Cystic Lymphangioma: A Rare Disease of the Adrenal Gland

Kistik lenfanjioma: Adrenal Bezin Nadir Bir Hastalığı

Nor Faezan Abdul Rashid[1], Rohaizak Muhammad[2], Suraya Othman[1], Shahrun Niza Abdullah Suhaimi[2], Norasyikin A. Wahab[3]

[1]Department of Surgery, University Technology Mara, Malaysia
[2]Department of Surgery, University Kebangsaan Malaysia Medical Centre, Malaysia
[3]department Of Medicine, University Kebangsaan Malaysia Medical Centre, Malaysia

ABSTRACT

Cystic adrenal neoplasms are uncommon and may be discovered incidentally or may be symptomatic but cystic lymphangioma is a rare benign tumour. The tumour usually occurs in the neck or axilla and rarely involves the adrenal gland. Here we report a case of a 42-year-old lady presented with symptoms of anorexia, vomiting and lethargy for 3 weeks. Apart from evidence of mild dehydration, there was no positive physical examination. Upon admission, her blood urea and creatinine was high and she was diagnosed with end stage renal disease secondary to chronic glomerulonephritis. Ultrasound of the abdomen revealed an incidental right adrenal mass and computed tomography (CT) scan showed a lesion with Hounsfield unit (HU) of 13, 20 and 21 on plain, venous and delayed phase, giving an absolute wash out of 12.5%. Biochemical tests were consistent with a non-functioning adrenal incidentaloma. Hence she was subjected for right retroperitoneoscopic adrenalectomy in view of suspicious of adrenal malignancy. However, histopathology reported as a rare adrenal cystic lymphangioma.

Key Words: Cystic adrenal lymphangioma, adrenalectomy, adrenal, cyst, lymphangioma

Received: 08.13.2017 **Accepted:** 11.18.2017

ÖZET

Kistik adrenal neoplaziler seyrektir ve tesadüfen bulunabilir veya semptomatik olabilirler, ancak kistik lenfanjiom nadir görülen benign bir tümördür. Tümör genellikle boyun veya aksillada görülür ve nadiren adrenal bezi tutar. Bu olguda, 3 haftadır anoreksi, kusma ve letarji belirtileri gösteren 42 yaşındaki kadın vakayı sunuyoruz. Hafif dehidratasyon dışında, herhangi bir pozitif fizik muayene bulgusu saptanmadı. Yatış sonrası, kan üre ve kreatinin değerleri yüksekti ve kronik glomerülonefrite sekonder son dönem böbrek hastalığı tanısı aldı. Karın ultrasonunda tesadüfen sağ adrenal kitle ve bilgisayarlı tomografide (CT) ise düz, venöz ve gecikmiş fazda Hounsfield ünitesi (HU) 13, 20 ve 21 olan bir lezyon gösterdi ve % 12.5'lik mutlak yıkanma sağladı. Biyokimyasal testler, fonksiyonel olmayan adrenal insidentaloma ile uyumlu idi. Bu nedenle, adrenal malignite şüphesi nedeniyle sağ retroperitoneoskopik adrenalektomi yapıldı. Bununla birlikte histopatoloji ender görülen adrenal kistik lenfanjiom olarak rapor edilmiştir.

Anahtar Sözcükler: Kistik adrenal lenfanjiom, adrenalektomi, adrenal, kist, lenfanjiyom

Geliş Tarihi: 13.08.2017 **Kabul Tarihi:** 18.11.2017

INTRODUCTION

Cystic lymphangioma is a rare tumour of the lymphatic vessels that occur more frequently in women(1). The location can be diverse and commonly occur in the neck or axilla(2). This tumour is less common in adult and most frequently occur in children and very rarely involves the abdomen(3). Evaluation of cystic adrenal lesion is important both in recognizing true adrenal cyst and in differentiating these lesions from malignant neoplasm of adrenal gland(1). Preoperative correct diagnosis depends on a good clinical discussion between the surgeon, endocrinologist and radiologist as well as experience in handling such case.

CASE REPORT

This is a 42-year-old lady with underlying bronchial asthma, presented in April 2016 with vomiting, anorexia and lethargy for 3 weeks. She also complained of unintentional weight loss about 5 kg in 3 months with reduced effort tolerance. She was later diagnosed with end stage renal disease secondary to undiagnosed chronic glomerulonephritis. Apart from mild dehydration, no abnormal physical examination was found. During her admission, several blood investigations and imaging were done including ultrasound of the abdomen where she was found to have a right adrenal mass.

Address for Correspondence / Yazışma Adresi: Nor Faezan Abdul Rashid, MD Department of Surgery, Sungai Buloh Campus, Universiti Technology Mara, UiTM, Jalan Hospital 47000 Sungai Buloh, Selangor, Malaysia E-mail: ezan_80@hotmail.com

doi:http://dx.doi.org/10.12996/gmj.2018.17

Computed tomography (CT) scan of the abdomen consistent with a right adrenal mass measuring 2.2 x 2.1 x 1.5 cm with fatty attenuation and Hounsfield unit of 13, 20 and 21 on plain, venous and delayed phase giving an absolute washout of 12.5%. Her serum renin, aldosterone and cortisol were within normal range. A repeat CT scan after 6 months showed an increase in the size of the mass by 1cm to become 3cm (Figure 1, 2) in size. She underwent endoscopic posterior retroperitoneum right adrenalectomy in view of suspicion of malignancy from her CT scan. Intraoperatively, there was a right adrenal complex cyst, which ruptured during manipulation, releasing a serous fluid. Minimal normal adrenal tissue was left behind. Post operatively, was uneventful and the histopathology report of the resected specimen was consistent with a cystic lymphangioma with no evidence of malignancy.

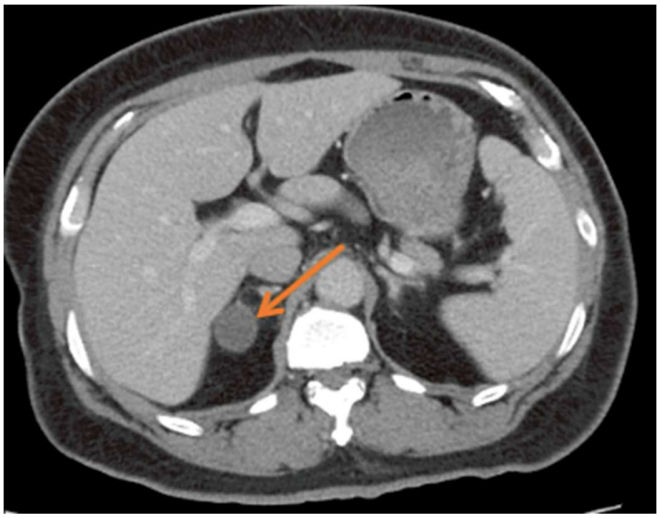

Figure 1: CT scan showed a 3cm right adrenal mass with homogenous appearance

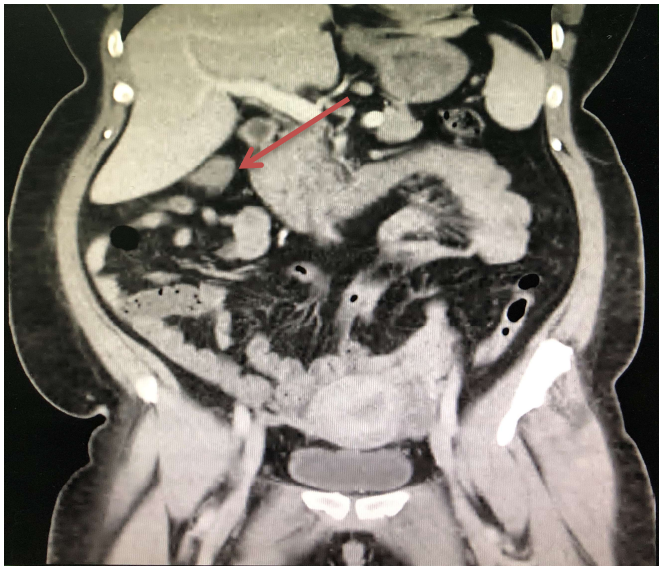

Figure 2: Arrow indicating right adrenal mass

DISCUSSION

Lymphangiomas are benign malformation of vessels, most frequently discovered during childhood(5). It is commonly found in the neck, axillary region and mediastinum (95%), with the remaining 5% are found in the abdomen cavity(8). An adrenal lymphangioma is a very rare benign vascular lesion that usually remains asymptomatic throughout life(4). It is nearly always asymptomatic and incidentally discovered during routine imaging work ups or investigations of other pathologies(9). It is usually unilateral, variable in size, smooth border with pure cystic internal structure(4). Adrenal cyst can be classified as endothelial cyst (45%), pseudocyst (39%), epithelial cyst (9%) and parasitic cyst (7%)(5). Within the endothelial cyst includes haemangioma, hamartoma and lymphangioma(5). Adrenal cystic lymphangioma can occur at any age with peak incidence between the third and sixth decades of life as in this case(4). Majority of the cases reported in literature showed that this tumour was found incidentally during abdominal imaging or abdominal surgery for other reason or at autopsy.

As for this case, the adrenal lesion was first detected during ultrasound of the abdomen looking for abnormality in her urinary system. She subsequently had CT scan of the abdomen, where the lesion at right adrenal gland was further delineated. Ming Zhao et al reported 3 cases of adrenal lymphangioma with average size of 3.2 cm were found during routine check up in two cases and one case in ureteral cancer detection. Most cases reported in literature describe the cyst is large and non-hormonal secreting. In our case, the cyst was around 3 cm in biggest diameter and non-hormonal secreting. Gaetan et al and Faten et al in their cases reported the cyst to be big and hypodense on CT scan and adrenal hormonal profile was not elevated. No specific laterality predilection reported in the literature, however Ming Zhao reported 3 cases of adrenal lymphangioma occurred on the left adrenal and so did Gaetan et al reported one case on the left adrenal. Ellis Cl et al on the other hand reported 9 cases of adrenal cystic lymphangioma where 3 cases occurred on the left adrenal and 6 cases occurred on the right adrenal. Most cases, clinical management of adrenal cyst depends on the findings of imaging studies. Several authors recommended managing the uncomplicated cyst with smooth wall lining and no thickening by aspiration instead of surgical excision(9). However, the shortcomings of this procedure including potentially unidentified histological type and high incidence of accumulation of cystic fluid(9). Surgical excision is recommended for cyst that is large, symptomatic, parasitic and considered likely to be malignant(10). In contrast to our case, patient was asymptomatic but the lesion grew in size by 1cm within 6 month and malignancy was suspected due to the high HU on a plain CT scan and delayed wash out on contrasted CT scan. Gaetan et al did performed aspiration of the cyst under image guided, however later proceeded with open excision as the cyst recurred and became symptomatic and technically difficult for a repeat aspiration.

CONCLUSION

Adrenal cystic lymphangioma is a benign lesion and mainly found incidental upon investigating for other disease. CT scan is a good modality in diagnosis and treatment mainly directed for symptomatic relief or suspicion of malignancy is high. Laparoscopic (minimal access) adrenalectomy is the best choice as it provides symptom relief and definite histopathology confirmation.

Conflict of interest

No conflict of interest was declared by the authors.

REFERENCES

1. Erickson LA, Lloyd RV, Hartman R, Thompson G. Cystic adrenal neoplasms. Cancer. 2004;101(7):1537-44.
2. Joliat GR, Melloul E, Djafarrian R, Schmidt S, Fontanella S, Yan P, Demartines N, Halkic N. Cystic lymphangioma of the adrenal gland: report of a case and review of the literature. World journal of surgical oncology. 2015;13:58.
3. Van Oudheusden TR, Nienhuijs SW, Demeyere TB, Luyer MD, de Hingh IH. Giant cystic lymphangioma originating from the lesser curvature of the stomach. World journal of gastrointestinal surgery. 2013;5:264.
4. Ellis CL, Banerjee P, Carney E et al. Adrenal lymphangioma: clinicopathologic and immunohistochemical characteristics of a rare lesion. Hum Pathol. 2011; 42: 1013-8
5. Ates LE, Kapran Y, Erbil Y et al. Cystic lymphangioma of the right adrenal gland. Pathol Oncol Res. 2005; 11: 242-4
6. Zhao M, Gu Q, Li C, Yu J, Qi H. Cystic lymphangioma of adrenal gland: a clinicopathological study of 3 cases and review of literature. International journal of clinical and experimental pathology. 2014;7:5051.
7. Faten Hadjkacem, Mouna Ammar, Ayman Maalej et al. Cystic lymphangioma of the adrenal gland: A case report and a review of the literature. American Journal of Medicine Studies. 2015; 3: 1-3
8. Poncelet V. Retroperitoneal cystic lymphangioma. Journal Belge Radiol. 1998. 81:245
9. Geramizadeh B, Yazdanpanah S, Salahi H, Marzban M. Adrenal Cystic Lymphangioma Presented With Hypertension: A Case Report. Nephro-urology monthly. 2015;7: :e27049.
10. Liu B, Li Y, Wang S. Adrenal lymphangioma removed by a retroperitoneoscopic procedure. Oncol Lett. 2013;5:539–40