A Retrospective Analysis of Retroperitoneal Fibrosis in North West London

Tomlinson JAP[1,2,3], Barwick T[3], Cairns T[3], Cook T[3], Gibbons N[3], Hrouda D[3], Mason J[3], McAdoo S[2,3], Pusey C[2,3], Tran K[3], Yusuf S[3], Tam FWK[2,3]

[1]Institute of Clinical Sciences, Imperial College, London, UK; [2]Renal and Vascular Inflammation, Faculty of Medicine, Imperial College London;
[3]Imperial College NHS Healthcare Trust, London, UK; Faculty of Medicine, National Heart and Lung Institute, Imperial College London

Introduction

Retroperitoneal fibrosis (RPF) is a rare fibro-inflammatory condition that develops around the abdominal aorta and frequently encapsulates one or both ureter(s) causing obstructive nephropathy. The aetiology of RPF is uncertain, the complication rate is high and the management is complex, requiring expertise from multiple specialties.

Objective

To better understand the disease burden and clinical characteristics of RPF in North West London

Methods

We performed a retrospective analysis of RPF cases treated at Imperial College Healthcare Trust and referring hospitals across North West London. RPF cases were identified through International Classification of Disease, Tenth Revision coding (ICD-10 N13.5) and specialty databases at Imperial College Healthcare Trust and referring hospitals, recorded between 1st January 2005 and 31st December 2016. Only cases with available imaging or reports compatible with RPF were included. Clinical data were extracted from paper case notes and digital records. Duration of follow-up was defined from the date of diagnosis by imaging to the last clinical attendance or death.

Results summary

A total of 72 retrospective RPF cases were identified over a 12 year period with a mean follow-up period of 90 months (range 1 – 468 months).

Table 1) Demographics

Mean age at diagnosis (years)	56 (range 26 - 82)
Male (%)	61
Ethnicity	
White British	30 (42%)
Black African	11 (15%)
Asian	9 (13%)
Black Caribbean	7 (10%)
White Other	5 (7%)
North African	4 (6%)
Middle-Eastern	3 (4%)
White Irish	3 (4%)

Table 2) Risk factors

Smoking history	45 (63%)
Pre-existing autoimmune disease or positive serum antibodies	30 (42%)
Known asbestos exposure	3 (2%)

Figure 1) Symptoms on presentation. Pain was the most common physical symptom recorded in 75% of individuals

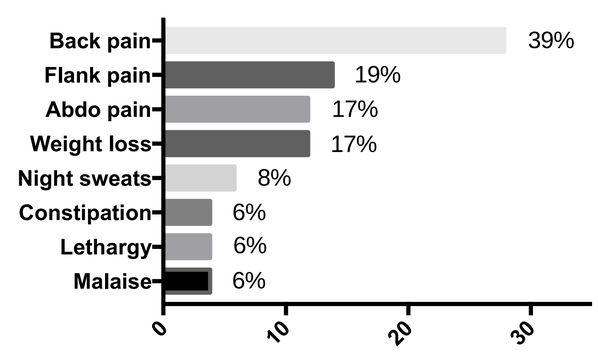


Figure 2) Imaging. A diagnosis of RPF was commonly first indicated by computed tomography; Figure 2a) and disease activity assessed using FDG-PET (Figure 2b).

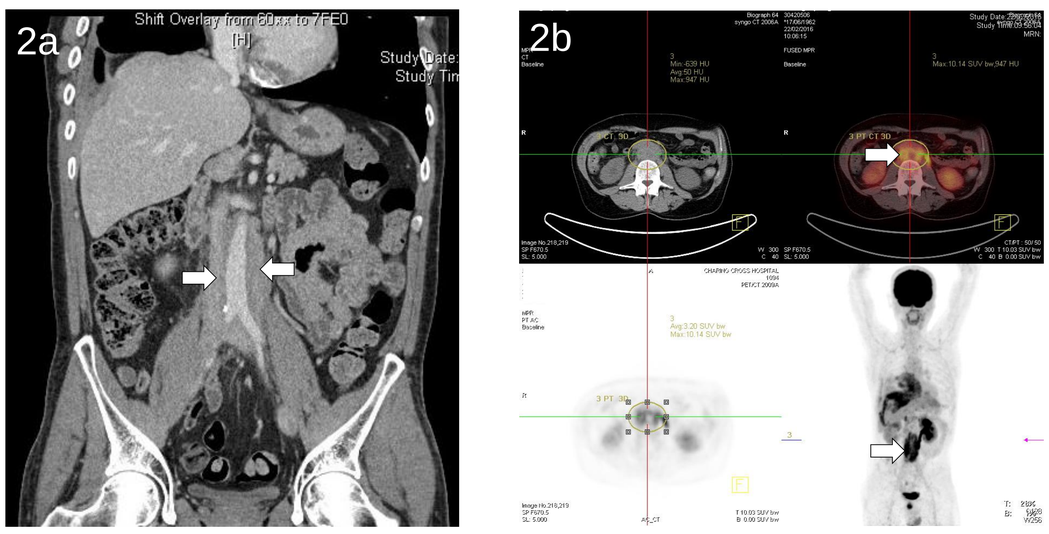
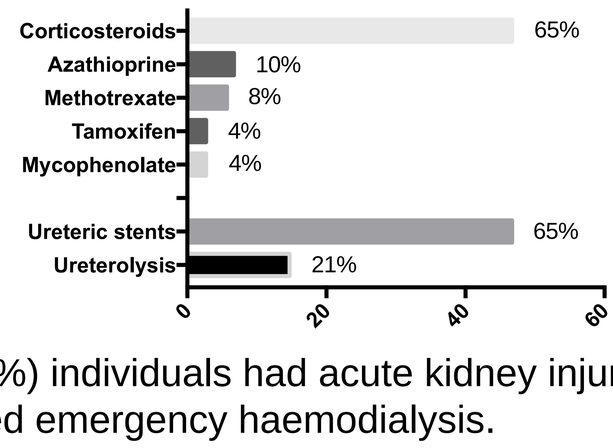


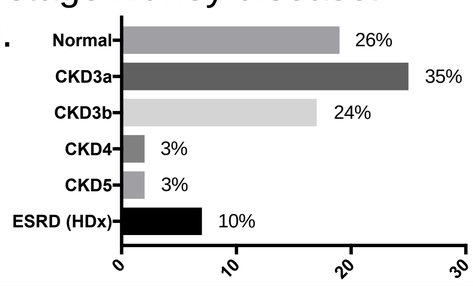
Figure 3) Treatment



Renal outcomes

At presentation 50 (69%) individuals had acute kidney injury and 4 patients (6%) required emergency haemodialysis.

Figure 4) On follow-up, significant renal impairment (eGFR<60mL/min/1.73m^2) was present in 51 (71%) of patients and 10 (14%) reached end-stage kidney disease. There were 8 deaths (11%).



Conclusion

To our knowledge, this is the first large multi-ethnic RPF retrospective study to be reported from the UK. Our ongoing work to improve understanding and outcomes for patients with RPF includes: founding a national RPF rare disease registry (RaDaR, Renal Association and Registry; http://rarerenal.org/rare-disease-groups/retroperitoneal-fibrosis-rdg/), an international online patient-focused study (see Poster PO-209) and formation of an RPF multi-disciplinary clinical and research group at Imperial College Healthcare Trust.