Laparoscopic excision of a giant adrenal myelolipoma and review of the literature

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Abstract

Introduction: Adrenal myelolipoma is a rare benign tumor. It is usually asymptomatic with variable sizes, where the small lesions are usually managed conservatively and the large and symptomatic ones indicate open or laparoscopic adrenalectomy.

Observation: A 45-year-old obese male patient presented with an accidentally-discovered right adrenal mass during abdominal sonographic examination. The mass was clinically-palpable in the right lumbar region. Abdominal computed tomography described a well-circumscribed lesion displacing the right kidney downwards with compression and displacement of the inferior vena cava. It was heterogeneous with 16 cm × 14 cm × 8 cm dimensions and low attenuation appearance. Adrenal myelolipoma was suspected and the patient was counseled for the laparoscopic approach with high possibilities of conversion to open surgery. Laparoscopic excision was done by expert surgeons with demanding dissection from the surroundings, especially the liver and inferior vena cava, but, the mass was successfully removed. The postoperative course was short and uneventful. Histopathological examination confirmed the diagnosis of adrenal myelolipoma.

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Introduction

Adrenal myelolipoma is a rare tumor with a progressively increasing reporting rate. Its incidence progressed from 0.08–0.4% to 10–15% through the last decade. Its pathological composition comprises adipose and myeloid tissues [1]. Although it may reach huge sizes, adrenal myelolipoma is usually a small-sized asymptomatic lesion [2]. Treatment approach usually considers the lesion size, where masses of <4 cm are recommended for conservation, while the larger ones are treated by surgical interventions [1]. Owing to the world-wide progressively growing laparoscopic skills, many giant adrenal myelolipomas have been excised laparoscopically [3,4]. Our aim from the following case presentation is the verification of the feasibility of laparoscopic approach for the giant adrenal myelolipomas in obese patients.

Case report

A 45-year-old obese male patient presented to us with an accidentally-discovered huge right adrenal mass. He consulted many urological centers for treatment in different countries before the presentation to our center. Physical examination revealed a body mass index of 34.64 kg/m². There was a non-tender palpable right-sided abdominal mass. Otherwise, no clinical findings were detected.

Abdominal ultrasound described a right adrenal hyperechoic lesion. Computed tomography described a large right adrenal mass markedly-displacing and rotating the right kidney with 16 cm × 14 cm × 8 cm dimensions and low attenuation heterogeneously hypodense appearance suggesting myelolipoma. The mass was well-defined with a clear demarcation from the compressed surroundings. Also, the inferior vena cava was displaced and stretched (Fig. 1A and B).

Laboratory work up of the adrenal tumor markers; metanephrines, serum cortisol, Vanillylmandelic Acid, and other routine and surgical fitness tests were unremarkable. The patient was counseled for the treatment options and associated risks and complications with a specific stress on the laparoscopic approach and possibilities of conversion to open surgery.

The patient had transperitoneal laparoscopic adrenalectomy. He was positioned in the left flank position with 45 tilt. Five laparoscopic ports were designated; the main port was 10-mm and placed as just supra-umbilical. A second 10-mm port created in the midline high in the epigastrium. Other two 10-mm ports were created in the mid-clavicular and lateral axillary lines two inches below the costal margin. Also, a 5-mm port was created in the mid-axillary line at a lower level than the previous ports. After pneumoperitoneum creation, instrumental adjustments, mobilization of the colon and retraction of the liver, dissection of the mass was carried out from the inferior vena cava with ligation of the right adrenal vein. Significant technical efforts were indicated for dissection of the mass extensions around the cava and the sub-hepatic plain (Fig. 2A–D). However, the operative course progressed without complications through a total time of about 3 h and 45 min. Blood loss was 180 ml with no blood transfusion.

Histopathological examination revealed benign mature adipose tissues and myeloid elements which confirmed the diagnosis of adrenal myelolipoma (Fig. 3A and B).

Discussion

Adrenal myelolipoma is a benign tumor composed of adipose and myeloid elements [5,6]. It has been commonly reported that adrenal myelolipoma has small sizes less than 4 cm [1]. However, with increased reporting rate, the sizes of the reported cases have been remarkably increased [4,6] (Supplementary Table 1). A recently published review article studied 440 histologically-verified clinical adrenal myelolipomas which were reported between 1957 and 2017 with an average size of 10.4 cm [7]. Adrenal myelolipoma is commonly a unilateral tumor with predominance of the right side [5]. It could be associated with other organs’ tumors [8] (Supplementary Table 1). Rao et al. [9] differentiated adrenal myelolipomas into 4 distinct clinico-pathological patterns; isolated adrenal myelolipomas, adrenal myelolipomas with acute hemorrhage, extraadrenal myelolipomas, and myelolipomas with other adrenal diseases. Giant adrenal myelolipomas may follow this classification. Accordingly, the current case belonged to the first pattern which is the simplest one. They are prone to complications, especially the spontaneous rupture and life-threatening hemorrhage [1,7] representing the second pattern in the above mentioned classification.

Extraadrenal manifestations of myelolipoma result from two different issues; the first issue is the extraadrenal origin of myelolipoma which is an extremely rare entity with multiple predilection sites principally including thorax and retroperitoneum. The second issue is the very rare states in which adrenal myelolipoma develops secretory activities leading to hypertension or manifestations of hypercortisolism [1,7,10]. This pattern may result from an isolated functioning lesion or due to another co-existing adrenal lesion to be up graded to the fourth pattern.

Adrenal myelolipoma is commonly reported as a hormonally-inactive tumor. However, there are many reported associations with functioning adrenal disorders such as congenital adrenal...
hyperplasia and Cushing’s and Conn’s syndromes. The commonest postulation of etiology of adrenal myelolipoma is metaplasia of the adrenal cortex due to chronic stressors [1,7,10]. However, these clinico-pathological associations with endocrine dysfunctions support the postulation of hormonal overstimulation pathways in etiology of myelolipomas [8]. Congenital adrenal hyperplasia due to 21-hydroxilase deficiency has been reported in association with giant adrenal myelolipomas more frequently than other variants and other congenital disorders or syndromes. Prolonged exposure of high levels of adrenocorticotropic hormone has been proposed as the underlying etiology in these cases [7,11]. Also, adrenal myelolipomas have been reported in association with multiple chronic congenital hematological disorders such as thalassemia and sickle cell anemia suggesting hematopoietic etiological stimuli and supporting the hormonal pathways [7]. Rarely, hypertension is being a presenting finding for a functioning adrenal myelolipoma. In spite of these controversies, still adrenal myelolipoma is having a big body of evidence in the literature that it is a non-functioning tumor [1,7,10].

Imaging of adrenal myelolipoma is an interesting subject in discussions of the majority of the reported cases, because the lesion is usually asymptomatic and incidentally-discovered tumor. It has characteristic findings in the modern imaging modalities that provide very low indices of false negative results [3,11]. Our case had a large size and it was discovered accidentally on abdominal ultrasonography. Also, computed tomography was very valuable, where it described the commonly reported characteristic findings such as the well-demarcated and heterogeneous low-attenuated adrenal mass compressing the surroundings [1,6]. It demonstrates variably low density values of Hounsfield Unit down to –200 due to high fat contents [12]. Other imaging modalities have been used for characterization of adrenal myelolipoma with high suspicion of malignancy including combination of computed tomography with positron emission tomography (PET-CT) scan to characterize the marrow elements and malignancy. Hematopoietic elements may occasionally provide an intense 18-fluorodeoxyglucose uptake [7]. Magnetic resonance imaging (MRI) may well characterize the adipose tissue using chemical shift imaging biotechnology [2,3,13]. However, these modalities seem to be complementary to computed tomography with a little or non-significant additional benefits in most of the instances [1,3,13].

Preoperative percutaneous biopsy or aspiration from adrenal myelolipoma is not a common policy in the late literature. It has been infrequently employed for confirmation of diagnosis in cases of giant adrenal myelolipoma, because of the high diagnostic capabilities of the modern imaging modalities represented mainly by computed tomography and magnetic resonance imaging. Also, fears of potential complications such as rupture and/or hemorrhage may limit the use of preoperative biopsy [3,11]. However, it is mainly preserved to diagnosis of undefined primary adrenal tumors with a suspicion of malignancy [14] or exclusion of the possibilities of metastases from malignancy of the other organs of the body that may co-exist with adrenal myelolipoma [15].

Adrenal myelolipoma represents 90% of all lipomatous adrenal tumors [14]. Differential diagnosis of adrenal myelolipoma necessitates its characterization from other adrenal tumors including adrenocortical carcinoma, adrenal lipoma, retroperitoneal liposarcoma, and adrenal adenoma with fatty changes [3,14]. Adrenocortical carcinoma may attain large sizes, but it has irregular outlines [7]. Liposarcoma is another adrenal lesion with fat contents that may be confused with myelolipoma, but it is usually non-homogenous and infiltrative to the surroundings [3,5,7,10]. Although adrenal lipoma is very similar to adrenal myelolipoma on imaging, its extremely low incidence may down estimate its clinical relevance [7].

Classically, the asymptomatic small-sized adrenal myelolipomas are considered for conservation, while the large-sized ones are subjected to open surgery [2,14]. However, laparoscopic approach has been

Figure 1  Abdominal computed tomography scan. (A) Transvers cut: giant right adrenal mass with heterogeneous pattern. Note the compression of the inferior vena cava. (B) Sagittal view: giant right adrenal mass displacing the right kidney inferiorly. Note the well-defined demarcation from surroundings.
employed for significant large-sized lesions with a progressively supervening role in large adrenal myelolipomas [4,16]. One of the determinant factors of treatment option is the tumor size, where sizes larger than 5–6 cm were considered previously as contraindications to laparoscopy. However, this issue has been changed by removal of challenging masses up to 15 cm (Supplementary Table 1). Also, body mass index represented an important factor which requires availability of high laparoscopic experiences [4,6,16]. Considering the large tumor size as a consistent indication for surgical intervention [1,7], observational or conservative management has been very rarely employed for large and giant adrenal myelolipomas (Supplementary Table 1). Adrenal myelolipoma is prone to progression and development of manifestations along its natural course. So, the rationale for surgical removal is the avoidance of potential life-threatening complications such as spontaneous rupture and hemorrhage that are related to the large sizes [7].

The term “giant” has been commonly employed to represent the extent of size of adrenal myelolipoma in context of symptom development, treatment option and surgical approach (Supplementary Table 1). Although this term is regularly used, its numerical value is variable and progressively increasing from small-sized to very large-sized lesions which have been subjected to successful laparoscopic excision [4,16]. In spite of this, the 10 cm value has been used as a cut-off point to define this term [1]. According to this definition, more than 180 cases of giant adrenal myelolipomas could be collected from all the published cases in the English literature (Supplementary Table 1). So, the current case size of 16 cm with regards to its laparoscopic excision, definitely, deserves the use of this term. However, open adrenalectomy is still representing a viable approach for tumors of similar sizes [2,14], especially in the presence of other indications such as tumors in other organs [8] and major complications like hemorrhage and intestinal obstruction [11].

Details of the laparoscopic adrenalectomy procedure have been described in few instances of myelolipoma [5,6,16]. This observation could be attributed to the smaller sizes in those cases relative to the large sizes in the most recent cases such as the current one. The anatomical extensions were surgically demanding and unreported previously in such cases of laparoscopic excision of adrenal myelolipoma. Moreover, large tumors may indicate a relatively long incision up to 6 cm for retrieval of the specimen after laparoscopic excision. One of the ports’ sites is usually extended for this purpose according to the size of the mass. In spite of the potential suspicion of malignancy and fears of rupture of large adrenal masses, the characteristics of benign nature of adrenal myelolipoma in imaging may play a role in assuring surgeons while they are retrieving these sizes [4,6,17].

In comparison to open adrenalectomy, no doubt that laparoscopic approach has the advantages of avoidance of the generous wound hazards, and reduced hospital stay and convalescence. However, still the limiting factors for this technique present like obesity, large tumor sizes, and surgeon’s experience. Accordingly, the more the surgeon’s experience grow, the larger the sizes of the tumor that could be managed laparoscopically [6]. In the current case, obesity,
Figure 3 Histopathological (Microscopic) features of adrenal myelolipoma. (A) Section of tissue showing the components of benign mature adipose tissue and myeloid elements (X400). (B) Fat cells and the three elements of hematopoietic marrow are seen by a higher power (X1000).

large tumor size, and difficult anatomical extensions represented significant challenges. However, these issues were managed and overcome, successfully. The previously reported technical difficulties were encountered, mainly, during ligation of the adrenal vessels and dissection of the mass from the inferior vena cava [3,5,6]. However, in the current case, significant challenges were encountered during the dissection of the difficult mass extensions.

In the context of the large-sized myelolipomas, concerns of technical failure and length of the procedure could be raised. However, these concerns have been gradually ameliorated with increasing experiences [3,5,6]. Our case was managed in a satisfactorily timing without conversion to open surgery. Also, blood loss may be massive in large tumor surgery, while laparoscopy has the advantages of good hemostasis even with the large-sized tumors [5,6]. Accordingly, in our case, the blood loss was low without indication for blood transfusion which could be attributed to the laparoscopic magnification powers.

Feasibility of the laparoscopic approach for excision of the giant adrenal myelolipomas is being progressively confirmed to replace the standard open surgical adrenalectomy approach [4,16]. Present-
Challenging giant adrenal myelolipoma

References


