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Retroperitoneal Fibrosis Presenting with Recurrent Episodes of Abdominal Pain and Post-prandial Vomiting

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Abstract

Presentation

A 74 years-old lady presented with a one-week history of abdominal pain, post-prandial vomiting and pyrexia.

Diagnosis

Computed tomography (CT) of the abdomen and pelvis demonstrated symmetrical proliferation of retroperitoneal fat resulting in anteromedial displacement of both kidneys and the mesenteric vessels.

Treatment

The patient has treated conservatively with analgesia and nasogastric tube insertion. She was referred to a specialist centre for further management of her retroperitoneal fibrosis.

Discussion

When investigating recurrent abdominal pain, one must consider a broad differential diagnosis to avoid missing rare but potentially treatable aetiologies.

Introduction

Retroperitoneal fibrosis (RF) also known as chronic periaortitis (CP) is a condition of variable aetiology. Approximately 75% of cases are of the idiopathic form (IRF), known as Ormond's disease. The remainder of cases are secondary to pelvic malignancy or infectious/inflammatory aetiologies. Secondary RPF is associated with ergot alkaloid use, biologic use, malignancy, prior radiation and infectious aetiologies such as tuberculosis, actinomycosis and histoplasmosis [1].

This rare fibroinflammatory disorder is characterised by an extensive retroperitoneal fibrotic mass extending inferiorly from the infrarenal para-aortic region. IRF is a benign condition but it may be locally aggressive, often resulting in bilateral ureteric compression and resultant hydroureter, hydronephrosis and impaired renal function, as well as compression of surrounding vascular structures [2–4]. IRF specifically occurs in the presence of a non-dilated aorta [2,3].

The pathogenesis of IRF is not fully understood but there are environmental factors as well as genetic determinants. IRF may be part of immunoglobulin G4 – related diseases (IgG4-RD), a spectrum of fibroinflammatory disorders characterised by inflammation, fibrosis and IgG4 plasma cell infiltration or may present in isolation [5]. IRF is also associated with systemic and localised autoimmune conditions [5].

The incidence of IRF is 0.1 – 1.3 per 100,000 people per year, with a mean onset age of 57 years. Men are affected 3 times more commonly than women [5,6].

Diagnosis may be made by computed tomography (CT) or magnetic resonance imaging (MRI) [1,5]. Biopsy is indicated if the location of suspected fibrosis is atypical [5,6]. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are usually raised at presentation. While the level of elevation may reflect symptomatic disease they are not useful prognostic indicators [1].

Case Report

We present a 74-year-old lady who presented with a one-week history of abdominal pain, post-prandial vomiting and pyrexia, on a background of recurrent similar episodes with indeterminate back-pain and a similar presentation in 2013. Her past medical history was also significant for a left lower limb deep vein thrombosis (DVT) and liver cirrhosis secondary to alcoholic liver disease.

At admission, routine laboratory investigations demonstrated a white cell count (WCC) of 4.6 10^9/L and a CRP of <0.3 mg/L. Creatinine was chronically elevated at 86 μmol/L. CT of the abdomen and pelvis noted symmetrical proliferation of retroperitoneal fat resulting in anteromedial displacement of the kidneys and ureters bilaterally but without obstruction (Fig 1). There was no aneurysmal dilatation of the aorta.

The patient's symptoms settled with conservative management and she was referred to a specialist urology service where she is awaiting further input.

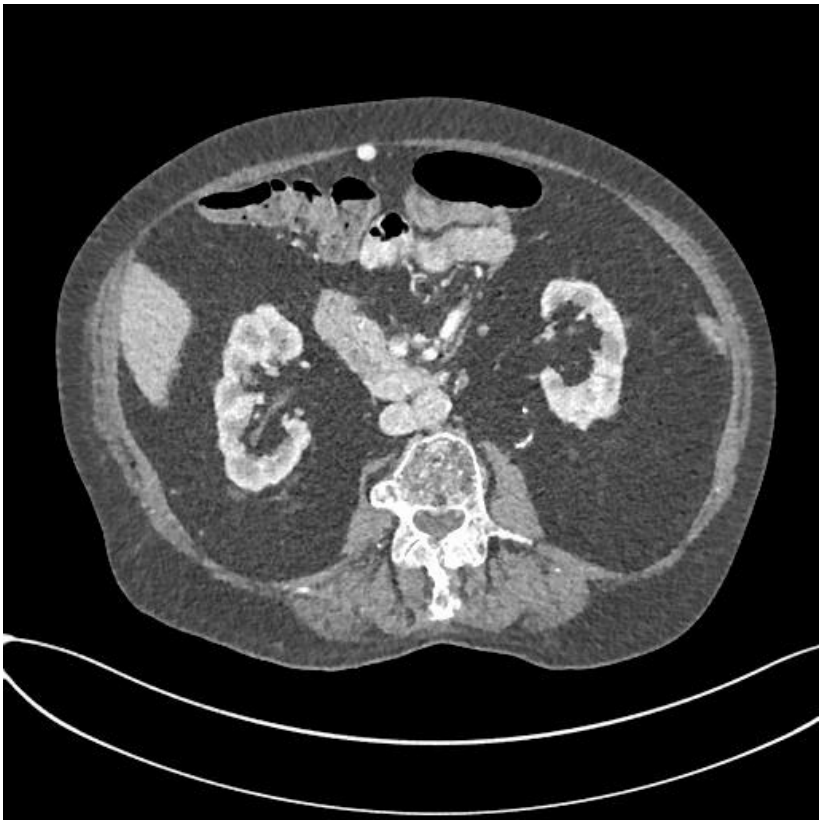

Figure 1. *Axial CT slice demonstrating a diffuse retroperitoneal fatty mass with medial displacement of the kidneys bilaterally.*

Discussion

The classic clinical picture of IRF is characterised by abdominal or back pain, with systemic features including fever and weight loss [5,7]. The extending inflammatory mass may cause unilateral or bilateral ureteral obstruction and if the later can present with acute kidney injury or failure. Seventy-five percent (75%) of patients with RF present with renal impairment. While IRF is not associated with aortic aneurysm, venous complications may occur. Compression of the inferior vena cava may result in lower limb swelling or deep vein thrombosis [1].

The low incidence of IRF has resulted in a paucity of randomised controlled trials [7]. Treatments with corticosteroid and immunosuppression regimes are based on case series data [8] but do appear to be effective [7] . Combination therapy appears to be more effective and allows for more timely tapering of steroids. The most commonly used immunosuppressants used in IRF are azathioprine and mycophenolate mofetil. In cases with obstructive uropathy unilateral or bilateral stenting or percutaneous nephrostomy are usually effective but ureterolysis may be required [7]. Fry et al. [8] have provided a useful algorithm for the management of RF which has been reproduced here with permission (Fig 2).

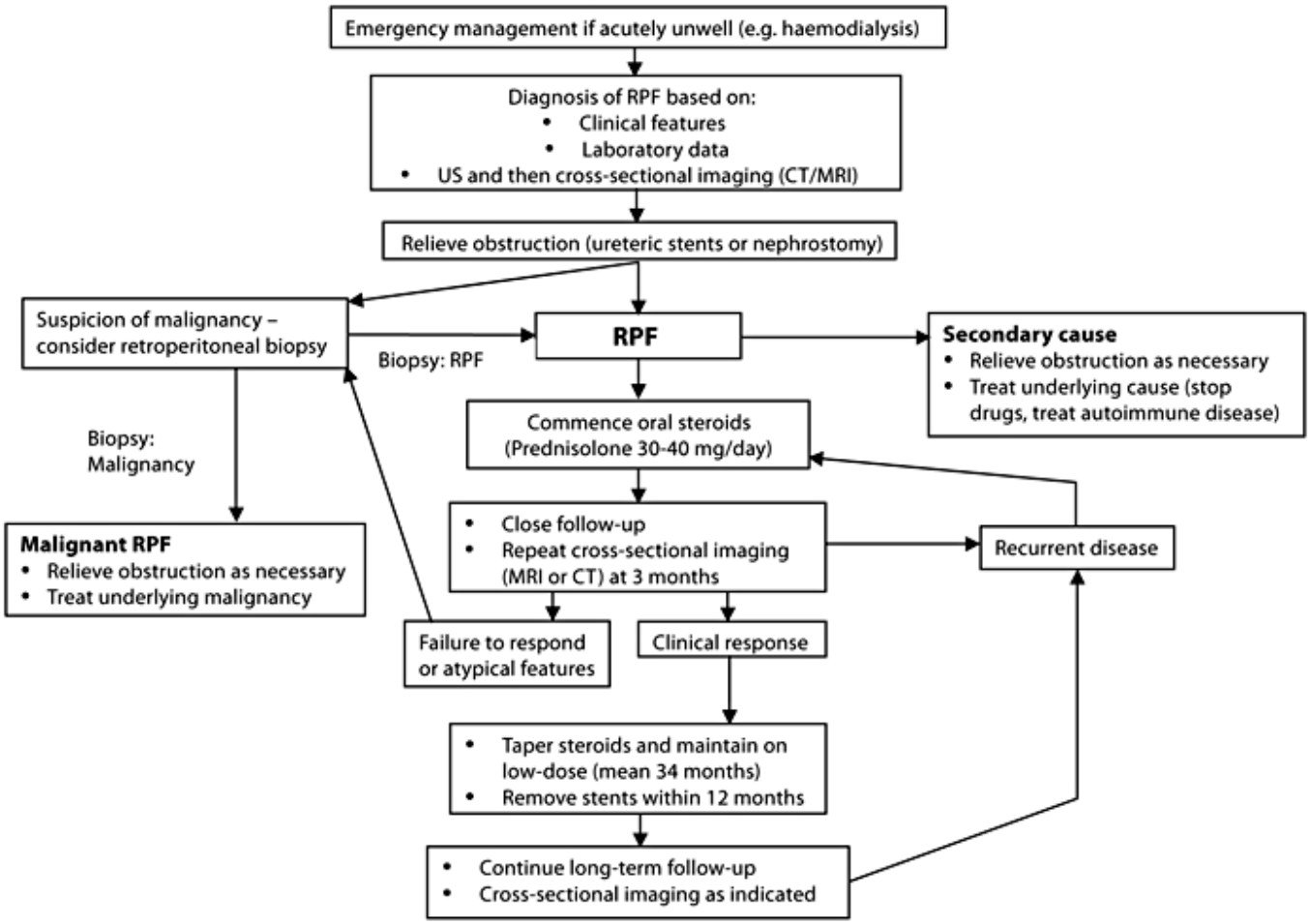

Figure 2. *RF management algorithm*

This case highlights the need to consider CP and IRF in patients presenting with a similar constellation of symptoms and a retroperitoneal fibroinflammatory mass. Considering the diagnosis of IRF in this cohort we may increase the knowledge base and tailor effective treatment regimens for what can be a debilitating disease.

Declaration of Conflicts of Interest:
No conflicts of interest to declare.

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References:

1. Vaglio A, Palmisano A. Clinical manifestations and diagnosis of retroperitoneal fibrosis - UpToDate [Internet]. [cited 2020 Jun 12]. Available from: https://www.uptodate.com/contents/clinical-manifestations-and-diagnosis-of-retroperitoneal-fibrosis?topicRef=7213&source=see_link
2. Idiopathic Retroperitoneal Fibrosis (Ormond's Disease) - American Urological Association [Internet]. [cited 2020 Jun 12]. Available from: https://www.auanet.org/education/auauniversity/education-products-and-resources/pathology-for-urologists/retroperitoneum/idiopathic-retroperitoneal-fibrosis-(ormonds-disease)
3. Nelius T, Reiher F, Lindenmeir T, Kalinski T, Rau O, Filleur S, et al. Die idiopathische retroperitoneale fibrose (Morbus Ormond). Aktuelle Urol. 2006 Jul;37(4):284–8.
4. Netzer P, Binek J, Hammer B. Diffuse abdominal pain, nausea and vomiting due to retroperitoneal fibrosis: A rare but often missed diagnosis. Eur J Gastroenterol Hepatol. 1997;9(10):1005–8.
5. Vaglio A, Maritati F. Idiopathic retroperitoneal fibrosis. J Am Soc Nephrol. 2016;27(7):1880–9.
6. Yachoui R, Sehgal R, Carmichael B. Idiopathic retroperitoneal fibrosis: clinicopathologic features and outcome analysis. Clin Rheumatol. 2016;35(2):401–7.
7. Kisial B, Kruszewski R, Juerk-Urbanowska A, Kidzinski R, Frankowska E, Sulek M, et al. Idiopathic retroperitoneal fibrosis: A case report. Polish Arch Intern Med. 2009;119(10).
8. Fry AC, Singh S, Gunda SS, Boustead GB, Hanbury DC, McNicholas TA, et al. Successful Use of Steroids and Ureteric Stents in 24 Patients with Idiopathic Retroperitoneal Fibrosis: A Retrospective Study. Nephron Clin Pract [Internet]. 2008 Apr [cited 2020 Jun 14];108(3):c213–20. Available from: https://www.karger.com/Article/FullText/119715