Rare disease

Fever of unknown origin: a rare retroperitoneal cause

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Correspondence to Mr Jason M Fitchett, jasonfitchett@hotmail.com**Summary**

The authors present a patient with fever of unknown origin and vague loin pain. Baseline investigations revealed elevated inflammatory markers and imaging eventually identified a renal mass. Radical nephrectomy was performed at a collaborative surgical list involving both the urology and vascular surgical teams. Histological examination confirmed a retroperitoneal paraganglioma and the patient made a rapid recovery and remains well at follow-up. This case draws attention to the rare diagnosis of retroperitoneal paraganglioma and the potential for diagnostic delay. Furthermore, the vital multi-disciplinary approach to the optimum management of patients with such tumours is highlighted.

BACKGROUND

We present a patient with a PUO (fever of unknown origin) that failed to settle on conservative treatment. Investigations eventually identified a renal mass. Only after surgical resection was a diagnosis considered and confirmed to be a paraganglioma. Although rare, such a diagnosis should have been considered, and would have substantially reduced time to treatment, and her subsequent discharge.

CASE PRESENTATION

A previously well 38-year-old housewife initially presented with a sore throat and pain radiating to her ear. Examination revealed right-sided cervical lymphadenopathy, and inflamed throat. Respiratory examination was unremarkable, but her white cell count was 16 000. She was commenced on antibiotics and steroids, improved and was discharged the next day. Ten days later she was readmitted with sore throat and vague loin pain. She was feverish (38.5°C), with elevated inflammatory markers including the C reactive protein. She was commenced on further antibiotics. Although clinically she improved, her fever and left-sided loin pain persisted.

INVESTIGATIONS

Basic routine investigations included chest and abdominal radiographs, mid-stream urine sample and haematological renal and liver function tests were all normal. However, she proceeded to an ultrasound scan that identified a non-specific renal mass, possibly an abscess. Since the fever continued, an abdominal CT scan was performed (figure 1) that localised a 6 cm mass at the lower pole of the left kidney and abnormal para-aortic lymphadenopathy.

TREATMENT

Eighteen days after initial presentation, a urology opinion was sought, and the case discussed at an MDT (multi-disciplinary team) meeting. The mass was thought to be a tumour and the patient underwent a CT guided biopsy that demonstrated sarcoma.

Six weeks after initial presentation, the case was discussed at a specialist sarcoma MDT meeting at the regional oncology centre. Radical nephrectomy was indicated as further imaging had ruled out metastatic disease. In view of the tumour proximity to major vascular structures the nephrectomy was performed at a collaborative surgical list involving both the urology and vascular surgical teams.

OUTCOME AND FOLLOW-UP

Local tumour clearance was achieved and the histology demonstrated a highly vascular tumour independent of the kidney that infiltrated adjacent structures. There was no vascular invasion and the cell cycle fraction was low, equating to slow growth. A second pathological opinion was sought, confirming a diagnosis of a retroperitoneal paraganglioma of uncertain malignant potential.

She made a rapid recovery and remains well at regular follow-up. A repeat CT scan at 6 months postoperatively showed no radiological evidence of recurrent disease, and following MDT discussion it was decided that further imaging should be guided by clinical assessment in order to extend the interval between CT scans to limit radiation exposure.

Figure 1 CT scan horizontal plane with tumour.

DISCUSSION

Paragangliomas are rare tumours that usually arise from catecholamine producing chromaffin tissue not located within the adrenals. Most occur in the head and neck, with chemodectoma being the well-known example. However, approximately 10% of these extra adrenal tumours arise in the retroperitoneal tissues.^{1,2} They may be classified as functional or non-functional according to their catecholamine secretory status, and therefore urine testing for catecholamines should be included in the investigations.³

The clinical presentation of these tumours is complicated by the wide variety of tumour sites and thus presentation. Furthermore, the diagnosis of malignancy is difficult histologically, requiring extensive capsular invasion.⁴ Metastases occur in up to 38%³ of cases, and confirm malignant potential. It is suggested that the aortic sympathetic group may have a greater malignant potential compared with other anatomical sites,³ possibly because retroperitoneal tumours can enlarge substantially before becoming apparent.

Our case presented with PUO and flank pain. The PUO may or may not be a direct result of the tumour, however, since resection she has remained afebrile. A review of 22 patients with retroperitoneal paragangliomas by Cunningham *et al*² identified the most common presenting symptoms to be hypertension or a palpable mass, but many presented with vague symptoms. The diagnosis in our patient was certainly delayed by her vague symptoms, and even when the mass was identified it was initially thought to be a tumour of renal origin.

Treatment for these conditions is difficult, but the literature would suggest that radical surgical excision is the treatment of choice as 5-year survival is vastly superior for those who undergo resection.² The use of radiotherapy has also been described.¹ Cunningham *et al*² suggest that a tumour larger than 7 cm is likely to require adjacent organ resection to completely excise the mass. They advise a

collaborative approach for resection when required, such as demonstrated in our case with joint vascular and urological operative input. This multi-specialty approach may indeed be vital for the treatment of paragangliomas as the quality of surgical resection is highly likely to affect long-term survival.⁵

These tumours are slow growing and therefore have a natural history of late recurrence, often many years after initial resection. Life-long follow-up is therefore indicated.²

Learning points

- ▶ Retroperitoneal paraganglioma are rare tumours that should be considered in the differential diagnosis of a retroperitoneal mass.
- ▶ Surgical resection is the treatment of choice, and complex cases require multi-specialty input.
- ▶ Life-long follow-up is required as recurrence may be very late.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. Bastin KT, Steeves RA, Gilchrist K. Retroperitoneal paragangliomas: natural history and review of the literature. *Wis Med J* 1995;**94**:83–5.
2. Cunningham SC, Suh HS, Winter JM, *et al*. Retroperitoneal paraganglioma: single-institution experience and review of the literature. *J Gastrointest Surg* 2006;**10**:1156–63.
3. Butterworth JE. Letter: Retroperitoneal paraganglioma. *Urology* 1975;**6**:131.
4. Madrigal B, Vara A, Fresno M, *et al*. Non-secreting, extra-adrenal retroperitoneal paragangliomas. *Arch Esp Urol* 2002;**55**:543–7.
5. Matsui H, Ikeuchi S, Onoda N, *et al*. Malignant paraganglioma of the retroperitoneum with lung metastases: a 13-year survivor after radical surgery. *Asian J Surg* 2007;**30**:75–9.

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