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Mature ganglioneuroma of the adrenal gland as a new rare cause of visible haematuria: A case report & literature review



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ABSTRACT

INTRODUCTION: Ganglioneuromas are benign tumours of the sympathetic ganglia and the adrenal glands medulla. We describe a case of a fit and well 18 year old Caucasian male patient who initially presented to primary care with intermittent episodes of painless frank haematuria as well as some non-specific right-sided loin pain.

PRESENTATION OF CASE: In this 18 year old man, initial ultrasound investigations at a 'one stop haematuria' clinic revealed the presence of an echogenic solid mass of 120 × 110 × 90 mm around the upper pole of the right kidney. A CT scan of the abdomen proved inconclusive to further determine the aetiology of the mass. Following a local multidisciplinary meeting (MDT) an MRI of the abdomen was undertaken which confirmed the presence of a large right adrenal mass. A biopsy was taken to determine the histology of the mass confirming a mature ganglioneuroma. The patient subsequently underwent surgery within 6 weeks of having presented to his general practitioner.

CONCLUSION AND DISCUSSION: The surgery was uncomplicated and excision of the mass was made via a thoraco-abdominal approach. The patient recovered well post operatively and was discharged home within 8 days with outpatient follow-ups organised.

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1. Background

Ganglioneuromas (GN) are benign tumours originating from the sympathetic ganglia and the adrenal medulla [1]. These tumours usually present in children and young adults, usually as mature GN before the age of 20. Females are commonly more prone to being affected by GN than males [1] and the vast majority are situated in the retroperitoneum. They are often discovered incidentally as these tumours commonly remain hormonally silent [2] and histological examination remains key to differentiating between a benign GN and the more malignant neuroblastoma [3]. Adrenal GNs, however, remain rare and have not been widely reported in surgical literature [4]. In this paper, we present the first reported case in medical literature of a mature GN originating from the adrenal medulla in a young adult male presenting with visible haematuria.

2. Case report

An 18-year-old male with no significant past medical history of note was referred to a 'one-stop' haematuria clinic by his General

Practitioner. The patient had become concerned after experiencing intermittent episodes of painless frank haematuria for over one week. There was no history of any prodromal illness, upper respiratory tract infection or trauma. The patient is a non-smoker and there is no family history of urological or malignant disease.

On examination blood pressure and pulse were normal. Urinalysis demonstrated non-visible haematuria with no evidence of glucose, protein or infection. There was a large palpable mass in the right loin which had recently started causing him some discomfort. A urinary tract ultrasound revealed a large well defined echogenic solid mass measuring approximately 12 × 11 × 9 cm located adjacent to the upper pole of the right kidney. The large mass was also found to be compressing and distorting the outline of the right lobe of the liver. Fortunately, both kidneys were of normal appearance on ultrasound. Flexible cystoscopy was normal.

This gentleman underwent a subsequent CT scan of the abdomen and pelvis which confirmed the presence of the large right renal mass seen on the ultrasound (Figs. 1 and 2). However, the CT scan did not determine the exact aetiology of the mass, thus making it difficult to differentiate between a haemorrhagic renal cyst, for example, or an adrenal mass. Following discussion at the uro-oncology multi-disciplinary team (MDT) meeting the consensus was that he should have an MRI of the abdomen which confirmed the presence of an encapsulated mass arising from the right adrenal (Fig. 3).

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Fig. 1. Cross-sectional CT abdomen with contrast showing right retroperitoneal mass (red arrow).

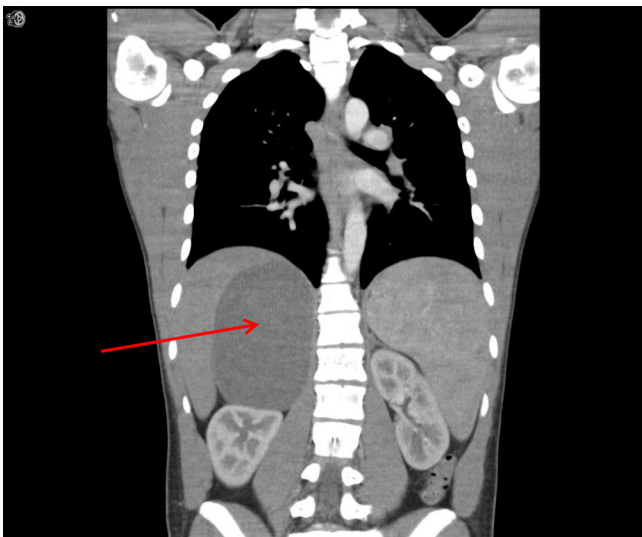


Fig. 2. Coronal CT abdomen with contrast showing the same right supra-renal mass (red arrow).



Fig. 3. Coronal MRI of the abdomen showing right supra-renal mass (red arrow).

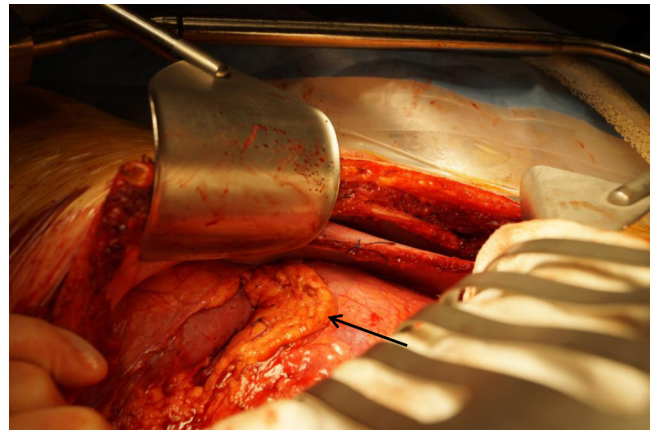


Fig. 4. Right thoraco-abdominal incision showing right supra-renal mass with yellow sleeve of normal-looking adrenal tissue (black arrow).

Endocrine tests such as serum cortisol, 24 h urinary catecholamines and vanillylmandelic acid (VMA) excretion were all within normal parameters pointing away from a functional tumour. This was further confirmed by a PET CT scan, demonstrating normal physiological uptake photopaenia from the large right adrenal mass making the poorer prognostic diagnosis of a neuroblastoma less likely. However, further MDT discussion recommended a right adrenal mass core biopsy would be required to determine if the mass was suggestive of a functional or non-functional tumour prior to surgical removal of the mass and this confirmed a GN with mature elements. There is the possibility that the biopsy may have missed potential malignant areas of the mass given its size so the consensus was that it should be completely excised.

An elective open right adrenalectomy via a right thoraco-abdominal incision was performed (Fig. 4) and the adrenal mass excised en bloc (Fig. 5) without complication. The patient had a chest drain inserted prior to wound closure and this was removed day 2 post-op. He was discharged home well on day 8 and histological analysis confirmed the a 14 × 11.5 × 7.5 cm mature ganglioneuroma, with no neuroblastomatous component – completely benign. He remains well 5 months post-op at last out-patient review.

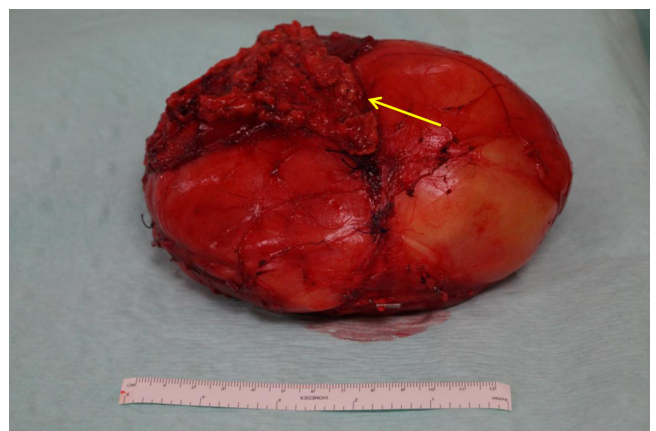


Fig. 5. Mature GN following open excision measuring up to x cm in maximum dimension with evidence of normal adrenal tissue on top of it (yellow arrow).

3. Discussion

Ganglioneuromas are rare benign tumours which have been widely reported in the literature and in the majority of cases they are asymptomatic or found incidentally [5]. It is even rarer to diagnose them following investigations for haematuria. A PUBMED search was performed for 'adrenal ganglioneuroma haematuria'. In 2000 Leavitt et al. reported the case of a 67 year old female who underwent surgical excision of an adrenal mass after presenting with asymptomatic non-visible haematuria [6]. In 2003 Fujita et al. reported two cases of GN of which one of the cases involved a 29 year old with an 8 cm right adrenal GN who also presented with asymptomatic non-visible haematuria [7]. Our case is the first reported in the literature of adrenal GN presenting with visible haematuria.

3.1. Should benign lesions on biopsy be excised?

In our case biopsy of the mass demonstrated mature GN which is known to be a benign lesion so there is the argument that putting a patient through surgery with the potential risks associated with it is unnecessary. However, composite tumours have been reported containing pheochromocytoma (PC) and GN as in a case report by Tohme et al. in 2006, albeit presenting with clinical signs of catecholamine hypersecretion [8]. Interestingly, a year earlier Bernini et al. reported on a similar case in which a composite GN tumour was associated with a non-functioning PC [9]. Therefore, the only definitive way of ensuring a tumour is completely benign and without any malignant potential is for complete excision and subsequent full histological analysis.

3.2. How should these masses be removed?

There are many documented cases of open surgical excision for adrenal GNs and in the era of minimally invasive surgery the laparoscopic approach is increasingly being more adopted. Zografos et al. reported on the complete laparoscopic excision of a 13 cm right adrenal GN in a 23 year old female using a trans-abdominal approach which went without complication [10]. This is certainly an option for small to medium-sized tumours and the patient was discharged well day 2 post-op. More recently, however, Abraham et al. described a similar case in a 33 year old female patient with a 17 cm left adrenal GN which was removed laparoscopically via the same approach without incident [11] suggesting this approach can be used for the larger tumours as well. The feasibility of the laparoscopic approach is further underlined in a series of 12 cases reported by Zhang et al., the largest being 16.8 cm, with a mean hospital stay of just over five days [12]. There also cases of successful laparoscopic procedures on paediatric patients too with Mukai et al., reporting on a successful laparoscopic adrenalectomy in a 14 year old girl for removal of a 4.4 cm GN [13].

In our particular case an open approach was recommended due to the large size of the tumour and its apparent adherence to the IVC on pre-operative cross-sectional imaging.

Overall, the method of excision can be either open or laparoscopic and will depend on surgeon experience and local facilities available.

4. Conclusion

Cross-sectional imaging by way of CT or MRI is essential but not diagnostic. We recommend thorough pre-operative work-up with testing for serum cortisol, urinary catecholamines and complete surgical excision of such large adrenal masses in order to obtain a definitive diagnosis even following benign biopsy results as some tumours can demonstrate heterogeneous architecture with potentially malignant areas that can be missed on biopsy. In addition, and although rare, it has been reported in the literature that some have the potential to metastasise. Lastly, the diagnosis of mature adrenal GN should be considered in patients presenting with loin mass and visible haematuria.

Conflict of interest

None.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at <http://dx.doi.org/10.1016/j.ijscr.2015.06.027>

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