Clinical Case Report

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A large pheochromocytoma requiring aortic and inferior vena caval reconstruction

A case report

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Abstract

Rationale: It is difficult to discriminate malignant pheochromocytoma (PCC) from benign PCC. The requirement of abdominal aortic and inferior vena cava reconstruction is extremely rare.

Patient concerns: We here report a case of a large pheochromocytoma in a 56-year-old woman who complained of only hand trembling and had no hypertension or other symptoms. The operation was difficult because of a tight adhesion to the circumference of great vessels. A replacement of the aortocaval vessels with grafts was necessary to remove the tumor completely.

Diagnoses: Ultrasonography, computed tomography (CT), and catecholamine assay revealed suspecting the retroperitoneal PCC.

Interventions: Tumor excision and reconstruction of the abdominal aorta and inferior vena cava by externally supported polytetrafluoroethylene (ePTFE) vessels were successfully performed.

Outcomes: A follow-up CT angiography showed no recurrence and graft vessels presented good patency after 7years. Catecholamine in urine and serum assay was normal.

Lessons: The complete resection of the tumor and infiltrated great vessels were necessary for the long-term survival of patients with a large PCC. The ePTFE graft is a good substitute for vessel reconstruction.

Abbreviations: AA = abdominal aorta, CT = computed tomography, IVC = inferior vena cava, PCC = pheochromocytoma.

Keywords: abdominal aorta, catecholamine, inferior vena cava, pheochromocytoma, reconstruction

1. Introduction

Pheochromocytoma (PCC) is a rare tumor, occurring in 3 to 8 per million people with a peak incidence in the third to fourth decade of life.[1] Hormonal hypersecretion in PCC can lead to hypertension, stroke, and even death. Classically, episodic headaches, palpitations, diaphoresis, and anxiety are thought to be hallmarks of a secreting PCC. Nevertheless, these symptoms do not occur in all patients except excessive catecholamines in the circulation,[2] which can sometimes hinder diagnosis. The mainstay of treatment is surgical excision. When a tumor abuts or invades a great vessel, surgical decision-making can be complex. Additionally, the prospective diagnosis of a malignant or benign PCC has been extremely difficult.[3] We here report a rare and complicated case of a large PCC adhering closely to abdominal aorta (AA) and inferior vena cava (IVC), which were simultaneously removed, and 2 great vessels were reconstructed with 2 externally supported polytetrafluoroethylene (ePTFE) grafts. Long-term patency of the aortocaval graft and no recurrence of tumor were confirmed by a follow-up CT angiography (CTA) at 7 years after surgery.

2. Case report

A 56-year-old postmenopausal woman presented with an abdominal mass, without a family genetic history, found by ultrasonography on a staff physical examination in June 2011. CT scan revealed a hypervascular mass located between the aortic bifurcation and the coeliac trunk, with a cross-sectional size of 9.5 × 6.5 cm (Fig. 1). The tumor adhered to the AA and IVC at over 180 degree. Given the complexity of the operation, she attended our hospital and was admitted for the retroperitoneal soft tissue tumor. She complained of only hand trembling and had no hypertension or other symptoms. Laboratory examinations showed a normal complete blood count, normal electrolytes and normal liver function. Urinary excretion of 17-ketosteroids and 17-hydroxycorticosteroids was also within the normal limits, plasma epinephrine and norepinephrine concentrations were 831 ng/L (normal range 54–122 ng/L) and 1046 ng/L (normal range 272–559 ng/L), urinary epinephrine and norepinephrine

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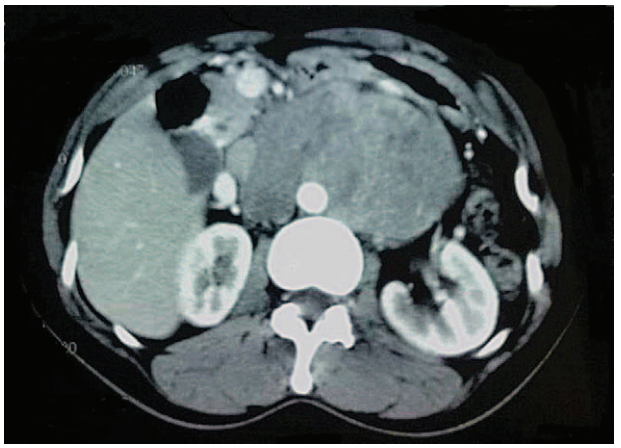

Figure 1. An enhanced computed tomography showed a large retroperitoneal tumor was detected in the 2011 year.

excretions were 427.4 μg (normal range 7.5–21.9 μg) and 526.5 μg per 24 hours (normal 16.3–41.5 μg per 24 hours). Based on these findings, the diagnosis of PCC was established and the patient was prepared for surgery.

After administration of oral phenoxybenzamine (10–20 mg twice a day) and supplementing the circulation volume for 2 weeks, surgery was performed through a midline abdominal incision. Intraoperative exploration confirmed that the mass was stiff, immobilized, and hypervascular on its surface (Fig. 2A). The mass encompassed the side and front of AA and IVC (Fig. 2A), and separating the tumor from AA and IVC was very difficult. Carefully detaching great vessels and tumor from the surrounding organs, especially the left ureter and renal arteries and veins, we simultaneously clamped the infrarenal AA and IVC from the upper and lower polar of the tumor, clamped lumbar arteries and veins in the posterior infiltrated great vessels, and scissored completely the mass including 8-cm segments of AA and IVC. AA was firstly reconstructed with a 14-mm externally supported ePTFE prosthesis (Gore-Tex; W. L. Gore & Associates, Inc., Flagstaff, Ariz.) anastomosing in an end-to-end fashion to the infrarenal and abdominal aortic remnants. After opening artery circulation, the reconstruction of the IVC was also accomplished with a 14-mm externally supported ePTFE graft (Fig. 2B). The tumor measured 12 × 20 cm. Sixteen units of red cell suspension and 1000 mL serum were transfused during the operation.

After surgery, the patient was transferred to the ICU and managed with leg elevation, intermittent mechanical compression of the lower extremities, and anticoagulation with nadroparin calcium. Urine and serum catecholamine returned to normal. The patient was discharged at 36 days after surgery without any complications.

The final pathology demonstrated a PCC originated from paraaortic sympathochromaffin cells that invaded the tumor capsule, surrounding adipose tissue and 1 lymph node. Vascular invasion and distant metastasis were not found. Pathological features of the tumor cells included diffuse growth, spindling, and nuclear pleomorphism (Fig. 3A). Immunohistochemistry examinations showed tumor cells were characterized by the CgA phenotype (+)

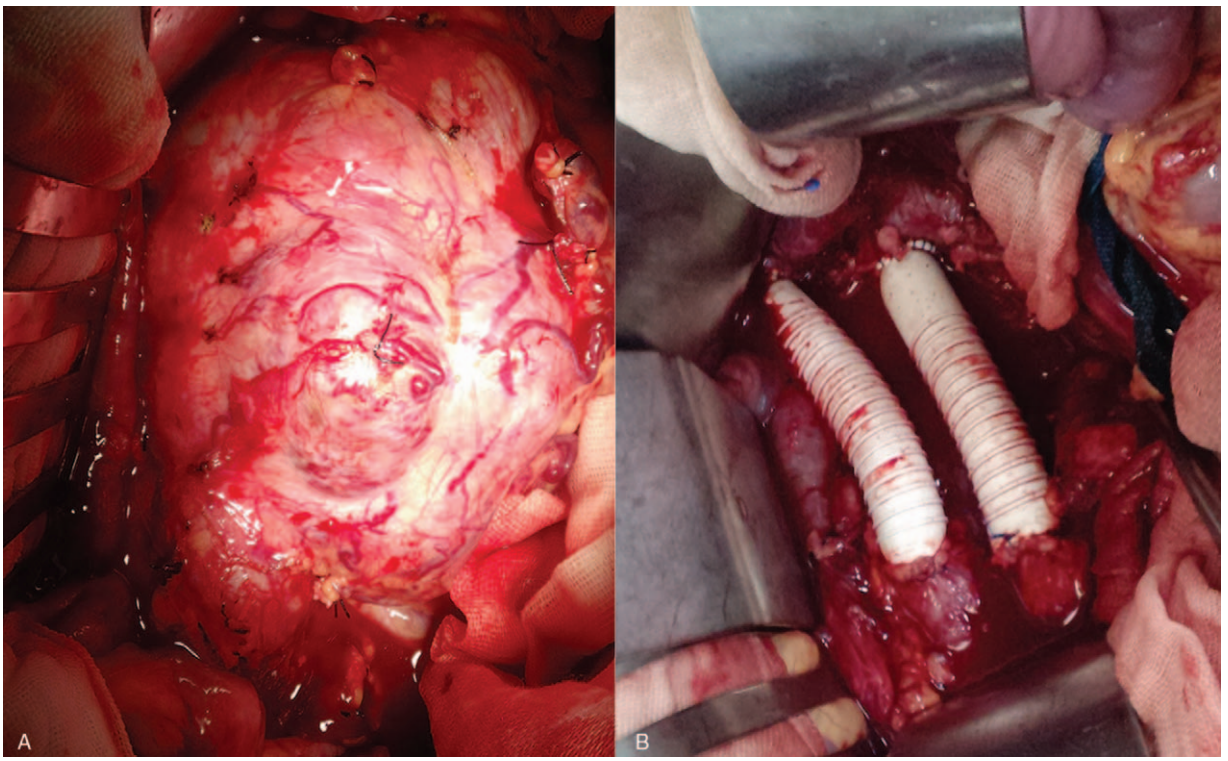

Figure 2. (A) Intraoperative show the tumor had tightly adhered with abdominal aorta (AA) and inferior vena cava (IVC) of around over180 degree, and was hypervascular on its surface. (B) AA and IVC were completely reconstructed with externally supported polytetrafluoroethylene graft.

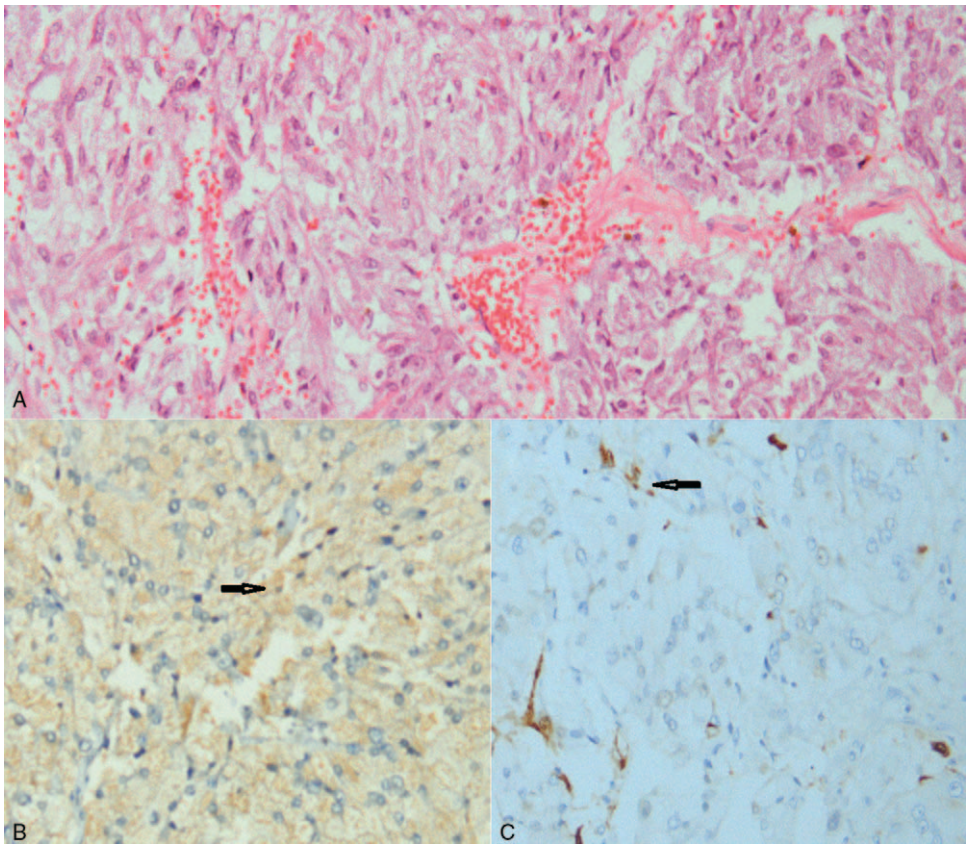

Figure 3. (A) Pheochromocytoma was diffuse growth, spindling, hypervascular, and nuclear pleomorphism. (B) CgA(+) in tumor cells. (C) Partial S-100(+) in the support cells.

(Fig. 3B), calretinin(−), PCK(−), EMA(−), and Ki-67 (positive rate <1%), and support cells presented with S-100 (Partial +) (Fig. 3C). This tumor was confirmed to be a potentially malignant PCC. An enhanced CT scan revealed good patency of aortocaval grafts (Fig. 4A) and no recurrence of PCC was observed at 7 years follow-up (Fig. 4B), except for the right hydronephrosis. Warfarin therapy has been continued since the time of surgery without complications.

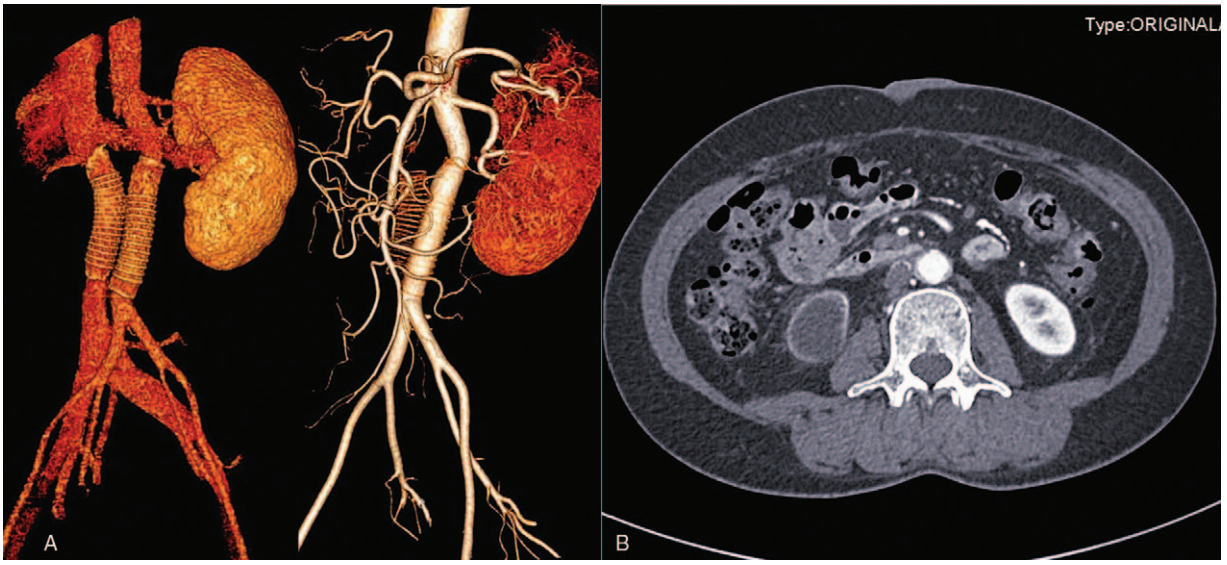

Figure 4. A follow-up computed tomography angiography scan discovered good patency of the aortocaval grafts (A) and no recurrence of pheochromocytoma (B).

3. Ethic statement

The case report was approved by the Ethics Committee of our institution. The patient has provided informed consent to all surgical procedures and clinical research

4. Discussion

The total replacement of the AA and IVC after resection of an extra suprarenal PCC was rarely reported. Additionally, it is difficult to keep good patency of grafts and achieve a long-term survival without tumor. The outcome was closely related to the benignancy or malignancy of the tumor and the success of surgery.

Differentiating benign from malignant PCC was difficult in our case based on current laboratory, imaging, intraoperative, and histological findings. According to the pathological findings, this PCC was stemmed from para-aortic sympathochromaffin cells, which is one of the extrarenal paragangliomas. It was reported that extra-adrenal PCCs are more likely to be malignant (29%–40%) than those found in the adrenal gland.[4,5] Malignant PCC is characterized by the features, such as the presence of large or heavy neoplasms, extraadrenal neoplasms, and high catecholamine secretion in plasma or urine.[5,6] Pheochromocytoma of the Adrenal Gland Scaled Score (PASS) weighting for these specific histologic features can be used to distinguish tumors with a potential aggressive behavior (PASS >4) from tumors that behave in a benign fashion (PASS <4).[3] Our patient had PASS =8: diffuse growth >10% of tumor volume, 2; extension into adipose tissue, 2; tumor cell spindling, 2;capsular invasion, 1; and profound nuclear pleomorphism, 1 (Fig. 3). Our case is a malignant PCC owing to the presence of an extra-adrenal tumor, high secretion of catecholamine, larger size ($20 \times 12 \times 7$ cm), PASS = 8 and 1 local lymph node metastasis. Actually, local tissue invasion or pathological evidence of nuclear pleomorphism or mitotic activity does not necessarily imply malignancy.[3] Distant metastasis is the only reliable criterion for confirming malignancy in the current WHO definition. Therefore, this tumor was confirmed to be a potentially malignant PCC. It was reported that 10% to 30% of patients with pheochromocytoma were found to have metastasis. If PASS reached 7 to 10 points, the metastasis rate was about 77% and a 5-year survival rate was about 22.4% for patients with PCC.

The best treatment for extra-adrenal malignant PCC is completely surgical resection. When a tumor cannot be resected, radiotherapy and chemotherapy have limited effectiveness in the treatment of malignant PCC.[7] However, the removal of PCC involving the AA and IVC was difficult because of the tight adhesion between the mass and great vessels, and surgery should be performed with caution. Rough manipulation inevitably caused cancer cells to fall off and metastasize with the blood circulation. In this case, we simultaneously clamped infrarenal AA and IVC from the upper and lower polar of the tumor and clamped lumbar artery and veins in the involved posterior great vessels.

For decades, various graft materials such as aortic homograft crimped Dacron and Teflon grafts,[8] spiral vein grafts, paneled superficial femoral veil, and bovine or horse pericardium[4,9] have been reported to reconstruct IVC. Moreover, biological materials had lower thrombosis rates than prosthetic grafts in the venous system. Dacron and Teflon grafts had been used simultaneously to reconstruct AA.[4] The lack of a suitable sized autologous conduit has led to the development of spiral graft or panel graft techniques, but uncertain patency rates with these grafts and the complexity of operation has led to less universal acceptance. In addition, Dacron and Teflon grafts are too rigid and lack flexibility. Therefore, the use of externally supported ePTFE for AA and IVC reconstruction is most encouraging up to date.[10] In our case, the14-mm externally supported ePTFE prosthesis completely substituted IVC and AA, and maintained good patency during the 7-year follow-up, which indicated that artificial ePTFE graft is a good alternative for vascular replacement. The patient lived happily and no recurrence was observed. The patient outcome verified that our preoperative decision and surgery were successful.

Author contributions

Conceptualization: Jian Wang, Jichun Zhao.
Data curation: Jian Wang, Ya Lu, Ding Yuan.
Investigation: Jian Wang.
Methodology: Yukui Ma, Bin Huang, Jichun Zhao.
Writing – original draft: Jian Wang.
Writing – review & editing: Jian Wang, Ding Yuan, Bin Huang, Yi Yang, Jichun Zhao.

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