CASE REPORT JSLS

Laparoscopic Resection of Periadrenal Paraganglioma Mimicking an Isolated Adrenal Hydatid Cyst

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ABSTRACT

We present the case of a 64-year-old female with a periadrenal paraganglioma. The mass was diagnosed during abdominal ultrasonography performed to investigate right flank pain. Magnetic resonance imaging showed a lesion 48mm in diameter with an irregular hyperintense central margin, thickened septa, centrally located cystic component, and contrast enhancing peripheral portions. These findings made us think it was an isolated adrenal hydatid cyst. The mass was excised via laparoscopy without complications. Histopathological examination was consistent with periadrenal paraganglioma. We believe that the paraganglioma can mimic the radiological appearance of an isolated adrenal hydatid cyst, which should be taken into consideration during diagnosis.

Key Words: Paraganglioma, Periadrenal, Laparoscopy.

INTRODUCTION

Laparoscopic adrenalectomy is one of the successful interventions of minimally invasive surgery techniques. Since the first laparoscopic adrenalectomy was performed in 1992,[1] this approach has become a standard operation, and its popularity is continuing to spread. Several studies[2-5] have reported that laparoscopic adrenalectomy is followed by lower complication rates, less operative blood loss, less postoperative pain, earlier return to normal activity, and shorter hospital stay.

We present here the case of laparoscopic resection of a periadrenal paraganglioma that was discovered during a workup for flank pain in a female patient and mimicked the radiological appearance of an isolated hydatid cyst.

CASE REPORT

A 64-year-old woman was referred from our Endocrinology and Metabolism Diseases Clinic for a right adrenal mass. In her medical history, she suffered from right flank pain with a 2-year duration. Physical examination revealed no evidence of hypertension, cushingiod features, or endocrine hyperactivity. Abdominal ultrasonography revealed a solid mass measuring 38mm x 41mm in size in the right adrenal gland. On magnetic resonance imaging (MRI), T2 images suggested that a 48mm in diameter lesion was in the center of the right adrenal gland. The lesion included an irregular hyperintense central margin, thickened septa, centrally located cystic component, and contrast enhancing peripheral portions. It was not suppressed in out-of-phase images. The adrenal hydatid cyst could not be excluded due to the centrally located cystic component on the MRI **(Figure 1)**.

Preoperative laboratory studies were as follows: serum cortisol was 10.6mg/dL (normal range, 6.7 to 22.6), ACTH was 5.3pg/mL (normal range, 4.7 to 48.8), renin was 1mIU/mL (normal range, 4.4 to 46.1), aldosterone was 166.4pg/mL (normal range, 35 to 300), 24-hour urine collection for epinephrin was 5μg/24h (normal range, 4 to 20), norepinephrine was 55μg/24h (normal range, 23 to 105), metanephrine was 240μg/24h (normal range, 74 to 297), normetanephrine was 75μg/24h (normal range, 105 to 354), and vanillylmandelic acid was 1.5mg/24h (normal range, 1.4

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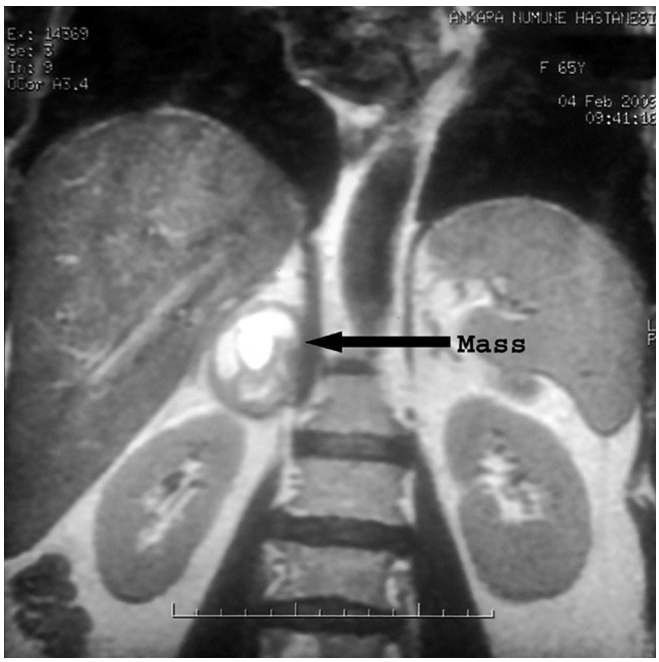

Figure 1. In magnetic resonance imaging, T2 images suggested that 48mm in diameter lesion in center of the right adrenal gland.

to 6.6). Serum and urine laboratory tests showed that the mass was nonfunctioning. The hydatid cyst latex agglutination test result was positive. In light of the above findings, after oral treatment of the patient for 15 days with Albendazole, laparoscopic transperitoneal excision of the adrenal mass was decided on.

After the induction of general anaesthesia, the patient was placed in a 30-degree lateral decubitis position. At surgery, pneumoperitoneum was established with a Veress needle. An 11-mm port was inserted via Visiport (Covidien Health Care, USA) just 2cm above the level of the umbilicus in the midclavicular line. Two 10mm and one 5mm ports were also inserted respectively: midline in the epigastrium to retract the liver by using Endo Retract Maxi (Covidien, Health Care, USA) at the lateral border of the rectus abdominis mid way between the umbilicus and the costal margin, and at the anterior axillary line in the subcostal region. Initially, we retracted the liver by using the retractor. Firstly, the right colon and the duodenum were mobilized to expose the inferior vena cava (IVC). Secondly, we mobilized the liver from its abdominal wall attachments high up along its lateral aspect. Also, the peritoneal attachments between the liver and the mass were mobilized. Then, we observed a thickened right adrenal vein in the posterolateral aspect of the IVC. Three 10-mm Hem-o-lok (Weck-Teleflex, USA) clips were used for clipping the adrenal vein, and two 10-mm Hem-o-lok clips were used for the distal and were cut. It was observed that the mass was attached to the adrenal. Thirdly, the attachments between the mass and the adrenal were dissected by using a 5-mm LigaSure (Covidien Health Care, USA) sealing device. It was observed that the medial aspect of the mass was attached to the IVC. The mass was dissected from the IVC by using the LigaSure sealing device through the lateral retraction with the laparoscopic nut. Later, the remainder of the tissue was dissected. When the right adrenal and the mass were completely free, we placed them in a retrieval bag, and we removed the mass and adrenal gland through a port site. After control of bleeding, we placed a Jackson Pratt drain. We did not encounter any intraoperative complications. The operative time and estimated blood loss were 100 minutes and 60cc, respectively. The drain was removed on the first postoperative day. The patient was discharged uneventfully on the second postoperative day without any postoperative complications. The patient is alive and has been doing well with no recurrence for 10 months.

Histopathologic Findings: Macroscopy

A smooth contoured, brown colored mass 48x45x40mm in size was observed in the immediate vicinity of the adrenal gland, which was 30x2x5mm in size. The cut sections of the mass were hemorrhagic and cystic. The cyst contained brown-colored hemorrhagic fluid. The cut sections of the adrenal gland were unremarkable **(Figure 2a)**.

Histopathologic Findings: Microscopy

Cut sections of the adrenal gland revealed no specific pathology. The cystic mass was separated from the adrenal gland and covered entirely by a thin fibrous capsule. The tumor harbored a rich vascular network and displayed nested and trabecular pattern. Tumor cells contained round, pleomorphic nuclei and eosinophilic cytoplasm. Neither mitosis nor necrosis was observed **(Figure 2b)**. The tumor cells showed diffuse and strong positivity with chromogranin, synaptophysin, neuron-specific enolase, S-100 protein, vimentin, and CD-56; on the other hand, cytokeratin, HMB-45, melan-A, desmin, inhibin, calretinin, estrogen receptor, progesterone receptor, CD-117, and low molecular weight keratin were all negative. The proliferation index with Ki-67 was evaluated by counting the positively stained nuclei in thousands of cells that was found to be 1%. Histopathological examination consisted of periadrenal paraganglioma.

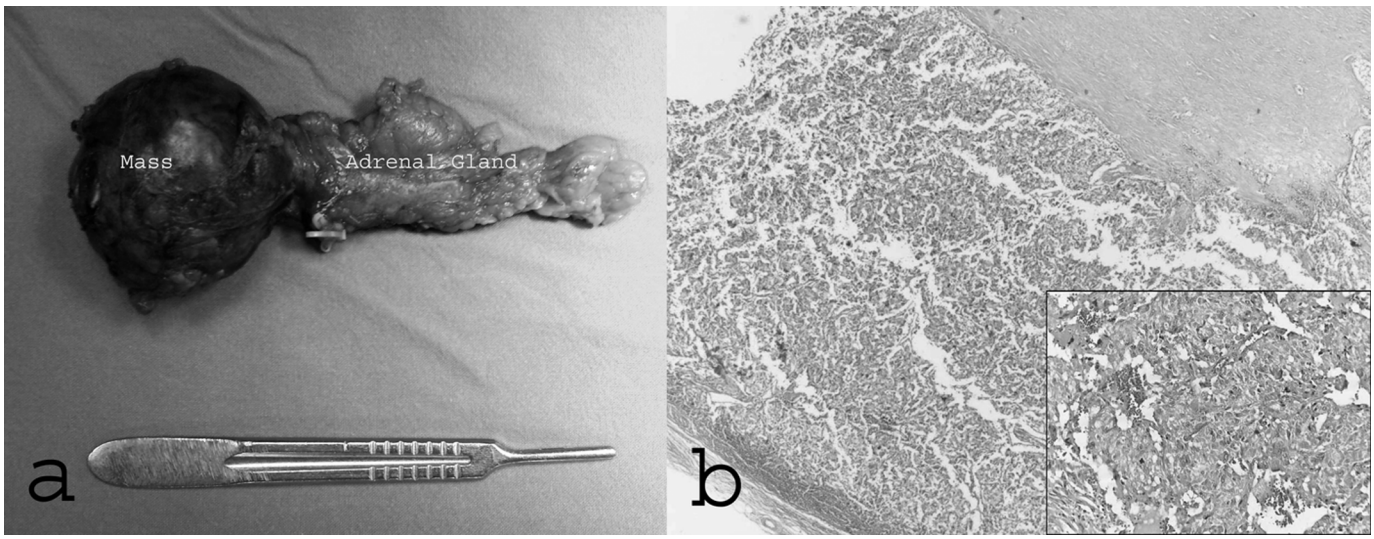

Figure 2. Thin capsulated tumor composed of pleomorphic cells forming nested and trabecular pattern (HE x 200).

DISCUSSION

Pheochromocytomas originate in the neural crest-derived chromaffin cells of adrenal medulla. Approximately 20% of pheochromocytomas are extraadrenal, arising along the migratory pathways of sympathoprogenitor cells, mainly in the abdomen (remnants of the organs of Zuckerkandl, urinary bladder, paraaortic), some in the thorax, and, rarely in the neck.[6] In the international literature, these tumors are frequently called extraadrenal paraganglioma. They often occur in persons 30 years to 45 years of age. Men and women are affected in approximately equal numbers in most series.[6,7] Back pain and palpable mass are the 2 most common presenting symptoms. About 10% of patients present initially with metastatic disease, and about 20% of the tumors are discovered incidentally at the time of autopsy. Symptoms related to production of norepinephrine occur in 25% to 60% of patients with these tumors.[6,8] When our patient's age was compared with the age range of the patients having paraganglioma in general specified in the above-mentioned literature, it was found that our patient was older. Moreover, our patient suffered from one of the most common presenting symptoms, flank pain. Also, our patient's urine and serum biochemical examination results were in normal ranges, and there were no symptoms related to increased catecholamines and their metabolites.

Extraadrenal paragangliomas are rarely diagnosed in the preoperative period unless the lesion is functional. In the latter instance, the diagnosis can be established by measuring 24-hour urinary catecholamines, fractioned plasma catecholamines, and metanephrines.[6] Localization of the mass is accomplished by several diagnostic methods. Computerized tomography (CT) and MRI are highly sensitive for detecting small tumors but are not specific to paraganglioma.[9] Scintigraphic localization of both adrenal and extraadrenal lesions has been accomplished by means of iodine 131 metaiodobenzylguanidine (MIBG), a structural analogue of norepinephrine.[10] MIBG scintigraphy is less sensitive than CT and MRI but is highly specific for paragangliomas and adrenal pheochromocytomas.[11] In our case, we did not perform MIBG scintigraphy due to normal urine and plasma catecholamine levels, and because of MRI findings that mimicked an adrenal hydatid cyst. The diagnosis of the hydatid cyst of the adrenal gland is based mainly on ultrasonography and CT.[12,13] In our case, ultrasonography findings were not consistent with an adrenal hydatid cyst. Additionally, MRI features and a positive cyst hydatid latex agglutination test made us think that the mass concerned was an isolated hydatid cyst. In our case, because the MRI findings were taken into consideration, a CT investigation was not performed.

A prospective randomized study of the role of laparoscopy in an adrenal tumor is not feasible due to the rarity of primary and metastatic adrenal tumors. A recent review[14] stated that metastatic lesions in the adrenal gland are more favorable for laparoscopy than is primary malignant disease. The authors concluded that laparoscopy should be the initial step in localized primary tumors as well as solitary metastasis. Paragangliomas are traditionally resected via open surgery due to their multicentricity, malignancy (29% to 40%), and close relation to these tumors and major anatomic structures.[15] Laparoscopic surgery has recently been used for extraadrenal paragangliomas. In a case report by Brewster and

Sundaram,[16] a 45-year-old male underwent transperitoneal laparoscopic surgery due to a functional 2.7cm in diameter mass at the L2-L3 level without invasion of the aorta or IVC. The mass was excised successfully, and histopathological examination showed an extraadrenal paraganglioma. In the postoperative first month, plasma metanephrine and normetanephrine levels were in normal ranges. The authors suggested that laparoscopy become a viable solution for the treatment of these tumors with fewer adverse effects compared with that of open surgery. Noda and associates[15] reported long-term follow-up of 2 patients who underwent laparoscopic resection of periadrenal paraganglioma. In their case report, the first patient had a functional 50mm in diameter solid mass in the periaortic region above the adrenal vein, and the second patient had a functional 20mm in size solid mass in the upper rear nephric area. The 2 masses were resected laparoscopically without complication. Until now, in addition to case reports as mentioned above, several cases of a laparoscopic resection for paragangliomas have been reported. In those reports, it was stated that extraadrenal paragangliomas could successfully be resected by laparoscopy.[11,17-19] In our case, the periadrenal paraganglioma could successfully and without any complication be resected by laparoscopy. The main difference in our case compared with the other cases mentioned in the literature is that in our case the mass in the MRI findings mimicked an adrenal hydatid cyst. That is why the radiological findings on the MRI of our case showed that paraganglioma could have similar features to those of a hydatid cyst, which is important to be aware of for diagnostic reasons.

CONCLUSION

We argue that the transperitoneal laparoscopic approach for the periadrenal paraganglioma is a safe, applicable method with less morbidity and rapid recovery. We believe that the fact that the paraganglioma can mimic the isolated adrenal hydatid cyst in radiological appearance, as seen in our case, should be taken into consideration.

References:

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

2. Guazzoni G, Montorsi F, Bocciardi A, et al. Transperitoneal laparoscopic versus open adrenalectomy for benign hyperfunctioning adrenal tumors: a comparative study. *J Urol.* 1996;153:1597-1600.

3. Gagner M, Pomp A, Heniford BT, Pharand D, Lacroix A. Laparoscopic adrenalectomy:lessons learned from 100 consecutive procedures. *Ann Surg.* 1997;226:238-247.

4. Imai T, Kikumori T, Ohiwa M, Mase T, Funahashi H. A case-controlled study of laparoscopic compared with open lateral adrenalectomy. *Am J Surg.* 1999;178:50-54.

5. Cobb WS, Kercher KW, Sing RF, Heniford BT. Laparoscopic adrenalectomy for malignancy. *Am J Surg.* 2005;189:405-411.

6. Lenders JW, Eisenhofer G, Mannelli M, Pacak K. *Phaeochromocytoma Lancet.* 2005;366:665-675.

7. Barfield R, Hill DA, Hoffer FA, Tekautz T, Spunt SL. Retroperitoneal paraganglioma. *Med Pediatr Oncol.* 2002;39:120-124.

8. Nakano S, Kigoshi T, Uchida K, Morimoto S, Tsugawa R, Matsunou H. Hypertension and unilateral renal ischemia (Page kidney) due to compression of a retroperitoneal paraganglioma. *Am J Nephrol.* 1996;16:91-94.

9. Sahdev A, Sohaib A, Monson JP, Grossman AB, Chew SL, Reznek RH. CT and MR imaging of unusual locations of extra-adrenal paragangliomas (pheochromocytomas). *Eur Radiol.* 2005;15:85-92.

10. d'Hermobez M, Gouze V, Huglo D, et al. Chromogranin A assay and (131)-MIBG scintigraphy for diagnosis and follow-up of pheochromocytoma. *J Nucl Med.* 2001;42:993-997.

11. Whitson BA, Tuttle TM. Laparoscopic resection of periaortic paragangliomas. *Am Surg.* 2005;71:450-454.

12. Horchani A, Nouira Y, Nouira K, Bedioui H, Menif E, Safta ZB. Hydatid cyst of the adrenal gland. A clinical study of six cases. *Scientific World Journal.* 2006;6:2420-2425.

13. Gharbi HA, Hassine W, Brauner MW, Dupuch K. Ultrasound examination of the hydatid liver. *Radiology.* 1981;139:459-463.

14. Zografos GN, Vasiliadis G, Farfaras AN, Aggeli C, Digalakis M. Laparoscopic surgery for malignant adrenal tumors. *JSLS.* 2009;13:196-202.

15. Noda E, Tetsuro I, Maeda K, et al. Laparoscopic resection of periadrenal paraganlioma:a report of 2 cases. *Surg Laparosc Endosc Percutan Tech.* 2008;18:310-314.

16. Brewster JB, Sundaram CP. Laparoscopic resection of an interaortacaval paraganglioma:diagnosis following a needle biopsy. *JSLS.* 2007;11:502-505.

17. Baghai M, Thompson GB, Young WF, Grant CS, Michels VV, van Heerden JA. Pheochromocytomas and paragangliomas in von Hippel-Lindau disease.a role for laparoscopic and cortical-spring surgery. *Arch Surg.* 2002;137:682-688.

18. Walz MK, Peitgen K, Neumann HP, Janssen OE, Philipp T, Mann K. Endoscopic treatment of solitary, bilateral, multiple, and recurrent pheochromocytomas and paragangliomas. *World J Surg.* 2002;26:1005-1012.

19. Hwang J, Shoaf G, Uchio EM, et al. Laparoscopic management of extra-adrenal pheochromocytoma. *J Urol.* 2004;171:72-76.