

Adrenal myelolipoma in a young male — a rare case scenerio

Muhammad Anis-ul-Islam, Abdul Hafeez Qureshi, Syed Zafar Zaidi

Abstract

Adrenal myelolipomas are rare non functional benign tumours which are incidentally diagnosed in asymptomatic patients. Despite the fact that several theories have been discussed, the origin of myelolipoma remains unclear. Our patient, 19 years old male presented with history of abdominal pain for past 7 years. Computed tomography scan of abdomen with oral and intravenous contrast revealed well defined round heterogenous enhancing mass seen in right supra renal area involving the adrenal gland. Patient underwent surgery by open access through thoracoabdominal approach. Histopathology of resected mass showed myelolipoma along with trilineage haematopoiesis with mature adipose tissues.

Keywords: Adrenal myelolipomas, Tumours.

Introduction

Adrenal myelolipomas are benign tumours with the reported incidence of 2% to 4% of all adrenal tumours. Myelolipoma is an unusual benign tumour, composed of mature fat tissue and haematopoietic elements including myeloid and erythroid cells.¹ Edgar von Gierke, a German pathologist was the first to notice such a lesion in the adrenal gland and described it in 1905, but the lesion was given its name, "myelolipoma", in 1929 by the French pathologist Charles Oberling.¹ Myelolipomas are generally adrenal lesions and extra-adrenal myelolipomas (EAMLs) are a very rare entity. Currently these tumours are incidentally discovered at ultrasound or computed tomography done for other purposes.² Adrenal myelolipomas are rare non functional benign tumours which are incidentally diagnosed in asymptomatic patients in the 4th and 5th decades. It has a prevalence of approximately 0.08-0.4% on autopsy findings. Myelolipomas are usually unilateral, and very rarely bilateral.³

Case Presentation

A 19-year-old male initially presented to Medicine

Department of Urology, the Indus Hospital, Karachi.

Correspondence: Muhammad Anis-ul-Islam.

Email: mislam_2011@hotmail.com

Outpatient Department in Nov 2013 with history of abdominal pain for past 7 years. He had generalized vague abdominal pain, which felt like heaviness, mild to moderate in intensity. Patient also had complaints of joint pain, backache and chest pain, but no history of dyspnea, nausea or vomiting. He had lost weight for past couple of months but was undocumented. He had no known medical illness.

On Examination, patient had blood pressure of 100/70mmHg. He was anaemic but no visceromegaly was appreciated. His HB before procedure was 11.0 mg/dl, anaemia was normocytic normochromic. Lab biochemistry showed LDH of 1182 IU/L (n=95-195). Patient had deranged liver function tests at presentation with elevated indirect bilirubin levels, also LDH levels remained persistently high post procedure as well, and so patient was referred to gastroenterologist with impression of Gilbert's syndrome. His 24-hour urinary

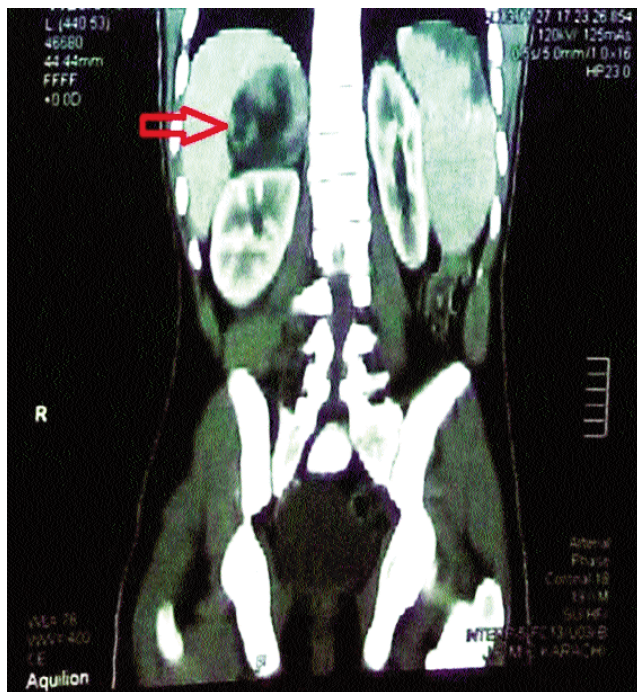


Figure-1: Computed tomography scan of abdomen with intravenous contrast revealing well defined round heterogenous enhancing mass seen in right supra renal area involving the adrenal gland.

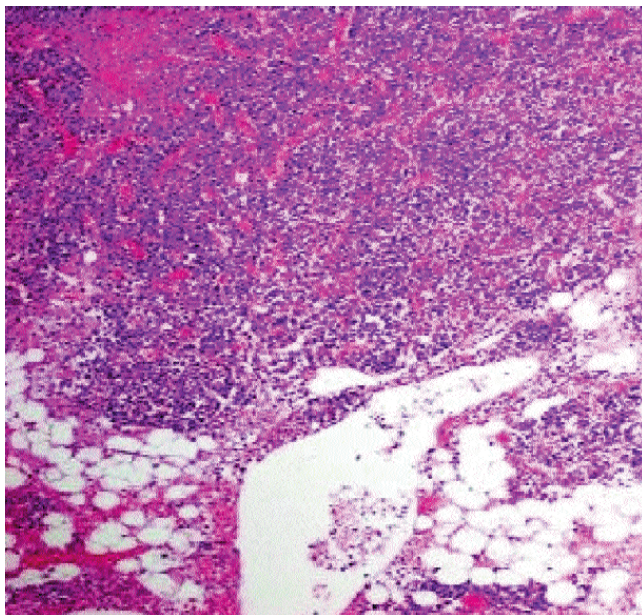


Figure-2: Microscopic appearance of adrenal myelolipoma. Typical histological features of myelolipoma comprising varying proportions of mature adipose tissue along with trilineage haematopoiesis.

vanillylmandelic acid level was 7.3 mg (reference range 1-11 mg/24 hour) to rule out adrenal malignancy as a CT was previously done without adrenal protocol.

Radiology work up was done and ultrasound whole abdomen showed large solid predominant right adrenal mass of around 10 x 6 cm. Patient had brought his computed tomography scan of abdomen and pelvis with oral and intravenous contrast, already done from outside revealing well defined round heterogenous enhancing mass seen in right supra renal area involving the adrenal gland. Approximately 3.5 x 3.1 cm hypodense area was also seen within it, representing necrosis and or haemorrhage, along with some calcifications (Figure-1). Other viscera were unremarkable. No lymphadenopathy was noted.

Based on work up, surgery was planned and preoperative preparations done. Patient underwent surgery by open access through thoracoabdominal approach in Jan 2014. Per operatively we found encapsulated right supra renal mass with smooth surfaces having solid and cystic components, adjacent to liver bed superiorly and inferiorly. It was in close proximity to right kidney. Also another two swellings with single pedicle seen arising from it. So complete resection of tumour was done and sent for histopathological analysis. Histopathology of resected mass showed myelolipoma along with trilineage haematopoiesis with mature adipose tissues (Figure-2). Post operatively he remained well with good recovery and

had followed us regularly with no active issues.

Discussion

Adrenal myelolipomas are benign tumours with a reported incidence of 2% to 4% of all adrenal tumours.¹ The histological basis of these mesenchymal tumours is still modest. Lipomas are known to occur on the right side with male predominance as opposed to myelolipomas, which have no gender or site predilection. Adrenal myelolipomas vary in size from several millimeters to more than 30 cm, and usually in the range of 2-10 cm in diameter.¹

Despite the fact that several theories have been discussed, the origin of myelolipoma remains unclear. Such theories include remnants of foetal bone marrow, embolism of bone marrow cells, and hyperplasia of heterotopic reticulum cells. Chang et al. described a case of adrenal myelolipoma with a translocation $t(3; 21)(q25; p11)$. A similar change, $t(3; 21)(q26; p11)$, is found in haematopoietic neoplasms, such as myelodysplastic syndromes and chronic myeloid leukaemia. This finding reflects that myelolipoma is derived from misplaced haematopoietic cells.³ A study performed by Bishop et al. revealed myelolipomas and displayed X-chromosome inactivation in both fat and haematopoietic elements, which suggests clonal origin of myelolipoma.⁴

Myelolipomas are usually asymptomatic and discovered incidentally on routine radiological examination. About 10% become large and can cause vague symptoms, such as pain.³ The most significant complication that could occur with large myelolipomas is acute haemorrhage, which can present with pain that localizes in the back or flanks, with nausea, vomiting, hypotension and anaemia.⁵ Even though myelolipomas are hormonally inactive, but about 10% cases are associated with endocrinopathies, such as Cushing's syndrome, congenital adrenal hyperplasia, Conn's syndrome, pheochromocytoma, hyperparathyroidism or adrenogenital syndromes.⁷

Massive accelerated growth of these neoplasms can result in symptoms such as flank pain and abdominal discomfort due to rupture, haemorrhage, or necrosis. More severe symptoms include haematuria, renovascular hypertension, and even surgical emergencies such as retroperitoneal haemorrhage.⁷ Therefore, it is critical to realize the significance of incidental findings and management should be targeted towards preventing such severe consequences.

Most myelolipomas are well circumscribed and true capsule is not seen, rather, compressed connective tissue surrounding the lesions represents a pseudo capsule.⁵

Haemorrhage is more common in larger lesions of more than 10 cm in diameter. Calcification may also be found. Microscopically, myelolipomas contain mature fat cells and megakaryocytes.¹ They are differentiated from true bone marrow in that they contain no reticular sinusoids or bone spicules.⁵

Recommendations propose that asymptomatic myelolipomas of less than 4 cm should be monitored expectantly with annual abdominal CT scans. Since the risk of spontaneous rupture or bleeding is minimal in these small myelolipomas, observation can prevent lifelong steroid replacement.⁵ Surgery is mandated if the tumour exceeds 7 cm because of increased risk of spontaneous rupture and retroperitoneal haemorrhage.⁸ Some surgeons recommend resection when tumour size is greater than 5-6 cm since potentially malignant tumours of the adrenal gland also manifest as nonfunctional tumours.⁹ Others recommend surgical removal of myelolipomas greater than 4 cm because of the rare chance of rupture and other associated complications.¹⁰

Junaid et al had performed right adrenalectomy on a 63 years old male through right subcostal incision taking extraperitoneal approach, who presented with upper abdominal pain and adrenal mass of 6.1 x 4.0 cm on CT scan.⁸ Brogna et al had also done surgery on a 52 years old man who presented with hypertension and had a 20 cm mass of left adrenal gland on ultrasound scan done for other reasons. Surgical left adrenalectomy was performed and patient remained normotensive after surgery.⁹

Conclusion

Giant adrenal lipomas are rare but now are being reported more frequently because of improved modern imaging technologies.

Our case had a relatively younger age at presentation and was associated with deranged liver function tests, again an infrequent accompaniment. Complete resection of the tumour reversed all the symptoms.

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