Case Report

A case of adrenal myelolipoma complicated with Prader-Willi syndrome

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Abbreviations & acronyms
CT = computed tomography
MRI = magnetic resonance imaging
PWS = Prader-Willi syndrome

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Introduction: Prader-Willi syndrome is a congenital disorder that occurs in one in 10 000–30 000 children and is characterized by obesity, short stature, and intellectual disability.

Case presentation: A 24-year-old male patient with Prader-Willi syndrome presented with an enlarged adrenal tumor. Computed tomography detected a well-defined mass. Magnetic resonance imaging revealed an increased signal intensity predominantly in fatty areas, suggesting adrenal myelolipoma. Laparoscopic left adrenalectomy was performed. Postoperatively, the patient developed mild pulmonary atelectasis, myelolipoma was confirmed by histopathology, and there was no recurrence at approximately 2 years postoperatively.

Conclusion: This is the first report of Prader-Willi syndrome complicated with adrenal myelolipoma, which was removed laparoscopically.

Key words: adrenal myelolipoma, intellectual disability, laparoscopic adrenalectomy, obesity, Prader-Willi syndrome.

Keynote messages

The case of a 24-year-old man with PWS who presented with an enlarged adrenal tumor is reported. Herein, we describe the successful treatment of adrenal myelolipoma complicated with PWS using laparoscopic left adrenalectomy. This is the first report of PWS and adrenal myelolipoma concomitantly.

Introduction

Prader-Willi syndrome (PWS) is a rare genetic disorder is characterized with obesity and intellectual disability.[1] Generally, adrenal myelolipomas are benign and enlarge slowly,[2–4] but, the long-term natural history is not yet so well understood. In patients with PWS, there are few reports on perioperative management. To our knowledge, this is the first report of an adrenal myelolipoma in a patient with PWS treated laparoscopically.

Case presentation

A 24-year-old man presented with left adrenal incidentaloma. His medical history consists of bilateral orchiopexy and type 2 diabetes at 3 and 14 years of age, respectively. There was no family medical history. An abdominal CT scan revealed an incidental 78 × 70 mm^2 fat-rich neoplastic lesion in the left adrenal gland, upon which the lesion gradually increased, and he was referred for surgery. At admission, his body mass index (BMI) was 41 kg/m^2. Dexamethasone suppression and blood tests led to the diagnosis of a nonfunctioning tumor. Enhanced CT revealed a poor-contrast, 8-cm mass rich in fatty components in the left adrenal gland (Fig. 1a). Abdominal MRI revealed a well-defined mass with fatty components, mostly in the left adrenal gland. Compared with abdominal CT scans performed at other hospitals 10 and 2 years earlier (Fig. 1b,c), the tumor was enlarged, and malignancy could not be

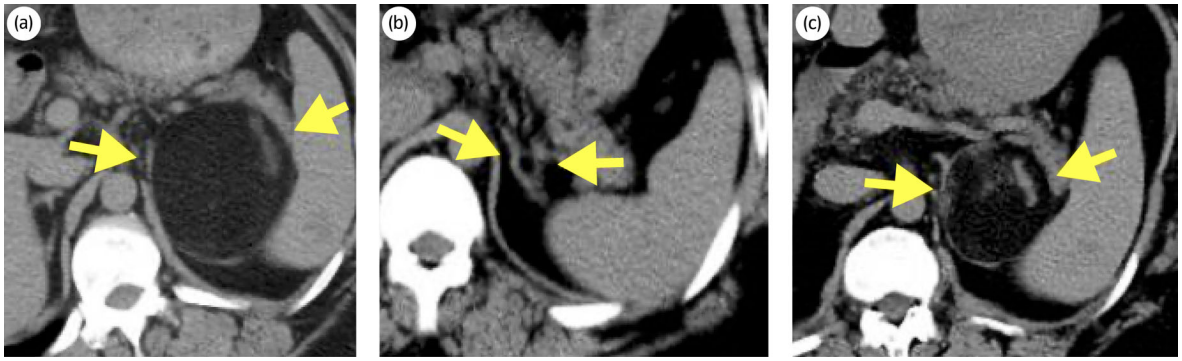
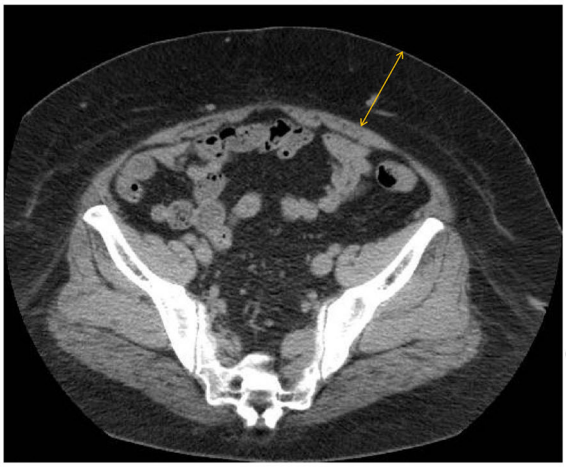


Fig. 1 Abdominal CT scan shows a retroperitoneal tumor of 78 × 70 mm^2, mostly consisting of fatty density areas (a). Changes in tumor size on CT images. (a) Current (78 × 70 mm^2); (b) 10 years ago (6.5 × 6 mm^2); (c) 2 years ago (57 × 45 mm^2).



orange arrow; subcutaneous tissure

Fig. 2 CT findings Axial view. Subcutaneous fat is 40–50 mm thick, indicated by an orange line.

ruled out. Informed consent was obtained from the guardian. A psychiatrist evaluated intellectual and behavioral problems preoperatively and postoperatively, and the patient's family members were required to accompany the patient. A laparoscopic left adrenalectomy was performed (insufflation time: 160 min, blood loss: 20 mL). The operation was performed under general anesthesia. The subcutaneous fat was very thick (40–50 mm) for trocar placement (Fig. 2). Chest radiographs were taken immediately postoperatively, revealing atelectasis in the right lung lower lobe. The anesthesiologist extubated the patient following bronchoscopic aspiration. The patient was admitted to the intensive care unit for postoperative management until postoperative day (POD) 1. A closed drain was placed and removed with a small amount of serous fluid at POD 2. The urinary catheter was removed the next day. The resected specimen was a capsule-covered large mass rich in fatty tissue (Fig. 3a). Histopathologically, bone marrow tissue of three lineages (bone marrow megakaryocytes, erythroblasts, and myeloblasts) and mature adipose tissue was observed; thus, myelolipoma was diagnosed (Fig. 3b). Subsequently, the postoperative course was uneventful, and the patient was discharged on POD 7. CT and ultrasound examinations were performed annually for approximately 2 years postoperatively, and recurrence was not observed.

Discussion

PWS is a rare genetic condition caused by a lack of expression of the paternally inherited chromosome 15q11-q13.[5] It is characterized by cognitive, behavioral, and endocrine abnormalities, growth hormone deficiency, diabetes mellitus, hypogonadism, developmental and mood disorders, and musculoskeletal and nervous system abnormalities.[1,6] Particularly, PWS is a common cause of morbid obesity.[1,7]

Although understanding of PWS has improved, the disease remains life-threatening, and mortality rates are reported to be approximately three times higher than the general population.[8] Hypoxemia and atelectasis were originally common complications after laparoscopic surgery in obese patients.[9] Therefore, respiratory management is important for perioperative management in patients with PWS. Moreover, intellectual disability required perioperative family accompaniment. Similar to PWS, adrenal myelolipoma is associated with a high incidence of obesity.[2,10] Our patient had a high BMI and thick subcutaneous fat, which made the port closure somewhat difficult.

Adrenal myelolipomas are benign, lipomatous tumors with myeloid cell components, the majority of which manifest as adrenal incidentalomas and account for 3.3% to 6.5% of all adrenal masses.[11] Tumors associated with adrenal

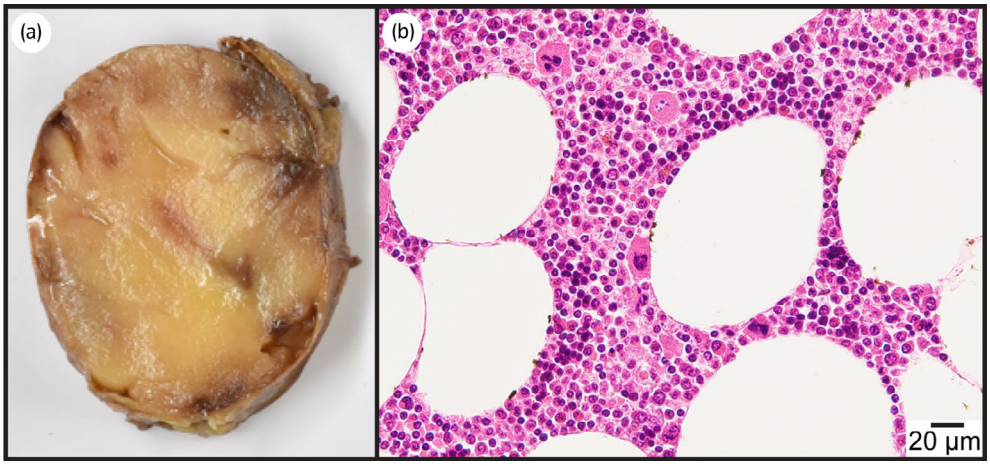


Fig. 3 (a) Macroscopic findings of the resected specimen show a large yellow solid tumor with a normal adrenal gland. (b) Histological examination shows focal hematopoietic elements in mature adipocytes (Hematoxylin and eosin staining, 200×)

myelolipoma range from 0.5 to 18 cm, although the median tumor size is 2–2.5 cm.[12] Adrenal myelolipomas are slow growing, and can be treated conservatively.[2–4] It remains unclear what determines the progression rate; some adrenal myelolipomas grow rapidly after a prolonged incubation period, while others grow slowly on a sustained basis.[2]

In adrenal myelolipoma, minor lesions should be retained. However, patients with large or symptomatic lesions may show new-onset symptoms of mass effect with tumor growth and become candidates for surgery. Although some consider tumors >4 cm to be an indication for surgery, so are symptoms, such as abdominal or lateral abdominal pain, especially in cases of compression of vital structures, such as the inferior vena cava, atypical imaging findings, and tumor size >6–8 cm.[10] Herein, the tumor size gradually increased to 78 mm; therefore, surgery was indicated since the possibility of malignancy cannot be ruled out. Recently, laparoscopic resection was introduced for adrenal myelolipoma management, even in giant masses and obese patients.[10]

Interestinglly, obesity, diabetes mellitus, and hypertension have also been reported to be present in the majority of adrenal myelolipoma,[2,10,13] similar to PWS patients. In our case, morbid obesity and diabetes mellitus due to PWS might be the cause of adrenal myelolipoma. However, the precise mechanisms for both diseases are unclear. The exact pathophysiology and histogenesis of adrenal myelolipomas itself remain poorly understood. Previous papers indicate that various postulates exist, that is, presence of ectopic myeloid tissue, and another hypothesis is that nonrandom X-chromosome inactivation is observed in the majority of adrenal myelolipoma, suggesting clonal development of these tumors.[14] Particularly, the most accepted theory of adrenal myelolipoma is metaplasia of reticuloendothelial cells of blood capillaries as a result of chronic stimulation of adrenals from necrosis, infection, trauma, stress.[4,11] The metaplasia results in cellular differentiation of mesenchymal stem cells into myeloid and adipose tissues.[15] In addiotion, several papers indicated the relationship between PWS and tumors. Davis *et al*. reported that 3% of PWS patients were diagnosed with benign or malignant tumors. Benign tumors included lipomas and fibroma.[16] Apart for myeloid leukemia, malignancy appear to be uncommon in PWS, possibly due to the high prevalence of growth hormone deficiency and low insulin-like growth factor 1 (IGF1).[16,17] There are a few reports of seminoma,[17] Wilms' tumor,[18] liver cancer.[19] The pathogenesis of PWS has yet to be elucidated. As life expectancy increases in the future, it is important to know whether or not neoplastic comorbidities are present. However, it has limitations because it is a single-case report.

Conclusion

This is the first report of a PWS patient who developed adrenal myelolipoma that was removed laparoscopically. Perioperative management of this case required careful consideration of intellectual disability and respiratory complications for the perioperative management of PWS patient.

Author contributions

Toru Inoue: Writing - original draft. Masahiro Todaka: Conceptualization. Yuichi Nakazono: Conceptualization. Yoko Fukata: Conceptualization. Toshitaka Shin: Supervision.

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Conflict of interest

The authors declare no conflict of interest.

Approval of the research protocol by an Institutional Reviewer Board

Not applicable.

Informed consent

Informed consent was obtained from the the family for the publication of this case report.

Registry and the Registration No. of the study/trial

Not applicable.

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