

Pediatrics

A large epithelioid angiomyolipoma of the kidney in a very young child with idiopathic chylothorax: A diagnostic dilemma

Ameer Kakaje^{a,*}, Raghad Mujahed^a, Othman Hamdan^c, Jihad Hakim^b

^a Damascus University, Damascus, Syria

^b Department of Urology, Children's University Hospital, Damascus, Syria

^c Department of Haematology, Children's University Hospital, Damascus, Syria



ARTICLE INFO

Keywords:

Paediatric epithelioid angiomyolipoma
Tuberous sclerosis
Idiopathic chylothorax
Octreotide

ABSTRACT

Epithelioid angiomyolipoma (EAML) is a subtype of angiomyolipoma, the most common benign renal tumour and usually asymptomatic until adulthood.

This paper reports a patient with EAML identified at 30 months, the youngest age of presentation with this tumour reported in the literature. In this case the tumour was extremely large and was successfully surgically removed. Furthermore, the patient had no other TS features, presented with acute symptoms, and had bilateral idiopathic chylothorax which was managed by octreotide and diet.

Introduction

Perivascular epithelioid cell tumours (PEComas) are a large family of neoplasms which include angiomyolipoma (AML), lymphangiomyomatosis (LAM), amongst others and are distinctive by their perivascular epithelioid cells. Epithelioid angiomyolipoma (EAML) is a rare variant of AML tumours in the kidney with a mean age of diagnosis being 38 years.¹ Only a few cases of EAML have been diagnosed in children and the youngest reported in the literature was a four-year-old girl with a tuberous sclerosis (TS).² In this paper, we present a case of a 30 month old male with EAML and idiopathic chylothorax, the youngest case reported to our knowledge date in the literature.

Case report

The 30 month old male complained of a dry cough for ten days, shortness of breath and intermittent fever of 38.5°. He had no history of TS or any other medical condition and his psychomotor development, weight and height were normal. Decreased respiratory sounds were auscultated in both lung fields, with rales and crackles across the base of the left lung. A soft mass was palpable in the upper right abdominal quadrant. Chest x-ray (CXR) showed pleural effusion around the lungs, mostly in the right lung space. Chest computed tomography (CT) showed free pleural effusion, without lymph node enlargements or any abnormal alterations in the lung tissue (Fig. 1). Abdominal

ultrasonography detected a retroperitoneum, hyperechoic lesion. Abdominal CT disclosed a nodular mass which surrounded the aorta and vena cava. The mass was also anterior to the vertebrae, extending from the left kidney to the right kidney. Furthermore, the mass partially replaced some of left renal parenchyma and pushed the right kidney laterally (Fig. 1). Bone marrow aspiration and biopsy, brain MRI, echocardiogram, and ophthalmoscopy were normal. Thoracentesis yielded a milky, lipemic fluid and the analysis showed elevated triglycerides with a high lymphocyte count. The ratio of fluid to serum for cholesterol was less than 1 and more than 1 for triglyceride which is typical for chylous fluid. Cytological examination was negative for malignant cells and tuberculosis bacillus. A medium-chain triglycerides diet was indicated with intravenous octreotide and a repeated thoracentesis. As metastases were negative in the work-up, an open surgery approach was adopted. In surgery, a solid tumour was found arising from the right kidney hilum. It protruded the renal parenchyma, extended through the right renal capsule, the mesentery, ascending colon, aorta and inferior vena cava and it pushed the left kidney laterally without protruding its capsule. The mass was very large, approximately 9 × 6 × 6 cm in size. The tumour was successfully dissected from the great vessels as the mass did not invade their walls (Fig. 2). Following this, the patient underwent right radical nephrectomy as it was adjacent with the mass. The resection of the tumour required removing the affected mesentery, and ascending colon. Finally, an anastomosis between the ileum and transverse colon was conducted and consequently

* Corresponding author. Damascus University - Faculty of Medicine, Children's University Hospital, Syria.

E-mail address: ameer.kakaje@hotmail.com (A. Kakaje).

<https://doi.org/10.1016/j.eucr.2019.101035>

Received 8 September 2019; Received in revised form 26 September 2019; Accepted 3 October 2019

Available online 3 October 2019

2214-4420/© 2019 The Authors.

Published by Elsevier Inc.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

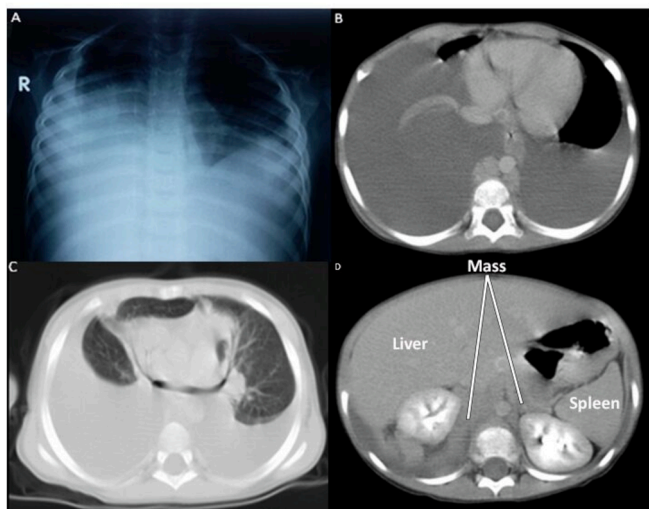


Fig. 1. A: a plain CXR that shows a bilateral pleural effusion, mostly on the right side. B + C: a thoracic CT-scan that shows free pleural effusion with normal lung parenchyma. D: an abdominal CT-scan that shows the mass with unclear extensions.

the entire tumour was successfully removed.

Pathological assessment demonstrated a mass arising from the right renal helium (Fig. 3), adjacent to the mesentery, and an ascending colon without signs of invasions. Histopathological findings detected giant mature polygonal epithelioid cells admixed with smooth muscle cells, scarce fat cells and vessels which is a picture typical of EAML. Although EAML is usually unpredictable in its behavior and can be considered malignant in some cases, it was considered benign in this case as there was no evidence of invasion or metastases. Octreotide was administered following diagnosis for four weeks until the chylothorax was no longer evident in the CXRs. This was followed for the next six months by weekly CXRs which remained normal. After six months, the patient had no dyspnea or a palpable mass. Furthermore, chest CXRs and abdominal and thoracic CT were all normal with no sign of relapse.

Discussion

AML is usually asymptomatic and discovered in TS follow-up, and when symptomatic pain usually being the primary reported symptom. . In CT scan, the paucity of adipose tissue renders EAML asresembling a renal cell carcinoma³ and distinguishable from classic AML.⁴ The patient reported in this paper however, did not meet the diagnostic criteria for TS⁵ and only developed EAML.

No genetic test was conducted. LAM, one of the major features of TS,⁵ was excluded in this case as the chylothorax was not in the typical form of LAM, the lungs parenchyma in the CT scan were normal and the patient did not have the typical LAM presentation as his age and gender

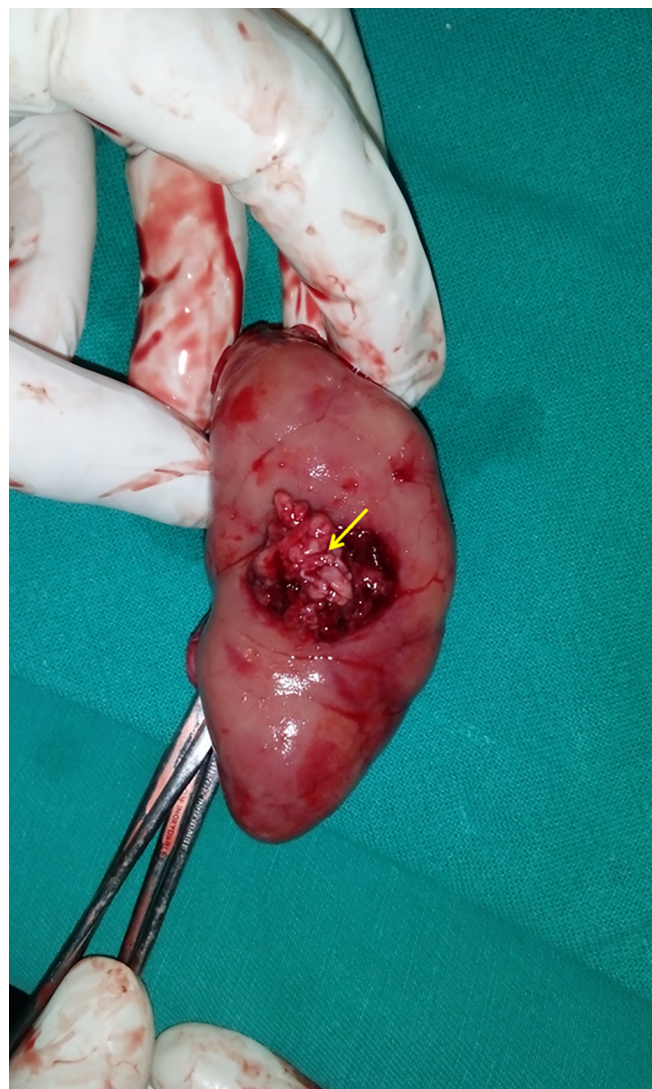


Fig. 3. The removed kidney with the dissected mass (arrow) originating from the kidney.

made the LAM diagnosis highly unlikely. The patient did not report any previous trauma or surgery prior to this surgical removal of this tumour, and as such no history could explain the chylothorax. Therefore, it was assessed as an idiopathic chylothorax and managed with a special diet and octreotide, and the chylothorax subsided in the following 4 weeks.

In conclusion, as EAML has a very strong correlation with TS, even stronger than classic AML, we could not assess with certainty whether the patient had TS, at least not at the present time as he may develop

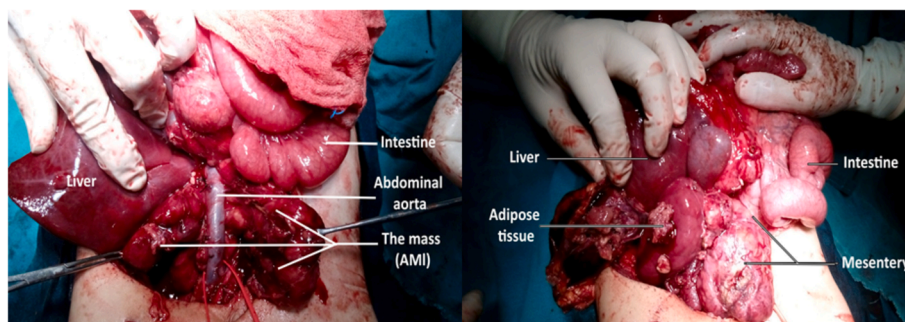


Fig. 2. Photos that show the abdomen after the mass being dissected from the great vessels.

more features in the future. However, the young age of presentation and the accompanying idiopathic chylothorax with octreotide treatment made this case highly unusual and to our knowledge the first of its kind reported in the literature.

Acknowledgements

We would like to thank Pro Lina Al-Khoury for her support and encouragement.

References

1. Hornick JL, Fletcher CD. PEComa: what do we know so far? *Histopathology*. 2006;48(1):75–82.
2. Gupta RK, Sharma MC, Chitragar S, Sinha K, Agarwala S, Sharma R. Epithelioid angiomyolipoma in a child—a diagnostic dilemma. *Urology*. 2014;83(6):1394–1397.
3. Bernardini S, Chabannes E, Algros MP, Billerey C, Bittard H. Variants of renal angiomyolipoma closely simulating renal cell carcinoma: difficulties in the histological diagnosis. *Urol Int*. 2002;69(1):78–81.
4. Esheba Gel S, Esheba Nel S. Angiomyolipoma of the kidney: clinicopathological and immunohistochemical study. *J Egypt Natl Cancer Inst*. 2013;25(3):125–134.
5. Northrup H, Krueger DA. International tuberous sclerosis complex consensus G. Tuberous sclerosis complex diagnostic criteria update: recommendations of the 2012 international tuberous sclerosis complex consensus conference. *Pediatr Neurol*. 2013;49(4):243–254.