Exceptional Case

No way in & no way out: a case of renal failure due to both pre- and post-renal obstruction

Oliver Monfredi, Colin Jones and Niall Warnock

Department of Renal Medicine, York Hospital, Wigginton Road, York YO31 8HE, UK

Keywords: Erdheim–Chester disease; histiocytosis; imaging; Langerhans cell histiocytosis; magnetic resonance angiography; renal failure

Introduction

Causes of renal failure can be classified as pre-renal, intra-renal and post-renal. We discuss a case of combined pre-and post-renal failure due to an unusual presentation of a rare disease. The case illustrates the usefulness of combining imaging modalities in investigating complex renal disease.

Case discussion

A 46-year-old lady presented to the renal physicians in 2002 with a biopsy diagnosis of Langerhans cell histiocytosis (LCH). Initial presentation had been with maxillary and retro-orbital disease. A CT scan of the thorax/abdomen/pelvis in 2002 had shown extensive irregular soft tissue around and extending into the sinuses of both kidneys (Figure 1), with renal calyceal but not pelvic dilatation, as well as disseminated involvement including bone, mediatinum and lungs. Her serum creatinine was normal and a MAG3 renogram showed no evidence of functional obstruction. She had no symptoms other than occasional loin discomfort.

Three years later she developed worsening hypertension, a rising serum creatinine and pulmonary oedema. MR angiography showed bilateral tight renal artery stenoses. It again showed the unusual pattern of strikingly dilated calyces but collapsed renal pelves. Dynamic renography and antegrade pyelography (Figure 2) indicated obstruction at the level of the renal pelvis bilaterally. Consequently, percutaneous nephrostomies were placed, but there was no improvement in renal function.

In view of the episode of pulmonary oedema, renal artery angioplasty was attempted. Digital subtraction angiography confirmed critically tight but focal renal artery stenoses (Figure 3). After intra-arterial heparin, the left-sided stenosis was traversed with a guide wire, but even this was sufficient to occlude the artery, and it was not possible to advance any catheter through the lesion. On removing the guide wire the artery remained occluded. Treatment on the right was not attempted. Despite maximal medical treatment including diuretics, ACE inhibitors and haemodiafiltration, the patient developed refractory episodes of pulmonary oedema and died. Some diagnostic uncertainty remained regarding whether the diagnosis was of true LCH or of the rarer Erdheim–Chester disease (ECD), since initial biopsies had not been exposed to the full panel of immunohistochemistry and electron microscopy required to differentiate these two closely related disorders (see discussion below).

Correspondence and offprint requests to: Oliver Monfredi, Department of Renal Medicine, York Hospital, Wigginton Road, York YO31 8HE, UK. Tel: +44-01904-631313 Ext 5374; Fax: +44-01904-726354; E-mail: oliver.monfredi@manchester.ac.uk

© The Author (2008). Published by Oxford University Press on behalf of ERA-EDTA. All rights reserved. For Permissions, please e-mail: journals.permissions@oxfordjournals.org
cells (LCs) and mature eosinophils. Initially described in 1868 by Paul Langerhans, the condition is now known to result from deficient presentation of antigen by the diseased LCs. Attempts at classification of LCH have been confusing. It is accepted that histiocytic disorders may be split into three different groups [3]:

1. dendritic cell histiocytoses
2. erythrophagocytic macrophage disorders
3. malignant histiocytoses

LCH belongs to group 1 and spans a spectrum from limited and indolent disease, to acute and fulminating. Letterer–Siwe disease, Hand–Schüller–Christian disease, histiocytosis X and eosinophilic granuloma were names previously used to signify different forms of LCH, but are now largely redundant. The current preferred means of classifying LCH is based on whether disease involves single or multiple sites, and single or multiple organs. Prognosis and treatment depend on the extent of the disease when classified like this, and whether certain high-risk organs are involved (e.g. liver, spleen, lung, bone marrow). Common presenting complaints include skin rash, dyspnoea, painful bony lesions, lymphadenopathy, weight loss, proptosis and polydipsia with polyuria (diabetes insipidus). Diagnosis relies on histological demonstration of abnormal Langerhans cells (show pathognomonic Birbeck granules on electron microscopy, CD1a-, CD14-, and CD52-positivity, and finally staining for antibodies to the granule protein 'Langerin'). Effective treatment can be given in the form of combination chemotherapy, e.g. etoposide, vinblastine and prednisolone. Survival in disseminated disease is ≤50% at 5 years.

Previous authors [4–9] have described renal artery stenosis and chronic kidney disease in ECD, a rare, class 2, non-LCH typified by extensive retroperitoneal infiltration with foamy histiocytes. This is often mistaken for LCH, and is differentiated by histological demonstration of absent Birbeck granules and absent S100 protein staining. In a case series and literature review reported by

---

**Fig. 2.** Left antegrade pyelogram showing extrinsically compressed pelvis and upper ureter with dilated calyces and normal lower ureter.

**Fig. 3.** Selective left renal arteriogram confirming critical stenosis. Appearances were similar on the right.
Haroche et al. [10] of 72 patients with cardiovascular complications of ECD, 8% had renovascular hypertension related to peri-renal artery stenosis. There are few reported cases of renal artery stenosis and severe renal impairment in true LCH. The above case illustrates well the need to employ all imaging modalities to fully assess extremely challenging cases of pre- and post-renal obstructive disease.

Conflict of interest statement. None declared.

References

3. www.histio.org

Received for publication: 13.2.08
Accepted in revised form: 28.2.08