

CASE REPORT**IDIOPATHIC RETROPERITONEAL FIBROSIS IN A YOUNG FEMALE;
THE FIRST CASE REPORT IN SRI LANKA**

Pradeep Jayantha Gamage, Saman Chandana, Upul Weerasinghe, Mohan Samarasinghe,
Sampath Jayawardhane, Nadun Dhanushka Gamage, Jalitha Thinna Arachchi and TN Sirimanne

Provincial General Hospital, Badulla, Sri Lanka

Corresponding author: Dr. Pradeep Jayantha Gamage
Email: prdp1990@icloud.com  <https://orcid.org/0000-0002-8455-7521>

Abstract

Idiopathic retroperitoneal fibrosis (IRF) is a rare disease with a better prognosis if identified earlier and treated promptly. This is thought to be secondary to an autoimmune process resulting in fibrosis, causing obstructive uropathy. Radiological aspects play a major role in diagnosis and follow up. We present a young female patient who was diagnosed with idiopathic retroperitoneal fibrosis and treated successfully. This case highlights the fact that diagnosing IRF is a result of a cascade of reasonable investigations and early diagnosis prevents irreversible renal damage. Classic radiological features with histology provide a guide to the proper diagnosis, and treatment with corticosteroids gives a marked response.

Keywords: radiological diagnosis, renal failure, corticosteroids

Introduction

Retroperitoneal fibrosis is a rare disease with a better prognosis if identified earlier and treated promptly¹. A majority of cases are idiopathic, whilst a secondary cause is identified in one-third of the cases. The non specific presentation often leads to a delay in diagnosis^{1,2}. Published literature on this phenomenon is limited, and case reports on IRF would pave the way for development of proper management guidelines.

Case presentation

A 39-year-old female presented with changes in bowel habits and mild lower abdominal pain of two weeks duration. Her inflammatory markers were elevated (C-reactive protein - 156 mg/L) and she had a highly elevated serum creatinine level (1120 µmol/L).

Ultra sound scan (USS) of the abdomen showed bilateral hydronephrosis with acute renal parenchymal changes (Figure 1). Computed tomography (CT) of the abdomen was done afterwards to look for the cause of obstruction (Figure 2). Meanwhile the patient underwent bilateral double "J" stenting to relieve the ureteric obstruction and her renal functions



This work is licensed under a Creative Commons Attribution 4.0 International License (CC BY)

gradually improved afterwards. The CT showed a soft tissue density mass in the retroperitoneal space at the level of the aortic bifurcation, along with medial deviation of ureters and tapering of ureters, with associated hydronephrosis and hydroureter.



Figure 1: USS of the abdomen showing right sided hydronephrosis

The patient underwent a histological assessment of the lesion with a mini laparotomy. Microscopic sections revealed fibrous tissue fragments with mixed inflammatory cell (lymphocytes, plasma

cells and eosinophils) infiltration. Fibroblastic proliferation was evident with no features of malignancy.

The patient was investigated to exclude secondary causes, and all investigations were reported as negative (antinuclear antibody, rheumatoid factor, blood picture, screening for tuberculosis) and there were no significant drug history.

The patient was treated with oral prednisolone 60mg daily and follow up CT was done three months later, while she was on treatment (Figure 3). This showed a marked reduction of the size of the mass along with improvement of hydronephrosis and hydroureter. The serum creatinine was 62 micromoles per liter and CRP was 37 mg/L after three months, indicating the marked improvement.

Discussion

Retroperitoneal fibrosis is a rare disease with an estimated prevalence of 1.38 in a

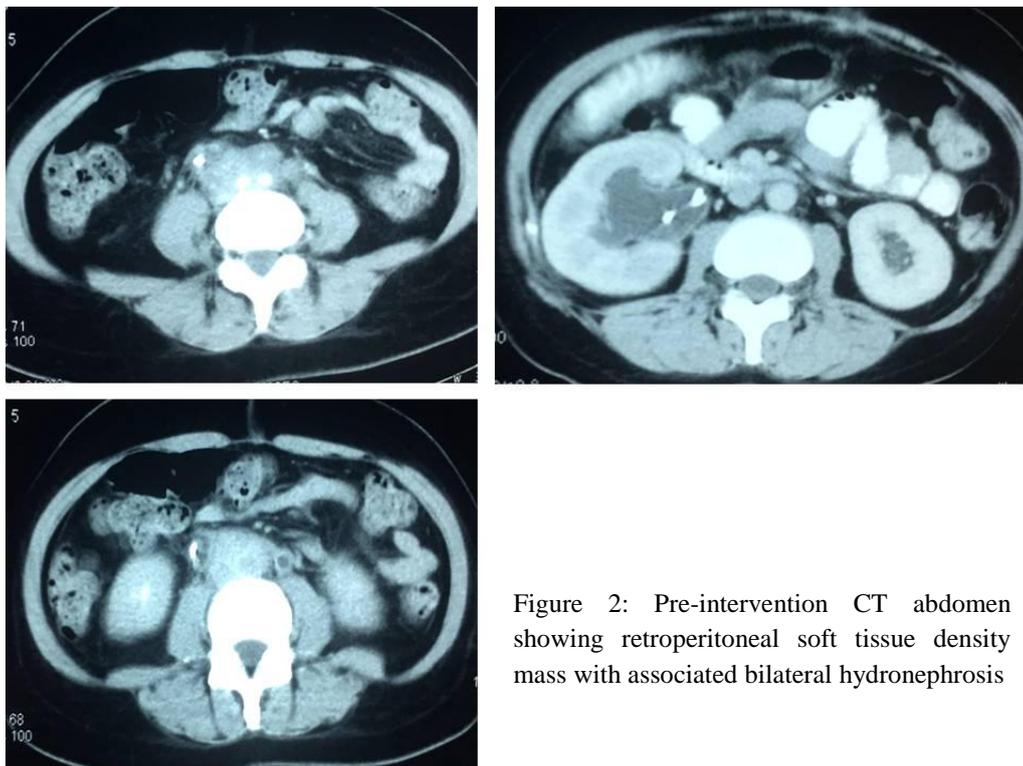


Figure 2: Pre-intervention CT abdomen showing retroperitoneal soft tissue density mass with associated bilateral hydronephrosis

100,000 population¹. There are no published cases in Sri Lanka. It is more predominant in males within 40 years to 65 years of age, though our patient was a female of 37 years of age, highlighting the rarity of this case presentation^{1,2}.

these are non-specific. USS has a minor role in diagnosis, whereas CECT (Contrast enhanced CT) with MRI (Magnetic Resonance Imaging) plays a major role¹. MRI is more advantageous compared to CECT, because of the low radiation exposure and more detailed anatomy of

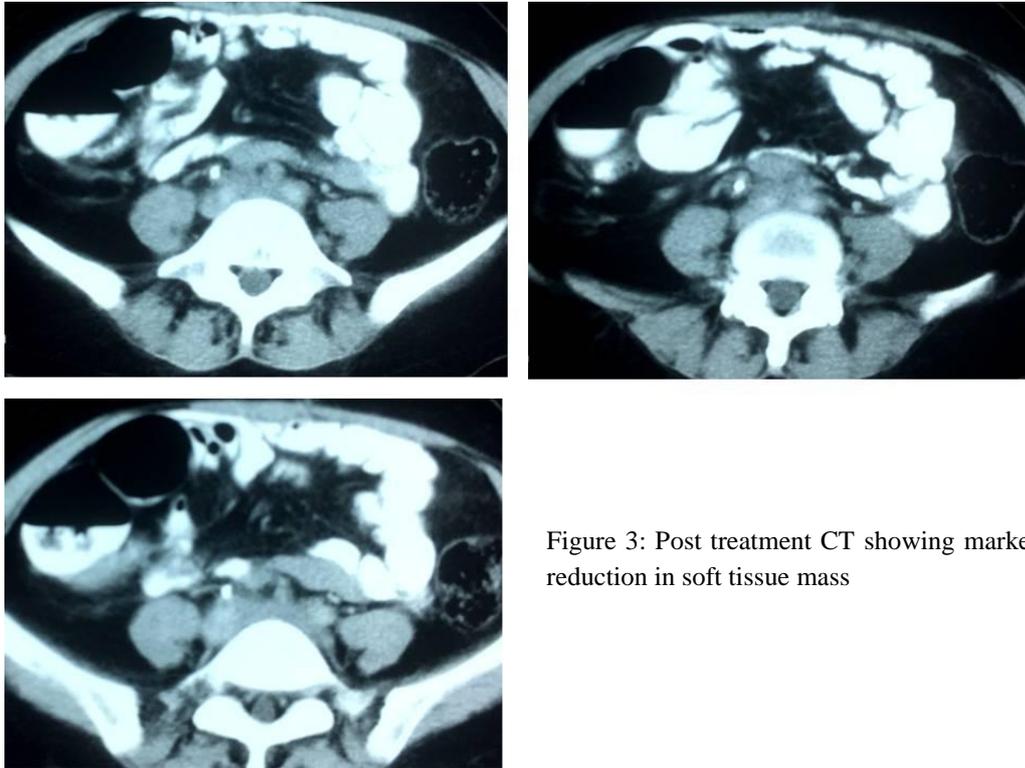


Figure 3: Post treatment CT showing marked reduction in soft tissue mass

The presentation may vary according to the underlying cause. The non-specific presentation leads to unnecessary investigations, leading to diagnosis delay³. This patient's main concern was change of bowel habits with a mild non specific lower abdominal pain. The majority of retroperitoneal fibrosis is idiopathic (more than 66%), whilst there are multiple secondary causes such as malignancies, drugs and infections².

Associated pathologically changes include inflammatory fibrosis of the retroperitoneal fat. The inflammation or the periaortitis occurs due to atherosclerotic plaques in the aorta^{1,4}. In most patients, inflammatory markers are increased. Special testing for autoimmune processes are positive in some cases, but

entrapped structures^{1,5}. But both the imaging modalities are not very sensitive regarding differentiation of IRF from a malignant lesion. Most authorities recommend confirmation of the diagnosis histologically, before commencement of treatment⁵. Open biopsy is preferred for obtaining histology from all the modalities⁵. Only CECT was performed in this patient due to limited availability of MRI, and this demonstrated a soft tissue density mass covering the aorta, inferior vena cava and both ureters, with associated bilateral obstructive uropathy. As secondary causes such as malignancies needed to be excluded, a biopsy was performed with the informed written consent of the patient, which confirmed the diagnosis of idiopathic retroperitoneal fibrosis.

Corticosteroids have been used as the main modality of treatment in the literature, with more than 200 reported cases showing a marked response rate. This treatment is associated with a 80% to 85% response rate with marked improvement of symptoms⁵. Our patient was commenced on oral prednisolone 60 milligrams per day, with bilateral double "J" stenting to relieve the ureteric obstruction. She was followed up with CECT after 3 months. This showed significant reduction of both the soft tissue mass size and degree of obstructive uropathy, with improvement of renal functions and inflammatory markers.

Conclusions

IRF should be considered as a differential diagnosis in young patients presenting with bilateral hydronephrosis without an identifiable cause. CECT or MRI may support to differentiate IRF from a malignancy. Histological confirmation should be carried out where imaging is suggestive.

IRF has a marked response to oral corticosteroids and imaging and inflammatory markers are useful in future follow up.

References

1. Caiafa RO, Vinuesa AS, Izquierdo RS, Brufau BP, Ayuso Colella JR, Molina CN. Retroperitoneal fibrosis: role of imaging in diagnosis and follow-up. *Radiographics*. 2013 Mar 5;33(2):535-52. <https://doi.org/10.1148/rg.332125085>
2. Majdoub AE, Khallouk A, Farih MH. Retroperitoneal fibrosis: about 12 cases. *The Pan African medical journal*. 2017;28:194. <https://doi.org/10.11604/pamj.2017.28.194.10092>
3. Van Bommel EF, Jansen I, Hendriksz TR, Aarnoudse AL. Idiopathic retroperitoneal fibrosis: prospective evaluation of incidence and clinicoradiologic presentation. *Medicine*. 2009 Jul 1;88(4):193-201. <https://doi.org/10.1097/MD.0b013e3181afc420>
4. Gilkeson GS, Allen NB. Retroperitoneal fibrosis: a true connective tissue disease. *Rheumatic Disease Clinics*. 1996 Feb 1;22(1):23-38.
5. Premachandra ND, Perera ND. Retroperitoneal fibrosis—an unsolved mystery. *Sri Lanka Journal of Urology*. (Vol 2010) 2012 Jan 31;11(1):7-13.