African Journal of Urology

CASE REPORTS Open Access

Spontaneous haemorrhage of an adrenal angiomyolipoma: case report

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Abstract

Background: Angiomyolipomas are rare mesenchymal tumours arising from the perivascular epithelioid cells consisting of variable amounts of adipose, thick-walled blood vessels and smooth muscle cells. These benign tumours commonly occur in the kidney with only a few case reports of adrenal angiomyolipomas which have the potential to reach a large size and haemorrhage.

Case presentation: A 45-year-old lady presented with a 3-week history of right loin pain, nausea and vomiting. A CT scan revealed a right adrenal angiomyolipoma measuring 6.3 × 6.8 cm with associated haemorrhage. The lesion was successfully treated with right open adrenalectomy, and histology confirmed the diagnosis of adrenal angiomyolipoma. The patient remained well with no evidence of recurrence at the 36-month follow-up.

Conclusion: Adrenal angiomyolipomas are rare benign tumours that have the ability to reach a large size and potential to bleed. Here, we report the second case of spontaneous haemorrhage in an adrenal angiomyolipoma, which was successfully treated with open adrenalectomy.

Keywords: Adrenal gland neoplasms, Adrenalectomy, Angiomyolipoma, Haemorrhage

1 Background

Angiomyolipomas are rare mesenchymal tumours arising from the perivascular epithelioid cells. They are benign endocrinologically inactive tumours with a histological structure consisting of variable amounts of adipose, thick-walled blood vessels and smooth muscle cells [1]. Commonly, angiomyolipomas occur in the kidney with few extra renal case reports in adrenals, bone, breast, colon, heart, liver, lung, parotid gland, retroperitoneum, skin and spermatic cord [2–5].

2 Case presentation

A 45-year-old lady, previously fit and well, presented with a 3-week history of right loin pain, nausea and vomiting. At presentation, her vital signs and abdominal examination were normal. A full blood count and urea and electrolytes were at baseline; C-reactive protein was elevated at 103 mg/L.

CT scan showed a right adrenal angiomyolipoma measuring 6.3 × 6.8 cm with intralesional haemorrhage (Fig. 1). Due to the large size, haemorrhage and symptoms, the patient underwent an open right adrenalectomy. She was discharged home just over 2 weeks post-operatively.

Histology showed a soft tissue tumour composed of mature adipose tissue, proliferating thick-walled vessels and in a smaller quantity proliferation of smooth muscle, with focal spindle and epithelioid cells (Figs. 2, 3). Immunostains for Melan-A (Fig. 4) and HMB-45 were focally positive. Desmin and smooth muscle actin were also focally positive, confirming the presence of smooth muscle in the tumour. EVG stain showed the blood vessels, with loss of elastic lamina, which is in keeping with vessels in angiomyolipoma (Fig. 5). These histological features are consistent with an adrenal angiomyolipoma [1].

The patient was reviewed in clinic 4 months post-operatively when she was asymptomatic, and a repeat CT scan showed no evidence of recurrence. At 36-month follow-up, she remained well.

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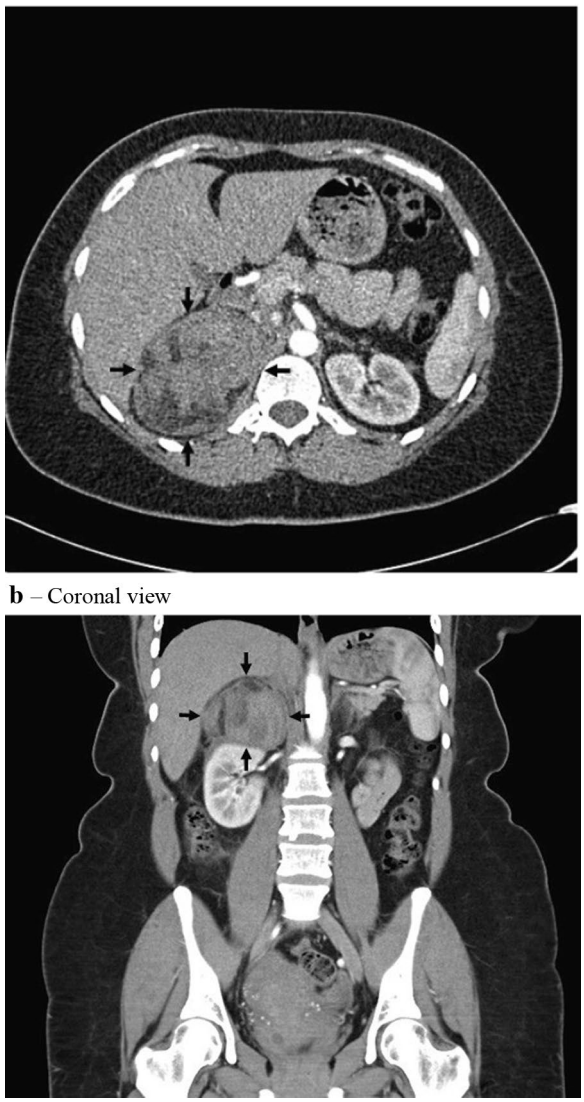

Fig. 1 CT on initial presentation shows haemorrhage of the right adrenal angiomyolipoma (arrowed)

3 Discussion

This case report describes spontaneous haemorrhage of an adrenal angiomyolipoma causing loin pain. Adrenal angiomyolipomas can also be picked up incidentally on imaging. The tumours are easily detectable from the pathognomonic feature of high-fat content. A review of the literature revealed 17 previously reported cases of adrenal angiomyolipoma. Of these, 14 were sporadic and 3 were reported in patients with tuberous sclerosis or lymphangioleiomyomatosis [2–17]. There was 1 reported case of spontaneous haemorrhage [4].

Angiomyolipomas can be difficult to differentiate histologically from sarcoma or sarcomatoid carcinoma.

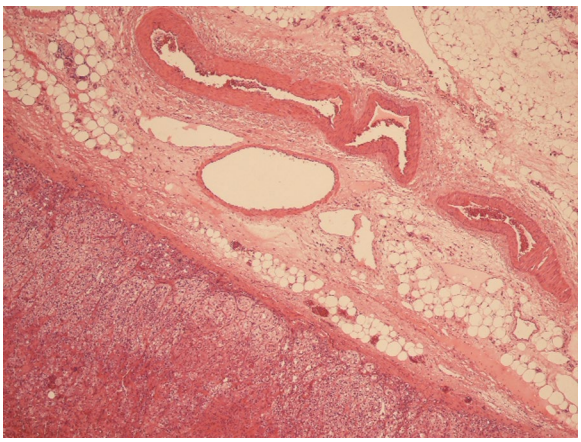

Fig. 2 Adrenal gland with tumour. Haematoxylin and eosin staining (× 4)

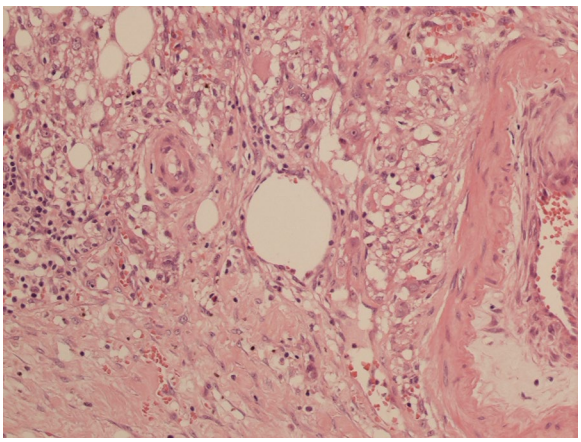

Fig. 3 Tumour with epithelioid cells, fat and blood vessels. Haematoxylin and eosin staining (× 20)

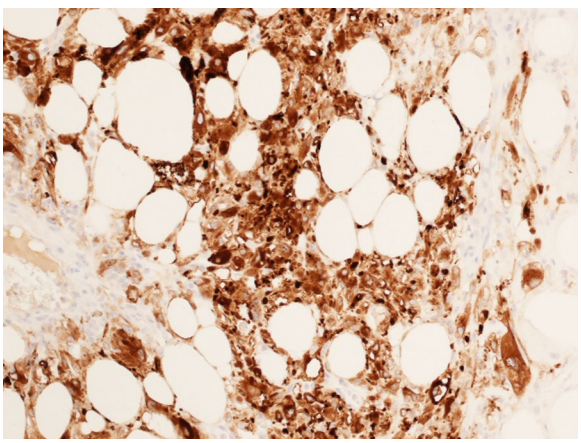

Fig. 4 Melan-A positive spindle and epithelioid cells (× 20)

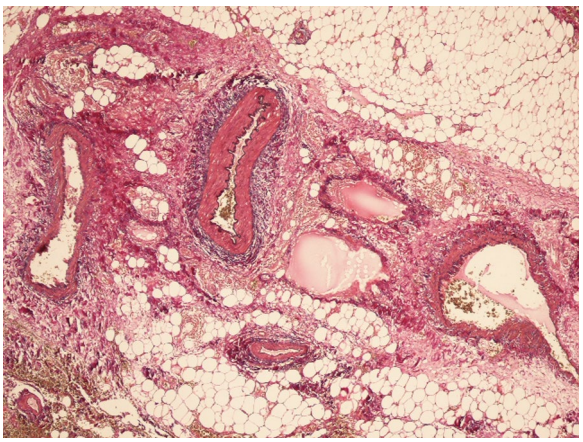

Fig. 5 Van Gieson's EVG stain with loss of elastic lamina in blood vessels (× 4)

Immunostaining can be used to help confirm the diagnosis. Markers that have shown a high specificity and sensitivity for angiomyolipomas include: HMB-45, Melan-A, SMA (smooth muscle actin) and microphthalmia transcription factor. S-100 is also often used in diagnosis although not all studies have found a high sensitivity for its use [1].

Management of these lesions has been guided by the limited number of case reports in the literature. It is general consensus that smaller lesions detected incidentally should be managed non-surgically and their size should be kept under surveillance. Surgical management has been proposed for lesions which are either symptomatic and/or greater than 5 cm. Laparoscopic adrenalectomy has successfully been used, but larger lesions are best removed using open adrenalectomy [13]. Adrenal angiomyolipomas have the potential to become very large with the largest reported case being 15 × 16 cm [5]. There have been no reports of malignant change in these lesions; nevertheless, long follow-up is recommended because of the unknown clinical progression of these tumours [1].

4 Conclusion

Adrenal angiomyolipomas are rare benign tumours that have the ability to reach a large size and potential to bleed. Here, we report the second case of spontaneous haemorrhage in an adrenal angiomyolipoma, which was successfully treated with open adrenalectomy.

Acknowledgements
Not applicable.

Authors' contributions
DW wrote the initial manuscript. IR redrafted the manuscript. AG provided the histological images and associated information. SS and SM provided critical review of the manuscript. All authors read and approvedthe final manuscript.

Funding
Not applicable.

Availability of data and materials
Not applicable.

Competing interests
The authors declare that they have no competing interests.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Ethics approval and consent to participate
Our institution does not require ethical approval for this case report.

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Received: 22 October 2019 Accepted: 29 October 2019
Published online: 27 November 2019

References

1. Esheba GES, Esheba NES (2013) Angiomyolipoma of the kidney: clinicopathological and immunohistochemical study. J Egypt Natl Canc Inst 25(3):125–134. https://doi.org/10.1016/j.jnci.2013.05.002
2. Godara R, Vashist MG, Singla SL et al (2007) Adrenal angiomyolipoma: a rare entity. Indian J Urol 23(3):319–320. https://doi.org/10.4103/0970-1591.33734
3. Goswami A, Sharma A, Khullar R, Soni V, Baijal M, Chowbey P (2014) Adrenal angiomyolipoma: a case report and review of literature. J Minim Access Surg 10(4):213–215. https://doi.org/10.4103/0972-9941.141531
4. Stolle H, Jonetz-Mentzel L, Krautschick-Wilkens AW (2006) Spontaneous rupture of an adrenal angiomyolipoma. Aktuelle Urol 37(6):443–444. https://doi.org/10.1055/s-2006-944287
5. Hu H, Xiaoqing X (2012) Giant Adrenal Angiomyolipoma. J Clin Endocrinol Metab 97(11):3835–3836. https://doi.org/10.1210/jc.2012-2319
6. Lam KY, Lo CY (2002) Adrenal lipomatous tumours: a 30 year clinicopathological experience at a single institution. J Clin Pathol 54:707–712
7. Elsayes KM, Narra VR, Lewis JS, Brown JJ (2005) Magnetic resonance imaging of adrenal angiomyolipoma. J Comput Assist Tomogr 29(1):80–82
8. Sutter R, Boehler A, Willmann J (2007) Adrenal angiomyolipoma in lymphangioleiomyomatosis. Eur Radiol 17:565–566. https://doi.org/10.1007/s00330-006-0206-5
9. Chee Kong CH, Mohmed Rose I, Singam P, Eng Hong G, Boon Cheok L, Zainuddin ZM (2010) Angiomyolipoma of the adrenal gland: a case report. Iran Red Crescent Med J. 12(4):489–491
10. Gupta P, Guleria S (2011) Adrenal angiomyolipoma: a case report and review of literature. Res J Med Sci 5(5):243–246. https://doi.org/10.3923/rjmsci.2011.243.246
11. Yener O, Özçelik A (2011) Angiomyolipoma of the right adrenal gland. ISRN Surg 2011, 102743. https://doi.org/10.5402/2011/102743
12. Monowarul I, Amanullah ATM, Alam AKMK (2012) Asymptomatic angiomyolipoma of the right adrenal gland. J Surg Sci 16(1):47–48. https://doi.org/10.3329/jss.v16i1.14449
13. Bhatti ABH, Dar FS, Pervez M (2013) Adrenal Angiomyolipoma. J Coll Physicians Surg Pak 23(9):663–664
14. Li W, Pang H, Cao Y, Guan L, Chen J (2015) High ^{18}F-fluorodeoxyglucose uptake in adrenal angiomyolipoma: case report and review of literature. Medicine (Baltimore) 94(22):e900. https://doi.org/10.1097/md.0000000000000900
15. Kwazneski D, Merrill M, Young J, Sell H (2016) Angiomyolipoma and malignant PEComa: discussion of two rare adrenal tumors. Case Rep Oncol Med 2016, 5204092. https://doi.org/10.1155/2016/5204092

16. Antar A, Boyle A, Patel T (2017) Angiomyolipoma of the adrenal gland: a case presentation and a review of adrenal lipomatous tumors. Urol Case Rep 12:59–61. https://doi.org/10.1016/j.eucr.2016.11.004
17. Ghimire O, Wenzhang L, Huaping L, Wenguang L, Yigang P, Jiale H (2017) Angiomyolipoma of the adrenal gland: a report of two cases and review of the literature. Am J Case Rep 18:989–994. https://doi.org/10.12659/ajcr.903908

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