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JSM Gastroenterology and Hepatology

Case Report

Single-Port Retroperitoneal Robotic Treatment for Adrenal Angiomyolipoma: Description and Review of the Literature

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Submitted: 25 August 2023
Accepted: 30 September 2023
Published: 30 September 2023

Keywords
- Angiomyolipoma
- Adrenal
- Retroperitoneoscopic
- Robot-assisted
- Single-port
- Adrenalectomy

Abstract

Angiomyolipomas (AMLs) are benign tumours of mesenchymal origin that derivate from perivascular epithelioid cells and are most commonly found in the kidney, with a strong association to disorders like Tuberous Sclerosis and lymphangioleiomyomatosis but there are several extra-renal locations where AMLs can arise, the most common being the liver, followed by the spleen, bone and lungs being the adrenal one of the rarest locations where they can appear. Concerning adrenal AMLs, only 21 cases have been described in the English literature so far, most of them using an open approach. Sizes reported range between 0.2 and 17.3 cm. We present a review of the literature and the presentation of the first adrenal AML treated by a robotic retroperitoneal approach via-single port being this a feasible and good approach to treat this kind of lesions despite de size with great cosmetic and algetic outcome.

INTRODUCTION

Angiomyolipomas (AMLs) are benign tumours of mesenchymal origin that derivate from perivascular epithelioid cells. Tumours derived from these cells have been recently grouped under the term PEComas and tend to have low malignancy rates. Its etiopathogenesis rely on alterations concerning the genes from the Tuberous Sclerosis Complex [1].

AMLs contain a mixture of blood-vessels, muscle cells and adipose tissue, with specific staining to HMB-45 marker [2] and are most commonly found in the kidney, with a strong association to disorders like Tuberous Sclerosis (TS) and Lymphangioleio Myom Atosis (LMA), but there are several extra-renal locations where AMLs can arise, the most common being the liver, followed by the spleen, bone and lungs [3]. Approximately half of AMLs are sporadic (non associated to TS or LMA) and are more commonly found in women (2:1 ratio) [2].

AMLs rarely cause any clinical symptoms, being non-specific abdominal pain the most common clinical feature; this is why they are usually diagnosed as incidental findings in image testing [2]. In extremely rare cases, the proliferation of blood-vessels can cause spontaneous bleeding, leading to a retroperitoneal haemorrhage that can cause shock and even the death of the patient.

On CT scan, AMLs are seen as non-homogeneous, well-defined masses with abundancy of fatty attenuation. These are usually sufficient findings for radiologic diagnosis. Treatment includes a wide range of alternatives. On small lesions, radiologic follow-up may be enough. However, in large lesions and bleeding-prone masses, surgical excision is generally recommended, with or without previous embolization [2,4].

Concerning adrenal AMLs, only 21 cases have been described in the English literature so far, being one of the rarest locations where AMLs can appear. The association to TS or LMA is very weak in adrenal AMLs compared to renal ones [5].

Here, our purpose is to make an exhaustive review of the present literature concerning adrenal AMLs and present a case of adrenal AML that was admitted to our inpatient clinic for surgical excision through Single-Port (SP) retroperitoneal laparoscopic approach, a minimally invasive surgery that has not yet been described for this kind of tumour.

CASE PRESENTATION

Here we describe a case of a 56-year-old woman who was admitted to our inpatient clinic to receive surgical adrenalectomy.

She had a medical history of AAS allergy, hypothyroidism and smoking, with no other associated comorbidities. On a routine urine sediment solicited by her general practitioner, asymptomatic microhaematuria was detected, so she was studied with an abdominal US, finding a retroperitoneal heterogeneous mass with a suspected adrenal origin.

She was referred to CT scan to further study the mass, finding a well-defined, non-homogeneous, hypodense mass with right adrenal origin. It measured 40x32x44 mm (Figure 1). MRI follow-up was recommended, so she underwent an MRI six months later which showed a right adrenal mass with heterogeneous intensity in T1-enhanced sequences and hyperintensity in T2-enhanced sequences. Comparing to previous imaging techniques, no growth was detected.

After that, the patient was proposed to receive surgical excision of the right adrenal gland, so she was admitted to our inpatient clinic. She underwent SP robot-assisted retroperitoneal adrenalectomy trough a single 4 cm incision Da Vinci Xi® platform was used using a multiarm setting with no need of accessory trocars or incisions. Total surgery time was 100 minutes with less than 100 cc bleeding (no drain tube was left); patient went on an uneventful recovery and was discharged one day after the surgery (hospital stay of 20 hrs) with no need of intravenous analgesia.

Pathology analysis of the mass showed an angiomyolipoma originating of the right adrenal gland with areas of haemorrhagic infarction. Margins were tumour-free.

No post-operative complications were detected after patient discharge and as of this day (6 months after the surgery), no tumour recurrence has been detected on follow-up imaging.

METHODS

For our review, we searched articles through the medline-pubmed data index using the words “Adrenal” AND “Angiomyolipoma” in the title and “adrenal angiomyolipoma” in any text field.

The results showed for the first search 28 results and 17 for the second. We crossed both search results and then we finally selected 20 case-report articles that were in English language and contained in their text a description of at least one case concerning an adrenal angiomyolipoma, being a total of 23 cases (plus the present case, a total of 24 cases) [3,5–23].

Of all the 20 case-reports we analysed, we decided to exclude the Article from Goswami et al. [22], because of discordance between AMLs and their pathological findings: their article talks about an adrenal AML, but they point out the finding of hematopoietic tissue in the microscopic examination of the tumour. This is a characteristic pathological finding of myelolipomas [24].

It is also worth noting the word-by-word similarity between Yener et al. (20) and Godara et al. [18], case-reports (being the latter the original of the two), though we decided to review both of them because of the clinical differences between both cases.

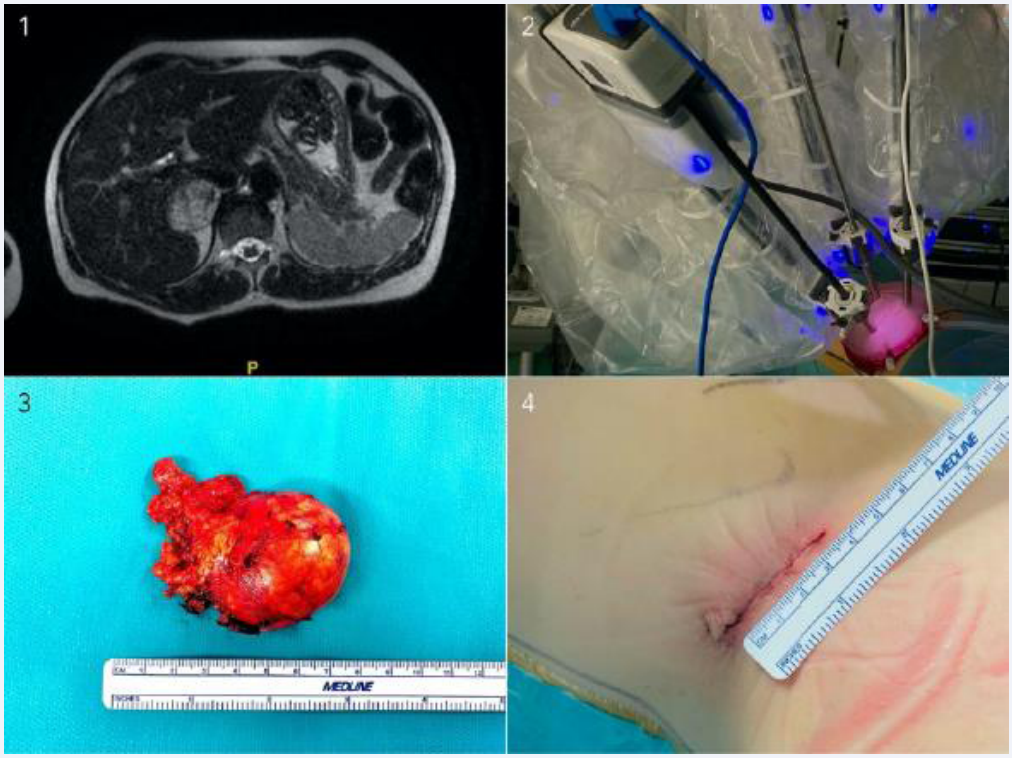

Figure 1 1. MRI-T2 showing the right adrenal mass. 2. Da Vinci Xi docking in a single-port setting. 3. Adrenal specimen. 4. Cosmetic result after specimen removal.

Table 1: Angiomyolypomas series in the literature

Case No.	Author / year	Sex/ age	Size (cm)	Side	Presentation	Imaging	CT features	MRI features	Surgical approach
1	Lam K / 2001	M/20	0.2 (specimen)	L	Incidental finding in nephrectomy (TS)	CT	AMLs in liver and kidney	-	Open L nephrectomy + adrenalectomy
2	Lam K / 2001	F/46	8.0 (CT)	L	Incidental CT finding	CT	Heterogeneous contrast enhancement	-	Open L adenalectomy
3	Elsayes K /2005	F/49	12.2x9.8x6.8 (MRI)	R	Asymptomatic	CT/MRI	-	Fat signal with suppression in fatty suppression sequence	R adrenalectomy
4	Sutter R / 2007	F/32	6.0 (CT)	R	Abdominal pain (LMA)	CT/MRI	Partial enhancement	-	R laparoscopic adrenalectomy
5	Godara R / 2007	F/45	15.0x12.0 (US)	L	Epigastric pain	CT	Non-homogeneous, well defined	-	Open L adrenalectomy
6	D'Antonio A / 2009	M/42	6.0x4.5 (CT)	L	Flank pain + shock: Intratumoral bleeding (eAML)	CT/MRI	Well defined with intratumoral bleeding	T2 hyperintensity	Emergent open L nephrectomy and adrenalectomy
7	Yener O / 2011	F/45	5.0x6.0 (CT)	R	Right flank pain	CT/US	Low density	-	R laparoscopic adrenalectomy
8	Hu H /2012	F/55	15.0x16.0 (CT)	R	Right abdominal pain	CT/US	Non-homogeneous, well defined	-	Open R adrenalectomy
9	Zhao J / 2014	M/70	8.0 (MRI)	L	Asymptomatic	CT/MRI	-	T1 and T2 hyperintensity	Open L adrenalectomy
10	Zhao J / 2014	F/47	9.5x8.0x2.0 (MRI)	L	Asymptomatic	CT/MRI	-	T1 and T2 hyperintensity	Open L adrenalectomy
11	Li W / 2015	M/53	9.0x6.0 (US)	L	Left abdominal pain	CT/MRI/ US/PET	Low density with PET enhancement	T2 hyperintensity	L laparoscopic adrenalectomy
12	Kwazneski D /2016	F/65	11.3x9.4 (CT)	R	Right abdominal pain	CT/US	Heterogeneous and calcifications	-	Open R adrenalectomy
13	Selviambigapathy J / 2017	F/60	12x10x8 (specimen)	L	Abdominal pain + general syndrome (sarcoidosis)	CT	Heterogeneous with intratumoral bleeding	-	Open Lpartial nephrectomy and adrenalectomy
14	Antar A / 2017	F/48	10.0 (CT)	R	Right flank pain	CT	Hypodense	-	Open R adrenalectomy
15	Ghimire O / 2017	M/36	5.2x4.2x3.1 (CT)	R	Incidental finding	US/CT/MRI	Well defined, oval with fatty density	-	Open R adrenalectomy
16	Ghimire O / 2017	F/61	8.6x9.5x8.1 (CT)	R	Incidental finding	US/CT/MRI	Well defined, with fatty density		Open R adrenalectomy
17	Duralska M / 2018	F/35	7.0x6.0x9.0 (MRI)	L	Abdominal pain	US/CT/MRI	Low attenuation	Heterogeneous, lobular mass with contrast enhance.	Open L adrenalectomy
18	Kord V / 2019	F/33	7.3x6.9 (CT)	L	Abdominal disconfort (TS)	CT	Enhanced soft tissue density mass	-	Open L adrnalectomy
19	Ahmed M / 2020	M/68	17.3x14.6x16 (CT)	R	Upper abdominal pain	CT	Fatty mass	-	Open R adrenalectomy
20	Yang Y / 2020	M/64	3.6x4.0 (CT)	L	Referred for follow-up CT	CT	Low attenuation with ring enhancement	-	L retroperitoneal laparoscopic adrenalectomy
21	Bai Y / 2021	M/40	1.6x2.3 (CT)	L	Referred for follow-up CT	CT/US	Calcification	-	L retroperitoneal laparoscopic partial adrenalectomy
22	Cicek M / 2022	F/64	9.5x6.8 (CT)	L	Microscopic hematuria. (eAML)	US/CT	Well defined, heterogeneous, spherical	-	L laparoscopic adrenalectomy
23	Present case	F/56	4x3.2x4.4 (CT)	R	Microscopic hematuria	US/CT/MRI	Well defined, non-homogeneous. Calcifications	T1 intermediate intensity with T2 hyperintensity	Single-port robotic retroperitoneal laparoscopic R adrenalectomy

Abbreviations: TS: Tuberous Sclerosis; LMA: Lymphangioleiomyomatosis; eAML: epidermoid Angiomyolipoma

DISCUSSION

Doing a comprehensive review of the existing literature, we see that most cases of described adrenal AMLs occur in women, in an almost 2:1 relation. Median age at diagnosis is 47 years old, with most of the cases occurring between the 30s and 40s.

Clinical manifestations vary from completely asymptomatic cases to extremely severe syndromes like retroperitoneal bleeding with associated hypovolemic shock [19]. Still, the most common symptoms are non-specific abdominal discomfort or pain; this leading to routine image examinations, more commonly abdominal ultrasonography, showing a mass in the adrenal area that needs further study to be correctly assessed. Completely incidental findings are not infrequent, with no clinical manifestations associated, achieving suspicion or diagnosis through non-related image examinations.

In opposition to renal AMLs, the association to TS or LMA is much weaker. Only two cases have been associated to TS in our review [11,16] and one has been associated to LMA [17]. Though one of the TS cases [16] presented AMLs in other regions (kidney and liver, the most common places), this finding was not present in any other case, aways being isolated lesions. An association to sarcoidosis was also described, but in this case, the conclusion reached was that the association was fortuitous and non-causal [8].

Radiologically, adrenal AMLs show similar characteristics to renal AMLs: non-homogeneous, well-defined tumours with fatty attenuation and mild contrast enhancement. Some cases showed less fatty content, usually being due to a variant, the epithelioid AML [11,19]. In another case, due to proximity between structures, an adrenal AML was thought to be from kidney origin [5], leading to radical nephrectomy, achieving the diagnosis only after pathological examination. On MRI, the most common feature is the hyperintensity of the tumour with suppression of its parenchyma in fatty-suppression sequences. Still, all of these features are shared with the more common myelolipoma, making very difficult to distinguish one tumour from other only by imaging examinations.

On one previous case, positron enhancement was proved under the suspicion of a lung-cancer metastasis, a false impression of malignancy [6].

Pathological findings do not differ from renal AMLs: tumours containing a mixture of blood vessels, muscle and fatty tissue, being the HMG-45 marker, the common marker associated to AMLs.

Concerning its treatment, all of the reviewed cases, including ours, were referred to surgical excision of the mass. The approach is not always defined, sometimes only referred as “adrenalectomy”. Still, most reviewed cases were operated under open surgery. Only six cases [6,13-15,17,20] plus our case were intervened under laparoscopic procedures.

Thus, in our case, we described an approach using retroperitoneal laparoscopy, only used in 2 of the reviewed cases [13,14], but with the singularity of being robot-assisted and using a single-port access, which is a minimally invasive procedure that allows surgery using multiple laparoscopic tools through a single incision that gives access to the retroperitoneum, fulfilling the surgery with only a minimal scar in the back of the patient (3-4 cm), minimising morbidity, and enhancing aesthetic results.

Post-operative recovery was also optimal thanks to this approach, which resulted on the patient being discharged on the first day after the surgery with no post-operative complications detected in any of the follow-up outpatient visits.

None of the reviewed cases used a SP access or a robot-assisted retroperitoneoscopic technique. We believe our approach to be the best alternative to reduce post-operative morbidity and minimise skin-scarring. SP access is well-known in our centre and has given excellent result thus far, including retroperitoneal robotic approach [25,26]. This is a feasible approach, even in a multi-arm setting, that provides different advantages in the treatment of benign adrenal lesions (Table 1).

Authors Contributions

Anxo González-Manso wrote the main manuscript and prepared the tables. Dario Vázquez-Martul prepared the figures and provided the original material for them. All authors reviewed the manuscript.

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