Cystic Lymphangioma: A Rare Disease of the Adrenal Gland

Kistik lenfanjioma: Adrenal Bezin Nadir Bir Hastalığı

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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

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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.
**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 et al reported 3 cases of adrenal cystic lymphangioma occurred on the left adrenal and so did Gaetan et al reported one case on the left adrenal. Ellis CI 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**