish the diagnosis, relieve any obstructive nephropathy, and exclude malignancy such as lymphomas. Management remains controversial. Corticosteroids with surgical

SMF: reported associations

- mediastinal fibrosis
- maxillary sinus disease
- sclerosing cholangitis
- Riedel's thyroiditis
- retroperitoneal fibrosis
- Sjogren's syndrome
- sclerosing lobulitis
- pseudotumours of the orbit
- Hashimoto's thyroiditis
- IDDM

intervention in the form of ureterolysis and/or ureteric stenting is the most common approach. Corticosteroid therapy alone may relieve obstruction, but is by no means always effective. Uterolysis alone is associated with recurrence and the development of obstruction in the previously unaffected kidney.⁶ Immunosuppressants such as azathioprine and penicillamine have been used with variable success rates.⁷ The relapse rate after cessation of steroids has been studied by Baker *et al* in a series of 60 patients in which 10 patients relapsed after 5 years and a further five as late as 10 years, illustrating the need for long term follow-up.⁸

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Gastric rupture secondary to successful Heimlich manoeuvre

Arun Majumdar, P C Sedman

Summary

A fatal case of gastric rupture following the Heimlich manoeuvre is reported. This life-threatening complication has only been reported previously in seven patients with a high mortality rate. All patients should be assessed immediately following this manoeuvre for any potentially life-threatening complications.

Keywords: gastric rupture; Heimlich manoeuvre

The Heimlich manoeuvre is an essential first-aid resuscitative measure to relieve acute upper airway obstruction. Henry Heimlich first described this life-saving procedure in June 1974, after successfully carrying out experimental studies on animals. Within the first nine months, 162 lives were reported saved. Heimlich only encountered fractured ribs as a complication when the manoeuvre was improperly performed. Since then, however, a number of potentially fatal complications have been reported. We

Department of
Surgery, Hull Royal
Infirmary, Anlaby
Road, Hull HU3 2JZ,
UK
A Majumdar
P C Sedman

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