Case Report

Acute hypotension induced by suction of cystic fluid containing extremely high concentrations of catecholamines during resection of giant pheochromocytoma

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Abbreviations & Acronyms
PASS = pheochromocytoma of the adrenal gland scaled score
WHO = World Health Organization

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Introduction: Since pheochromocytomas present with various complications due to catecholamine hypersecretion, their perioperative management needs special attention.
Case presentation: A 45-year-old man visited our hospital with a complaint of abdominal swelling. Radiological and endocrinological assessments determined the tumor as a giant (>20 cm) cystic pheochromocytoma. After administration of doxazosin, the patient underwent radical surgery. Since the tumor was extremely large and fixed to surrounding structures, we punctured it and aspirated cystic fluid to improve the tumor's mobility. However, during the aspiration, the patient developed acute hypotension, which could be reversed by suction withdrawal and vasopressor administration. A similar event occurred during a second aspiration. Eventually, the tumor was successfully excised with negative surgical margin. The cystic fluid proved to contain extremely high concentrations of catecholamines, which might result in the hypotension.
Conclusion: We report the first case who developed acute hypotension due to aspiration of cystic fluid from giant pheochromocytoma.

Key words: adrenal tumor, catecholamine, complication, giant, pheochromocytoma.

Keynote message

Pheochromocytomas present with various complications due to catecholamine hypersecretion, and thus their perioperative management needs special attention. We report a case who developed acute hypotension due to aspiration of tumor cystic fluid during resection of giant pheochromocytoma. The cystic fluid proved to contain extremely high concentrations of catecholamines, which might result in the hypotension. The present report would promote physicians' awareness of this possible intraoperative complication and lead to safer pheochromocytoma surgery.

Introduction

Pheochromocytomas are rare neuroendocrine tumors that originate from the chromaffin cells of the adrenal medulla.[1] Since they present with a variety of complicated and potentially lethal complications due to catecholamine hypersecretion, their perioperative management is recognized as requiring the utmost attention.[2] Morphologically, pheochromocytomas frequently have areas of cystic degeneration as well as hemorrhage and necrosis.[3] We herein describe a patient who experienced acute hypotension by suction of cystic fluid during resection of giant pheochromocytoma. Biochemical examination of the cystic fluid showed extremely high concentrations of catecholamines, which might be the cause of the hypotension.

Case presentation

A 45-year-old, otherwise healthy male visited our hospital with a complaint of progressive abdominal swelling. Radiological assessments including computed tomography and magnetic resonance imaging revealed a giant (>20 cm) cystic adrenal tumor. The tumor compressed right lobe of liver upward, right kidney downward, and inferior vena cava inward, respectively (Fig. 1a). Both serum and urine norepinephrine values showed high scores above normal range (serum: 1423 pg/mL; urine: 569.3 μg/day), whereas serum and urine epinephrine values were slightly high or within normal range (serum: 72 pg/mL; urine: 27.8 μg/day). Comprehensive endocrinological assessments determined the tumor as pheochromocytoma; the tumor was also positive for ^{123}I-MIBG scintigraphy (data not shown).

After administration of doxazosin (gradually increased up to 8 mg/day)[2] and preparative fluid replacement (24 h before surgery), the patient underwent radical surgery, that is resection of the adrenal tumor together with the right lobe of the liver and right kidney. The tumor reached a maximal diameter of 27 cm at the time of surgery and could be identified on the surface of the body (Fig. 1b). In the surgical procedure, right lobe of liver was first resected as planned through a midline incision combined with a right thoraco-abdominal incision (ninth intercostal thoracotomy). We then attempted to mobilize the tumor but could not because of its large size and fixation to surrounding structures. Consequently, we decided to puncture a lateral cystic part of the tumor which strongly compressed the right abdominal wall and aspirate cystic fluid to improve the tumor's mobility. However, during the aspiration with an aspiration tube, the patient's blood pressure suddenly went down to 45/26 mmHg without any sign of bleeding (Fig. 2). Blood pressure soon recovered by withdrawal of aspiration and administration of vasopressor. To improve the tumor's mobility, we punctured another cystic part of the tumor and suctioned the content fluid again. During this second suction, blood pressure decreased to 41/25 mmHg, then recovered with vasopressor therapy. We collected a total of 2450 mL cystic fluid (1250 and 1200 mL in the first and second aspirations, respectively). After these events, the tumor was successfully mobilized and eventually excised together with right lobe of liver and right kidney with negative surgical margin (Fig. 1c) (operation time: 9 h 35 min; total blood loss: 2603 mL; blood transfusion volume: 16 units of red cell concentrates and 8 units of fresh frozen plasmas; total infusion volume: 4700 mL; total tumor weight: 6950 g [including the aspirated 2450 mL fluid]). Pathological diagnosis was pheochromocytoma with a PASS of 15 points (Fig. 1d). Biochemical examination of the cystic fluid demonstrated extremely high concentrations of catecholamines (norepinephrine: 814 500 pg/mL; epinephrine 23 825 pg/mL).

Discussion

While pheochromocytomas are reported to present with various complications in the perioperative period,[2] the present case is the first to develop acute hypotension due to aspiration of cystic fluid of pheochromocytoma. The hypotension occurred reproducibly during a second application of fluid suction and could be reversed by withdrawal of suction and administration of vasopressor. Notably, the cystic fluid was demonstrated to contain extremely high concentrations of

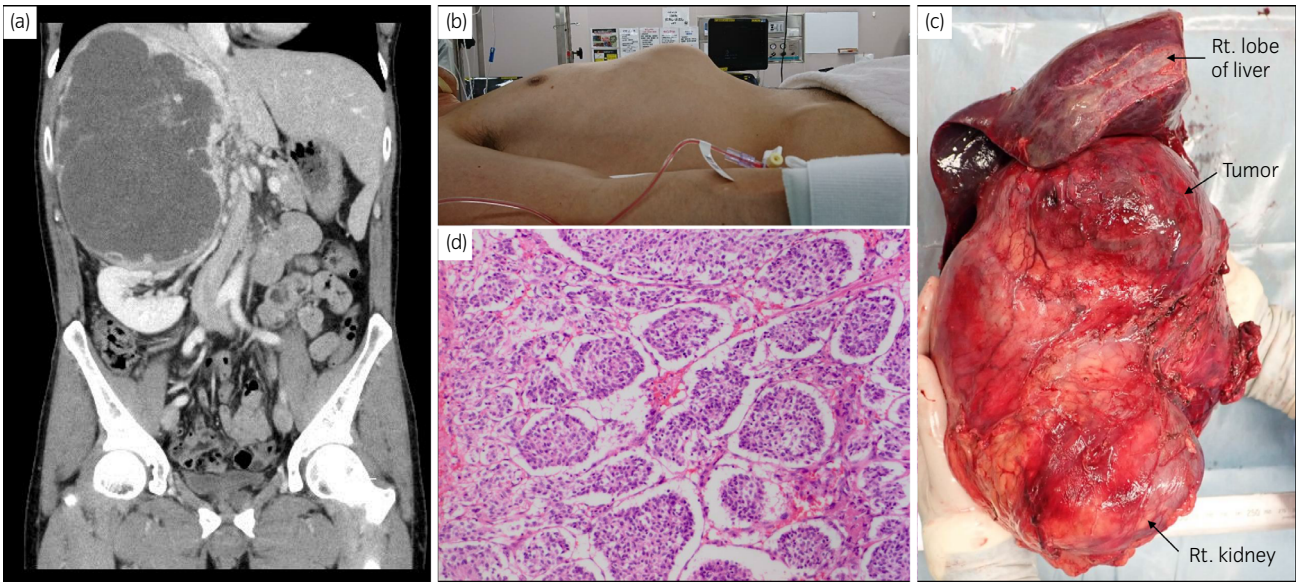

Fig. 1 (a) Preoperative computed tomography demonstrated that a giant right adrenal tumor compressed right lobe of liver upward, right kidney downward, and inferior vena cava inward, respectively. (b) The tumor could be identified on the surface of the body at the time of surgery. (c) A macroscopic image of the removed tumor: it was excised together with right lobe of liver and right kidney (arrows). (d) A microscopic image of the tumor: hematoxylin and eosin staining highlights the "Zellballen" pattern characterized by small nests of tumor cells surrounded by delicate fibrovascular stroma, which is typical for pheochromocytoma or paraganglioma.

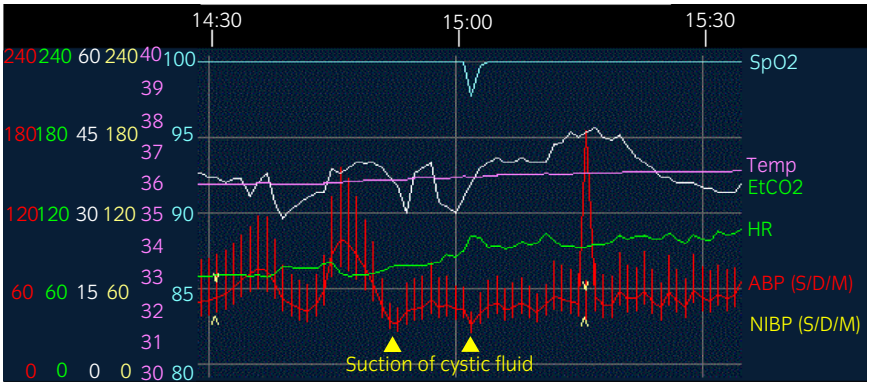

Fig. 2 An anesthesia chart of the present case including the event of acute hypotension due to suction of tumor cystic fluid. Yellow arrowheads indicate timings of the fluid suction.

catecholamines, which might be absorbed via the cystic wall and thus maintained the patient's blood pressure. Another possible explanation would be that aspiration of intracystic hemorrhage simply caused the hypotension, whereas the patient did not present with typical signs for hemorrhagic shock such as tachycardia. Therefore, the extremely concentrated intracystic catecholamines seemed the most likely cause of acute hypotension, despite no proof of decreased levels of circulating catecholamines during the event.

Surgery for the present case was considered challenging because the tumor was extremely large (>20 cm) and adjacent to surrounding organs including the liver, kidney, and inferior vena cava. The tumor reached a maximal diameter of 27 cm at the time of surgery, being one of the largest reported cases of pheochromocytomas.[4–9] Before the surgery, we held a joint conference with related departments (e.g. Departments of Urology, Surgery, Anesthesiology, and Internal Medicine) and prepared for possible perioperative complications. Accordingly, despite an unusual event or the described hypotension, the tumor was successfully excised together with adjoining organs without serious sequela. "Team medicine" is considered essential to performing surgery safely especially in such a complicated pheochromocytoma case.[2]

Currently, all pheochromocytomas are believed to have metastatic potential and thus categorized as malignant tumors by the latest version of the WHO Classification of Tumours of Endocrine Organs (4th ed., 2017).[10] Furthermore, pathological assessments revealed that the present tumor had a PASS of 15 points (PASS ≥4 suggests the malignant potential of a tumor; maximum: 20 points).[3] Therefore, the patient needs careful monitoring for metastasis, despite surgical margin being microscopically negative.

In summary, we have reported a case of acute hypotension by aspiration of cystic fluid during resection of giant pheochromocytoma. This report would promote physicians' awareness of this possible intraoperative complication and contribute to safer surgery for pheochromocytomas.

Conflict of interest

The authors declare no conflict of interest.

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