Journal of Advances in Medicine and Medical Research

33(17): 88-93, 2021; Article no.JAMMR.71744
ISSN: 2456-8899
(Past name: British Journal of Medicine and Medical Research, Past ISSN: 2231-0614, NLM ID: 101570965)

Case Report of a Giant Adrenal Myelolipoma in a Patient with Sickle Cell Anaemia

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Authors' contributions

This work was carried out in collaboration among all authors. Author UPI Patient management, conception and supervision. Author CAU Data collection and literature review, Author JUU Data collection and radiological imaging. Author EN Scholarly review. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JAMMR/2021/v33i1731032
Editor(s):
(1) Dr.Chan-Min Liu, Xuzhou Normal University, PR China.
Reviewers:
(1) Abel Decmann, Semmelweis University, Hungary.
(2) Giou-Teng, Yiang, Tzu Chi University, Taiwan.
Complete Peer review History: https://www.sdiarticle4.com/review-history/71744

Case Study

Received 23 May 2021
Accepted 29 July 2021
Published 02 August 2021

ABSTRACT

Adrenal myelolipomas (AMLs) are rare benign adrenal tumours containing adipose and hematopoietic tissue as a result of reticuloendothelial cell metaplasia. In this case report, we describe the diagnostic evaluation and the operative management of a giant adrenal myelolipoma in a 14-year-old male who has sickle cell anaemia. He presented with a one-week history of bilateral leg swelling. A physical examination revealed a mass in the right lumbar region. Ultrasound of the abdomen revealed a well-defined rounded echo-complex encapsulated supra-renal mass impinging on the upper pole of the right kidney, displacing it downwards. There was also marked compression of the inferior vena cava. A computed tomography scan showed a large mass occupying the right adrenal gland. The patient had an exploratory laparotomy with excision of the right adrenal tumour. Histopathological (Immunohistochemistry) evaluation of the mass confirmed the diagnosis of adrenal myelolipoma. The diagnosis of an adrenal myelolipoma requires a good history, physical examination, a high index of suspicion, imaging techniques, and a thorough histopathological evaluation. Surgery is the main treatment modality and good post-operative management minimizes complications and guarantees rapid recovery.

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Keywords: Adrenal myelolipoma; case report; sickle cell anaemia; surgery.

1. INTRODUCTION

An adrenal myelolipoma is a rare, non-functioning, benign tumour. It was first described by Gierke in 1905 and named by Oberling in 1929 [1]. It is a benign adrenal neoplasm predominantly composed of mature adipose tissue and intermixed myeloid tissue. Adrenal myelolipomas comprise 6-16% of adrenal incidentalomas and are the second most common cause after adrenal adenoma [1,2]. The increased prevalence of adrenal myelolipomas is due to increased detection because of the wider accessibility of computed tomography (CT) and magnetic resonance imaging (MRI) scans [2].

The pathogenesis of adrenal myelolipomas is either believed to be due to metaplastic changes in the mesenchymal cells or as a result of overstimulation by adrenocorticotrophic hormone (ACTH) [2]. Clinically, they may be asymptomatic or present with abdominal pain, nausea, or vomiting. Surgical resection is indicated in cases of significant growth or hormonal hyper secretion [2].

2. CASE PRESENTATION

A 14-year old male presented to our facility with a one-week history of bilateral leg swelling. The swelling was said to have started on the right and gradually progressed to involve the left leg. The patient is a known sickle cell anaemia patient who was diagnosed at the age of 3 months. He was, however, not consistent with follow-up visits, as his last visit was 2 years prior to presentation. He had surgery for hypospadias 10 years earlier.

A physical examination revealed bilateral pitting leg oedema up to the thighs. There was abdominal distension with an abdominal girth of 75 centimetres (cm) 10 cm from the xiphisternum. There was an undefined mass on the right flank, which was difficult to characterize. The liver was 6cm palpable below the right costal margin. His blood pressure was normal and a urinalysis showed mild to moderate proteinuria.

Liver function tests, serum urea and creatinine were within normal limits. A full blood count done on presentation gave a haemoglobin of 4.8g/dl and a total white blood cell count of 15,300/mm3. An abdominal ultrasound scan revealed a well-defined rounded echo-complex clearly encapsulated supra-renal mass, impinging on the upper pole of the right kidney, displacing it downwards. The mass measured about 12.55cm in diameter. There was also marked compression of the inferior vena cava (IVC), just close to the intra-hepatic portion. The spleen measured about 9.5cm in length, with a coarse echo texture, harbouring multiple irregular foci of calcification. A CT scan revealed a large mass occupying the right adrenal gland, measuring 96 x 97 mm. (See Figs. 1,2 and 3).

A Paediatric surgical consultation was requested, and the patient was booked for an exploratory laparotomy. He received blood transfusions both pre-and intra-operatively. Intra-operative findings were a right, well-encapsulated adrenal tumour, measuring about 16 x 16 cm, impinging on the IVC and the right kidney. The post-operative condition was satisfactory, and the patient recovered gradually, with remarkable regression of the leg oedema.

Histopathological analysis of the specimen revealed an 11 x13 x 6cm mass, with haemorrhagic areas. The immunohistochemistry revealed a tumour composed of few mature fat cells, and sheets of haematopoietic cells, showing normal trilineage haematopoiesis and markedly increased megakaryocytes. A rim of normal adrenal gland was seen. The overall features were those of an adrenal myelolipoma (See Fig. 4).

Subsequent follow up visits still had the patient presenting with leg oedema, albeit less severe than it was before the surgery. A Paediatric nephrology consult was requested, and serum lipid profile, albumin, total protein, and serum electrolytes were all within normal limits, even though the urine still showed mild proteinuria. Another abdominal ultrasound scan revealed splenomegaly and cholelithiasis. He was then referred to a paediatric gastroenterologist and a paediatric nephrologist for further evaluation and management, while continuing his paediatric haemato-oncology follow-up visits.

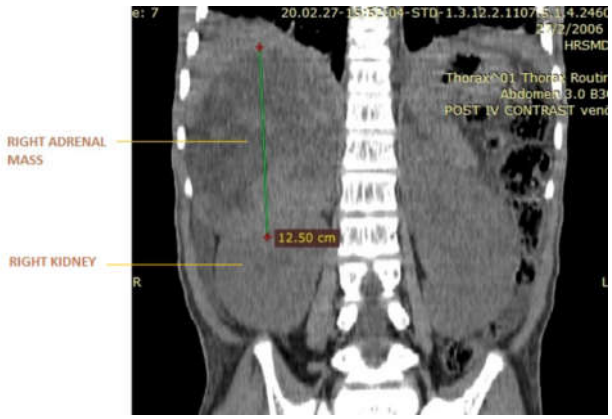

Fig. 1. Coronal view of abdominal CT

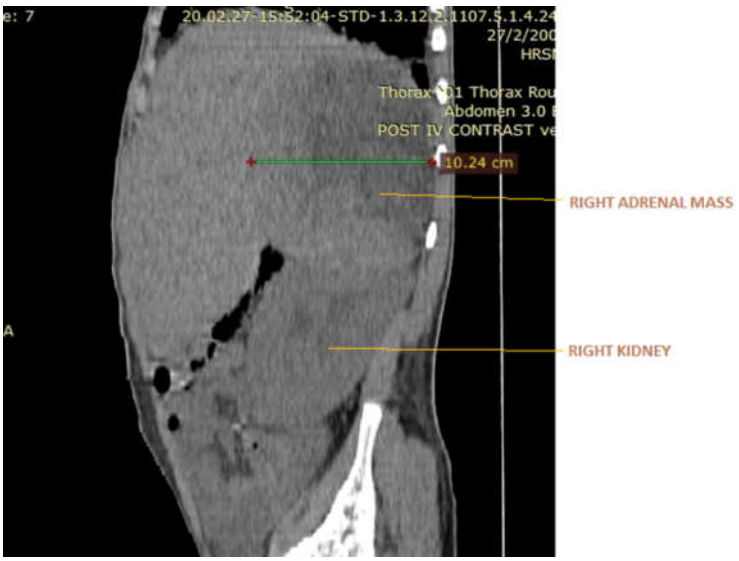

Fig. 2. Sagittal view of the abdominal CT scan

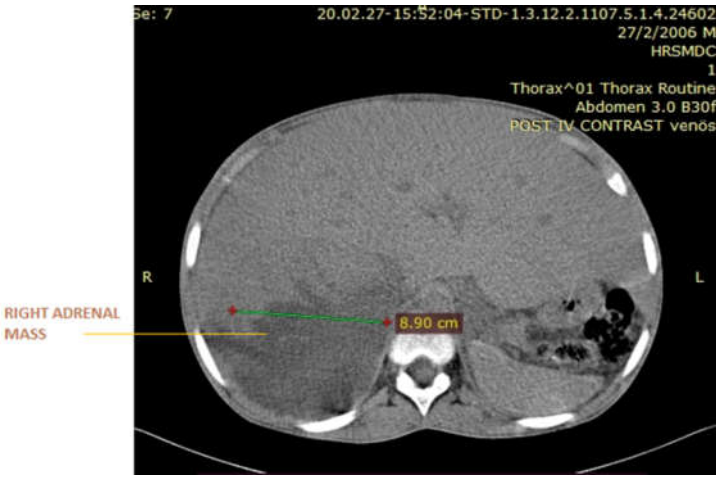

Fig. 3. Axial view of the abdominal CT scan

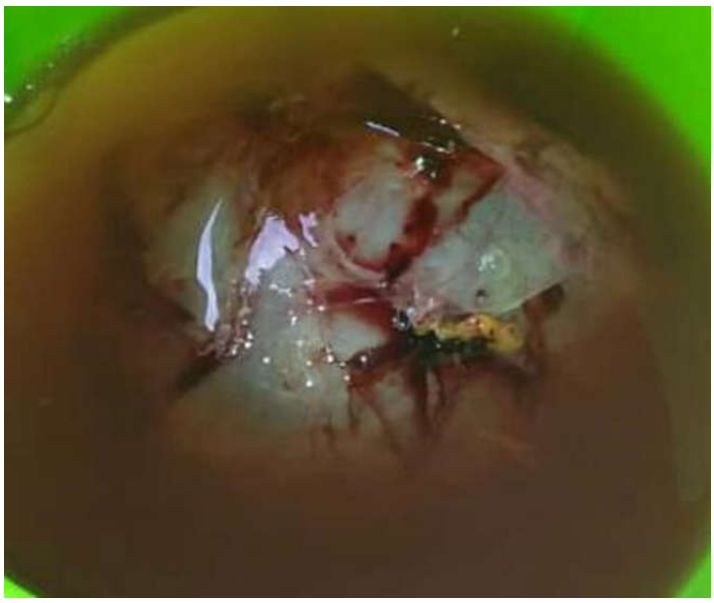

Fig. 4. Gross appearance of the tumour

3. DISCUSSION

Adrenal myelolipoma is a rare benign adrenal neoplasm [2]. The etiopathogenesis of adrenal myelolipomas is not definitively known. One hypothesis, which is the most widely accepted, suggests that stimuli, such as necrosis or inflammation, could lead to the metaplasia of the reticuloendothelial cells, which in turn could lead to the development of adrenal myelolipomas [3]. Another hypothesis suggests that adipocytes develop from the mesenchymal stem cells in the endothelium. This results in inflammation leading the adrenal cortex to secrete mediators responsible for the recruitment of hematopoietic progenitors [3]. Adrenal myelolipoma is often associated with conditions like Cushing's disease, obesity, hyperlipidemia, hypertension, and diabetes, which can be considered adrenal stimulants [4]. Adrenal myelolipomas are usually recognized in adulthood, with a median age of diagnosis of around 51 years, and there is no gender predilection [5].

Many cases of adrenal myelolipoma have been reported in sickle cell disease (SCD) and thalassemia, but the pathogenesis is not clear [6,7]. However, it is postulated that the persistent anaemia in SCD may be responsible for the stimulation of metaplasia in the haemopoietic elements sited in the adrenal stroma, leading to rapid tumour growth [7,8]. This may explain why our patient developed the tumour in childhood, when the median age at diagnosis is usually in adulthood.

Mass-forming extramedullary haematopoiesis (EMH) is another type of fat-containing mass that can occur in patients with haematologic disorders [8]. EMH is a physiological response secondary to insufficient blood cell production and bone marrow function and usually occurs in the reticulo-endothelial system outside the bone marrow. However it rarely occurs in the adrenals [8,9].

The size of adrenal myelolipomas is variable and can range from a few millimetres to greater than 10 cm, where they are termed giant adrenal myelolipomas [2]. They are usually unilateral, but they can also involve both adrenal glands [2] In the case of bilateral neoplasms, left-sided masses are usually larger than the right. It has been speculated that the asymmetric growth of these myelolipomas is due to the space limiting constraints of the liver on the right side [10].

The most common presenting symptoms of adrenal myelolipomas are as follows: Abdominal pain-22.5%, hypochondrial pain-13.9%, flank pain-13.9%, abdominal mass-5.2%. [2] Rarely, dyspnoea, back pain, fever, weight loss, and virilization can be the presenting symptoms of adrenal myelolipomas [2].

On histopathological examination, myelolipomas are predominantly composed of fatty areas with

interspersed hematopoietic tissue components. These fatty elements and hematopoietic areas may be clearly separated, or they are often intermixed [11]. Tissue analysis often reveals a variable amalgamation of myeloid and erythroid cells, megakaryocytes, and occasionally lymphocytes [11]. In an isolated adrenal myelolipoma, a peripheral rim of normal adrenal cortical tissue can be commonly identified distinctly from the mass, which was the case in our patient.

Adrenal myelolipomas can be diagnosed in 90% of cases by ultrasonography, CT, and magnetic resonance imaging, with CT scan being the most sensitive for identification of fat within the lesions [3,12]. CT is the preferred imaging modality for the diagnosis of adrenal myelolipoma [2]. It is also the most sensitive for detecting haemorrhage, which may be hyper to hypodense depending on the age of evolution [2,8]. On MRI, the predominantly fatty areas in myelolipoma appear hyperintense on T1 MRI images and intermediate to hyperintense on T2 [2]. The differential diagnoses include renal angiomyolipoma, benign adrenal adenoma, adrenal carcinoma, retroperitoneal liposarcoma, and lipoma [2].

Management of adrenal myelolipoma should be decided upon based on the size of the lesion and the presence of symptoms. Small lesions measuring less than 5cm and those that are asymptomatic are usually monitored via imaging over a period of one to two years [13]. According to various studies, it is suggested that symptomatic tumours or myelolipomas larger than 7 cm, as was the case in our patient, should undergo elective surgical excision [2,14]. The approach is based on the reported incidence of life-threatening emergencies caused by spontaneous rupture and haemorrhage within large lesions [2,14]. Conventional or endoscopic access may be chosen depending on the size of the tumour [15]. In conventional surgeries, the extra peritoneal approach is preferred to a midline incision because of the quicker recovery of the patient and the smaller postoperative complication rate [15]. Mini-invasive and endoscopic techniques are best utilized for smaller-sized tumours, depending on the expertise of the operator [11,12,16].

4. CONCLUSION

Adrenal myelolipomas are rare tumours which are usually benign. A high index of suspicion is required to make an early diagnosis. Late presentation and diagnosis may result in obstructive symptoms and compression of blood vessels, which was the case in our patient. Imaging techniques and histopathological evaluation are the mainstays of diagnosis. Surgery is the treatment modality of choice in cases where the tumours are large enough for resection.

CONSENT

The patient's parents were informed about the peculiarity of the case and the willingness of the authors to publish it as a case report. They understood and gave written informed consent for their history, radiographic images and specimen photos to be used for the case report.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle4.com/review-history/71744