



# Incidental giant adrenal lymphangioma presenting as nonfunctional cystic mass

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## ABSTRACT

Surrenal masses can be encountered with many different clinical manifestations and a diverse spectrum of etiologies in clinical practice. Recent advances in imaging and laboratory studies as well as their increasingly widespread use and easy accessibility have currently made it possible to diagnose a greater number of surrenal masses than ever. The basic approach principles vary for incidentally detected masses, benign/malignant masses, and hormonally active masses. Lymphangiomas are benign congenital malformations of lymphatic channels that primarily affect the neck and head region. They are typically found in children younger than 2 years of age, they are uncommon in adults, and they rarely involve surrenal glands. In this paper, we present a woman with a hormonally inactive right giant surrenal mass showing recent rapid growth, which was diagnosed to be a lymphangioma with an atypical localization in the histopathological examination. The patient was operated with right surrenalectomy and total mass excision via laparoscopic lateral transperitoneal approach.

**Keywords:** Adrenal masses, laparoscopic surgery, lymphangioma

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## INTRODUCTION

When deciding to proceed with surgery for surrenal masses, their size, hormonal activity, imaging signs suggestive of malignancy, and growth rate at serial examinations should be taken into account. There is no consensus for a size threshold beyond which a surgical intervention becomes necessary. While some authors have advocated that the surgical mass excision should be limited to masses larger than 6 cm based on the knowledge that masses smaller than 6 cm are associated with a negligible risk of malignancy, others have recommended surgery for masses larger than 3 cm, and still others for masses larger than 4 cm (1). Herein we present a woman with a nonfunctional giant right surrenal mass that turned out to be a lymphangioma, a tumor that is uncommonly considered in the differential diagnosis in this localization.

## CASE PRESENTATION

A 39-year-old woman had been followed up at an outside center for a right surrenal mass for 8 years. She presented to our urology department after the mass had grown rapidly and caused abdominal pain over the past 6 months. Her history was not notable for any disorder. On the physical examination, she had tenderness in her right lateral and right upper quadrants. Biochemical tests and hemogram parameters were in a normal range. A preoperative endocrinological evaluation including a 24-hour urine collection for vanilylmandelic acid, epinephrine, metanephrine, norepinephrine, normetanephrine, dopamine; plasma renin and angiotensin levels; and 1 gr dexamethasone suppression test were all normal. An abdominal computed tomography (CT) revealed a well-bordered mass lesion 86x70 mm in size and millimetric calcifications in the right surrenal gland; the mass was primarily considered to be a surrenal cyst shifting surrounding structures. A magnetic resonance imaging (MRI) examination demonstrated a cystic lesion measuring 9x7 cm in the right surrenal gland, which appeared hypointense on the axial T1A and axial fat suppression T1A images and hyperintense on the axial fat suppression T2A images (Figure 1). A surgical intervention was scheduled based on the recent rapid growth, signs of compression, and patient's desire to become pregnant. The patient underwent a laparoscopic lateral transperitoneal right surrenalectomy in which the mass and the right surrenal gland were excised. The patient had no problems during the postoperative period and was discharged 2 days later. Immunohistochemical examination showed CD31- and D2-40-positive cells on the wall of the multilocular cystic mass, and the lesion was identified as a lymphangioma (Figure 2). Informed consent was obtained from the patient who participated in this study.

## DISCUSSION

Despite the general knowledge that surrenal mass lesions are typically benign and do not release any hormones, each mass should also be evaluated, and its differential diagnosis should be done for hypersecretory syndromes or tumor development (2). The management of hormonally inactive surrenal

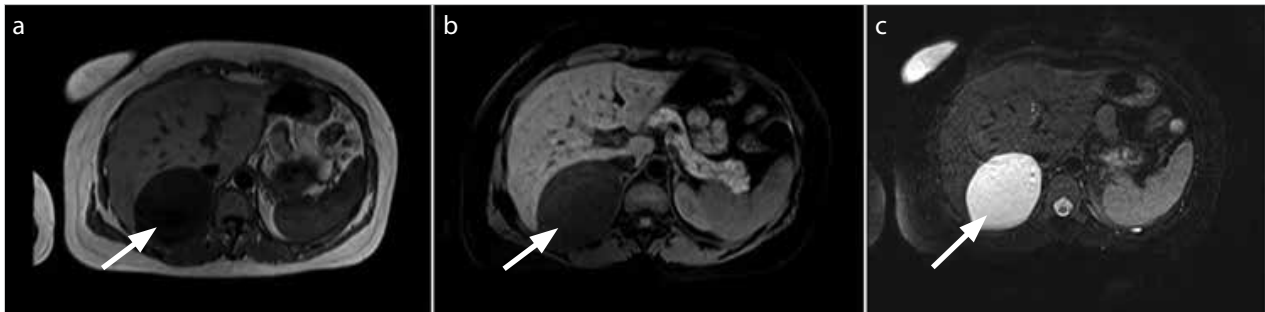


Figure 1. a-c. A cystic lesion in the surrenal gland, which appears hypointense on the axial T1A (a) and axial fat suppression T1A images (b), and hyperintense on fat suppression T2A images (c)

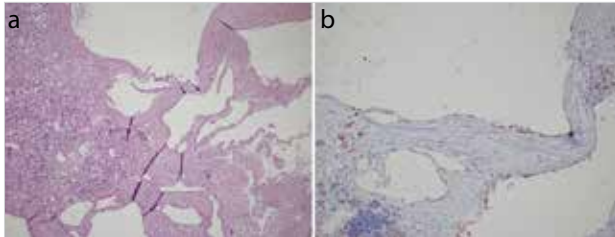


Figure 2. a, b. A histological view of the lymphangioma of the right adrenal gland; multiloculated cyst within the adrenal gland (a), cells positively stained with CD31 and D2-40 on the wall of the multilocular cystic lesion that contains the areas of focal dystrophic calcification and is divided by fine septae (b) (H&E: 200×)

masses is primarily based on the lesion size. As inactive masses smaller than 3 cm are typically of benign character, their conservative follow-up is usually recommended. Inactive masses between 3 and 5 cm in size can be conservatively managed when they appear homogenous in radiological imaging studies. However, surgery should be considered whenever radiological studies indicate growth. Hormonally active masses should be surgically excised irrespectively of their size (3).

CT and MRI are the most appropriate imaging modalities for differentiating adenoma, carcinoma, and pheochromocytoma from one another. These imaging modalities are also very beneficial for determining the surgical candidacy from an anatomicopathological standpoint (4).

Following the introduction of laparoscopic surgery for surrenal adenomas, it has been rapidly incorporated into the clinical practice, and studies comparing laparoscopic and open surgeries have been published. Laparoscopic surgery has the main advantages of short hospital stay, reduced postoperative pain, rapid recovery, and a better cosmetic outlook. Since its first introduction, the laparoscopic approach has been the technique of choice for the treatment of benign functional and nonfunctional surrenal mass lesions (5). Recently, the indications of laparoscopic interventions have been extended to larger surrenal masses and surrenal metastatic lesions. Different laparoscopic techniques have been defined for the resection of surrenal masses, including lateral transabdominal, lateral retroperitoneal, anterior transabdominal, and posterior retroperitoneal approaches. Among these, the lateral transperitoneal approach is widely used for surrenal mass lesions (6).

Adrenal cysts are rare, typically asymptomatic lesions that are usually detected postmortem. They are clinically important since they can be confused with malignant lesions. Symptomatic lesions manifest with the triad of pain, palpable mass, and inferior displacement of the kidney. These lesions have four major groups, which are the parasitic, endothelial, epithelial, and pseudocystic types. Despite affecting every age from birth to old age, they are most commonly observed in middle-aged women (7).

Lymphangiomas are benign congenital malformations of lymphatic channels that primarily affect the neck and head regions. Approximately 50% of these lesions are diagnosed at the time of birth, and 90% during the first 2 years of life. They are quite rare in adulthood. Ninety-five percent of lymphangiomas are located in the neck and axillary region, while the rest develop in the mediastinum, mesentery, omentum, retroperitoneum, and bones. Their diagnosis is made by physical examination, anamnesis, and imaging studies (8).

Cystic lymphangiomas are composed of sequestered lymphatic sacs. Although the histogenesis of lymphangiomas is still debated, some researchers assume that they are acquired lesions secondary to the obstruction of chylous vessels by inflammatory, traumatic, and degenerative conditions. In contrast, cystic lymphangiomas have been reported to occur congenitally, as a result of the proliferation of the embryonic lymph sac remnants. It is the general opinion that lymphangiomas are composed of sequestered lymphatic sacs that fail to establish a link with main lymphatic channels (9).

Despite having a benign character, cystic lymphangiomas may lead to the compression of adjacent organs and the obstruction, and they may also invade surrounding structures. No spontaneous regression is expected in adulthood lymphangiomas. A total surgical excision is necessary for their treatment. Incision, drainage, and repeat aspirations have been used as primary therapies, although recurrences and infections have proved to be ineffective (9, 10). As the mass lesion of our patient had recently grown rapidly and compressed the surrounding structures, surgical excision was decided despite the lack of any suspicion for a malignancy.

## CONCLUSION

Albeit rare, lymphangiomas should be considered in the differential diagnosis of surrenal masses in adult patients. The definite diagnosis is made after the surgical removal by histological and immunohistochemical examinations. As they are

located deeply in the retroperitoneal area, lateral transperitoneal laparoscopic surgery can be readily used for their treatment owing to its advantages such as a shorter hospital stay, early return to daily life, and superior cosmetic outcomes.

**Informed Consent:** Informed consent was obtained from the patient who participated in this study.

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