CASE REPORT JSLS

# Laparoscopic Adrenalectomy for Unsuspected Unilateral Primary Adrenal Lymphoma

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## ABSTRACT

**Background:** Rarely, a patient presents to a surgeon for evaluation of an adrenal incidentaloma where the final pathology is primary malignancy. For primary adrenal lymphoma, fewer than 100 cases have been reported in the literature.

**Case Report:** We report a case of unilateral primary adrenal aggressive B cell lymphoma discovered incidentally in a 41-year-old female. Preoperative testing demonstrated the 6-cm mass to be biochemically silent. Subsequently, the patient underwent a laparoscopic adrenalectomy. Following pathologic diagnosis of B cell lymphoma, a metastatic workup was negative, and she underwent treatment with systemic chemotherapy. She is currently disease free 6 months postoperatively.

**Conclusion:** Primary adrenal lymphoma should be considered in patients with unilateral adrenal incidentaloma. We believe that adherence to guidelines of resection of incidentalomas allowed for early surgical intervention and possible cure.

**Key Words:** Laparoscopy, Adrenalectomy, Lymphoma, Adrenal incidentaloma.

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DOI: 10.4293/108680811X13125733356639



## INTRODUCTION

Laparoscopic adrenalectomy was first introduced in 1992.[1,2] Since that time, it has become the procedure of choice for simple adrenal masses,[3–5] even those that are large.[6,7] We present here a case report of a laparoscopic adrenalectomy performed for unilateral adrenal enlargement with the unexpected pathologic diagnosis of B cell lymphoma.

## CASE REPORT

A 41-year-old African American female with BMI of 49kg/m$^2$ and past medical history significant for hypertension and noninsulin dependent diabetes was in a motor vehicle collision. The trauma workup was unremarkable, but the patient was noted to have a left adrenal gland that was enlarged to 3.6cm. She had no other relevant past medical or past surgical history, and she did not smoke tobacco or use alcohol or illicit drugs.

The patient continued to have intermittent left flank pain, which prompted repeat imaging 3 months later. At that time, she was noted to have a 5x6x7-cm adrenal mass with an additional cystic component **(Figure 1)**. The patient was otherwise well and had no specific medical complaints. She had been tolerating a regular diet and had no systemic symptoms, such as fever, chills, or night sweats.

Preoperative testing suggested that the mass was biochemically silent. Specifically, 24-hour urinary catecholamines, metanephrine, and normetanephrine were within normal limits. Urinary free cortisol was unremarkable. Serum rennin, aldosterone, and potassium were within normal limits.

We decided to take the patient to the operating room for a laparoscopic left adrenalectomy. The patient was placed in standard lateral decubitus positioning, and was well padded. Pneumoperitoneum was established with a Veress needle through a left subcostal position, and a 5-mm camera with an optical access trocar was placed in the left epigastrium. An additional 5-mm working port was placed in the subxiphoid region, and a 10-mm working port was placed in the left lateral subcostal position. The spleen was mobilized medially and the splenic flexure taken down using a Harmonic scalpel. Dissec-

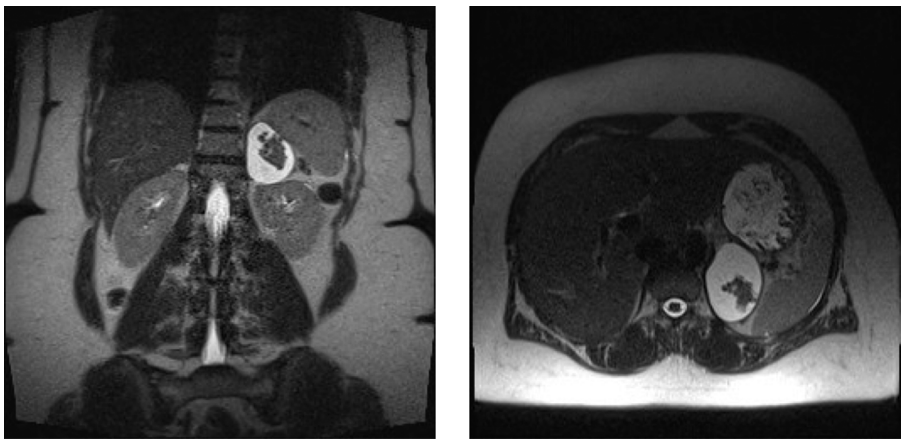

**Figure 1.** Magnetic resonance imaging (MRI) of the abdomen, demonstrating enlarged left adrenal gland.

tion of the adrenal gland proceeded uneventfully, but the gland appeared even larger than the preoperative imaging suggested. There was a dominant arteriole in the inferior aspect of the gland that was clipped, and the adrenal vein itself was separately dissected and clipped prior to division. The gland was placed in a retrieval bag, and removed via the 10-mm port site. The fascia and skin were closed in the usual fashion. The patient had an uneventful postoperative course and was discharged home on postoperative day 2.

Final pathology measured the gland at 7.2cm x 5.3cm. Within the borders were a 3.1-cm cyst and a 4.1-cm fungating mass that was positive for CD45, CD20, and Bcl-2 on immunohistochemistry, diagnostic for B cell lymphoma. In situ hybridization for Epstein Barr Virus was positive, and Ki-67 proliferation index was >95%.

The patient underwent a full lymphoma workup. Whole body PET/CT imaging was negative. Bone marrow aspirate and biopsy were also negative for malignancy. No EBV DNA was found in the serum.

The patient was treated for aggressive B cell lymphoma with systemic chemotherapy using dose-adjusted R-EPOCH (Rituximab, Etoposide, Prednisone, Vincristine, Cyclophosphamide, and Doxorubicin). At 6 months postoperative, the patient has no evidence of active disease, and has resumed work full time.

## DISCUSSION

Primary lymphoma of the adrenal gland is a decidedly rare condition, with fewer than 100 cases having been described in the literature.[8,9] Indeed, there have only been 9 case reports of primary adrenal lymphoma treated surgically.[9,10] More recently, 5 cases of lymphoma have been reported in the context of larger series, and are listed in **Table 1**.[11–15]

**Table 1.**
Laparoscopic Cases for Primary Adrenal Lymphoma Reported in Surgical Series

| Author | Year | Number of Patients | Incidence |
|---|---|---|---|
| Buurman[11] | 2006 | 2 | 2/282 adrenals, 10 yrs |
| Kirshstein[12] | 2008 | 1 | 1/26, 10 yrs |
| Liao[13] | 2006 | 1 | 1/210, 9 yrs |
| Naya[14] | 2005 | 1 | 1/169, 9 yrs |
| Yavuz[15] | 2005 | 1 | 1/23, 3 yrs |

Primary adrenal lymphoma accounts for less than 1% among non-Hodgkin lymphomas, and most are large B cell lymphomas.[16] It is presumed to derive from hemopoietic cells within the adrenal gland itself and may be more likely to occur in immunosuppressed patients.[8] Most cases involve both adrenal glands, and patients present with clinical symptoms of adrenocortical insufficiency.

The average survival seems quite poor, with a reported mean survival of only 4 months.[17] Particularly poor prognostic factors are listed in **Table 2**. Chemotherapy seems the mainstay of treatment, with the role of surgery and radiation therapy not well defined at present. Despite its rarity, chemotherapy regimens have been described, and complete remission has been achieved.[18]

An adrenal incidentaloma is defined as a serendipitously discovered adrenal lesion >1cm. While the patient's adrenal mass fulfilled these criteria upon her presentation to

**Table 2.**
Poor Prognostic Factors in Primary Adrenal Lymphoma (after Zhang[10])

| Poor Prognostic Factors |
|---|
| Large size of tumor |
| Spread to other organs |
| Elevated lactate dehydrogenase |
| Old age |
| Adrenal insufficiency on initial presentation |

the trauma center, further workup of the mass was expedited secondary to its symptomatic nature. Thus, in our patient, surgery seems to have occurred at the earliest stage of the disease process. Additionally, the patient has none of the poor prognostic factors listed in **Table 2** save for large tumor size. We opted for an aggressive chemotherapy regimen, but a good outcome may be related to the early stage as much as the adjuvant therapy. In any event, the laparoscopic approach was feasible, and resulted in minimal surgical morbidity to the patient.

## CONCLUSION

Laparoscopic surgery for the incidentally enlarged adrenal mass may reveal unexpected findings. In this case, primary adrenal lymphoma, of the large B cell type, was clinically and radiographically unsuspected due to its extreme rarity. The patient has done well, and is being treated with systemic chemotherapy given the overall poor prognosis of the condition.

**References:**


1. Gagner M, Lacroix A, Bolte E. Laparoscopic adrenalectomy in Cushing's syndrome and pheochromocytoma. *N Engl J Med.* 1992;327:1033.

2. Petelin JB. Laparoscopic adrenalectomy. In *Proceedings.* Third World Congress, Endoscopic Surgery. Bordeaux, France. September 1992.

3. Lee J, El-Tamer M, Schifftner T, et al. Open and laparoscopic adrenalectomy: analysis of the national surgical quality improvement program. *J Am Coll Surg.* 2008;206:953–959.

4. Smith CD, Weber CJ, Amerson JR. Laparoscopic adrenalectomy: new gold standard. *World J Surg.* 1999;23:389–396.

5. Assalia A, Gagner M. Laparoscopic adrenalectomy. *Br J Surg.* 2004;91:1259–1274.

6. Rosoff JS, Raman JD, Del Pizzo JJ. Laparoscopic adrenalectomy for large adrenal masses. *Curr Urol Rep.* 2008;9:73–79.

7. MacGillivray DC, Whalen GF, Malchoff CD, et al. Laparoscopic resection of large adrenal tumors. *Ann Surg Oncol.* 2002; 9:480–485.

8. Grigg A, Connors JM. Primary adrenal lymphoma. *Clin Lymphoma.* 2003;4:154–160.

9. Wang J, Sun NC, Renslo R, et al. Clinically silent primary adrenal lymphoma: a case report and review of the literature. *Am J Hematol.* 1998;58:130–136.

10. Zhang LJ, Yang GF, Shen W, Qi J. Imaging of primary adrenal lymphoma: case report and literature review. *Acta Radiol.* 2006;47:993–997.

11. Buurman H, Saeger W. Abnormalities in incidentally removed adrenal glands. *Endocr Pathol.* 2006;17:277–282.

12. Kirshstein B, Yelle JD, Moloo H, Poulin E. Laparoscopic adrenalectomy for adrenal malignancy: a preliminary report comparing the short-term outcomes with open adrenalectomy. *J Laparoendsoc Adv Surg Tech A.* 2008;18:42–46.

13. Liao C-H, Chueh S-C, Lai M-K, et al. Laparoscopic adrenalectomy for potentially malignant adrenal tumors greater than 5 centimeters. *J Clin Endocrinol Metabol.* 2006;91:3080–3083.

14. Naya Y, Suzuki H, Komiya A, et al. Laparoscopic adrenalectomy in patients with large adrenal tumors. *Int J Urol.* 2005;12: 134–139.

15. Yavuz N. Laparoscopic transperitoneal adrenalectomy using the LigaSure vessel sealing system. *J Laparoendsoc Adv Surg Tech A.* 2005;15:591–595.

16. Mozos A, Ye H, Chuang WY, et al. Most primary adrenal lymphomas are diffuse large B-cell lymphomas with non-germinal center B-cell phenotype, BCL6 gene rearrangement and poor prognosis. *Mod Pathol.* 2009;22:1210–1217.

17. Singh D, Kumar L, Sharma A, et al. Adrenal involvement in non-Hodgkin's lymphoma: four cases and review of literature. *Leuk Lymphoma.* 2004;45:789–794.

18. Kim KM, Yoon DH, Lee SG, et al. A case of primary adrenal diffuse large B-cell lymphoma achieving complete remission with rituximab-CHOP chemotherapy. *J Korean Med Sci.* 2009; 24:525–528.