Angiomyolipoma in the adrenal region: adrenalectomy converted from laparoscopic to open laparotomy case

Abstract Angiomyolipomas (AMLs) located in the adrenal region are exceptionally rare. The preoperative diagnosis of AMLs is difficult.

We report our experience with a 68-year-old woman who presented with right-sided back pain. A computed tomography scan showed a 66 × 45 mm fatty adrenal mass. Hormonal function was within normal limits. Laparoscopic adrenalectomy was performed. Because bleeding occurred from the tumor surface, we decided on an open conversion and removed the tumor together with a part of the normal adrenal gland. The histopathological features confirmed the diagnosis of AML in the adrenal region. The patient made an uneventful recovery and was discharged one week following the operation.

When we perform laparoscopic adrenalectomy for a fatty adrenal tumor, we must handle the tumor considering the possibility of an AML. Additionally, to reduce excessive bleeding, we must use an open conversion in cases of uncontrollable bleeding.

Key words: adrenal gland, adrenal tumor, adrenalectomy, angiomyolipoma

Introduction Angiomyolipomas (AMLs) are tumor-like lesions most commonly arising in the kidney, that are composed of an admixture of mature adipose tissue, convoluted thick-walled blood vessels, and irregularly arranged sheets and interlacing bundles of smooth muscle. Although uncommon, extrarenal AMLs have been described in various sites as the liver, retroperitoneum, lung, bone, and ovary. AMLs in the adrenal region are particularly rare. To our knowledge, the number of issues surrounding a laparoscopic adrenalectomy for AML in the adrenal region is small. In this report, we describe the difficulty of performing laparoscopic adrenalectomy for AML in the adrenal region and the differential diagnosis of adrenal lesions involving lipomatous component and review the literature.

Case report

A 68-year-old female presented with right-sided back pain. Plain computed tomography (CT) scan conducted at another hospital revealed a well-defined 66 × 45 mm mass in the retroperitoneum. In our hospital, laboratory investigations, including serum catecholamine, cortisol, and urinary vanillylmandelic acid (VMA) were within normal limits. Enhanced abdominal CT and magnetic resonance imaging (MRI) defined the mass as of right adrenal origin. CT attenuation value of the tumor was nearly equal to that of fat tissue (~80 Hounsfield units [HU]). As illustrated in Fig1, the tumor was enhanced in the early phase. Fluorodeoxyglucose Positron emission tomography (FDG-PET) was negative, thereby excluding the possibility of an adrenocortical carcinoma.

We diagnosed the non-functional adrenal tumor containing fat tissue as myelolipoma. Given the size of the tumor, we decided on adrenalectomy as the best course of action. Right laparoscopic adrenalectomy was performed using the lateral transperitoneal approach. First we exfoliated the right kidney and adrenal gland along a layer of Gerota’s fascia. Secondly, we separated the tumor from the upper pole kidney. Bleeding occurred from surface of the tumor while its posterior layer was being separated. The location of bleeding was covered with gauze and the carbon gas pressure was raised to 15 mmHg. After covering the posterior side of the tumor with gauze, we tried to exfoliate the adrenal tumor from liver. Bleeding also occurred from the upper surface of the tumor. We decided on open conversion and removed the tumor together with a part of normal adrenal gland. Total operation time was 293 minutes.
and blood loss was 1430 ml. Sectioning of the tumor revealed a mass that was yellow and homogeneous in texture. Histopathological examination revealed mature fat cells, smooth muscle fibers, and thin-walled blood vessels with peripherally compressed adrenal cortical tissue. Additionally, the immunohistochemical study revealed that epithelioid cells were positive for HMB45 while the immunoreactivity for cytokeratins was negative. These findings suggested this tumor to be an AML located in the adrenal region. (Fig. 2a, b) However it was unclear that the tumor was originated from adrenal gland because normal adrenal gland was too small to evaluate pathologically. The patient could walk on postoperative day 1 and consume food on postoperative day 2. Following an uneventful one week recovery the patient was discharged. There were no signs of recurrence at six months after the surgery.

Discussion

AML is part of a family of neoplasms that derive from perivascular epithelioid cells. Specifically, AML is a rare mesenchymal tumor that is usually found in the kidney. Lam et al.\textsuperscript{9} reported that adrenal AMLs accounted for 0.5% and 0.8% of the primary and surgically resected primary adrenal tumors, respectively. Meanwhile, myelolipomas accounted for 2.6% of the primary adrenal tumors, and liposarcomas accounted for 0.2% and 0.4% of the primary and surgically resected primary adrenal tumors, respectively. Furthermore, teratomas accounted for 1.3% of the surgically resected primary tumors and 0.7% of all primary adrenal tumors. Finally, lipomas accounted for 0.7% of the primary adrenal tumors.

Myelolipomas are usually non-functional unilateral benign lesions with variable mixture of fat and myeloid components.\textsuperscript{6} The presence of gross fat on CT (CT attenuation value of $-30$ HU) is diagnostic.\textsuperscript{5} The pres-

Fig. 1 A right adrenal gland mass. a CT attenuation value of the tumor was $-80$ Hounsfield units (HU) by unenhanced CT. b The enhanced tumor in the early phase.

Fig. 2 Pathological examinations. a Mature fat cells, smooth muscle fibers, and thin-walled blood vessels (hematoxylin-eosin, original magnification $\times 40$). b Epithelioid cells were positive for HMB45 (immunoperoxidase, original magnification $\times 40$).
ence of fat on all MRI sequences is also characteristic of myelolipoma. In this case, CT attenuation value of the tumor was ~80 HU, which was considerably less than the ~30 HU value associated with myelolipomas. This AML was enhanced in the early phase. However, it is difficult to distinguish AML from other lipomatous tumors by merely using imaging methods. Because up to 52% of patients with an AML that is larger than 4 cm in diameter are symptomatic and have an increased risk of bleeding, surgery or selective arterial embolization has been suggested for such cases. In recent years, laparoscopic adrenalectomy has been recommended because it is less invasive compared with open surgery. Management should be the same as that for removal of any adrenal mass. For instance, an assessment of the functional status of the tumor should be conducted. Since the risk of malignancy increases with tumor size, surgery is indicated if the patient is symptomatic or the tumor is more than 5 cm in diameter. The risk of spontaneous rupture also increases with size, owing to the presence of abundant and abnormal elastin-poor vascularity in the tumor. This tumor was maximum length 66 mm. If we had diagnosed the tumor AML presurgically, we decided to remove surgically because of the risk of malignancy and spontaneous rupture in this case.

Successful laparoscopic adrenalectomy has been previously performed on a 5 cm adrenal AML. In this case, as soon as we touched the tumor surface, bleeding occurred. Touching the tumor led to the rupture of vulnerable vessels on the surface of the AML. AMLs can vary significantly according to the percentage of the angiomatous component. We should have to perform the operation more carefully especially in touching tumor surface and exfoliate peripheral right kidney in early stage. However we think that it is not necessary to perform only open laparotomic adrenalectomy in the same cases. It is beneficial in laparoscopic adrenalectomy for avoiding excessive blood loss while exfoliating peripheral regions.

We performed laparoscopic surgery via the peritoneal approach because the retroperitoneal space was too narrow for dissecting a large tumor and limited the mobility of instruments. However, a retroperitoneal lateral approach was selected for small tumors at our institution. Naya et al reported that tumor size significantly correlated with estimated blood loss during surgery. Indeed, larger tumors have a larger surface to dissect and usually have richer vascularization. Avoidance of blood loss and complications when treating larger tumors requires extra caution and time. An AML that is located in the adrenal region is exceptionally rare. Thus, when we perform laparoscopic adrenalectomy for a fatty adrenal tumor, we must be careful to handle the tumor acknowledging the possibility of AML. Additionally, to reduce excessive bleeding, we must use an open conversion in cases of uncontrollable bleeding.

The authors have thier self-reported conflicts of interest to disclose: No

References


