A 34-year-old man presented with monomorphic ventricular tachycardia (VT) accompanying syncope (Figure 1A). Frequent episodes of sustained VT after paroxysmal elevation of heart rate and blood pressure were noted during his stay in the intensive care unit. He had no family or personal history of syncope or cardiac arrest. Coronary angiography and echocardiographic examination revealed no significant abnormalities. Chest and adrenal computed tomography scans revealed a left adrenal mass (7.2×5.3 cm), lymph node enlargement, and several pulmonary nodules (Figure 2A–2D). Elevated levels of 24-hour urinary catecholamines (Figure 3) and abnormal uptake on I-123 metaiodobenzylguanidine (MIBG) imaging (Figure 2E) suggested a malignant pheochromocytoma with lymph node and pulmonary metastasis.

Frequent episodes of pulseless VT (Figure 1B) were noted even after the administration of an α-blocker (phenoxybenzamine 10 mg twice daily) and an antiarrhythmic (amiodarone, 300 mg IV followed by a maintenance dose of 1 mg/min for 6 hours and then 0.5 mg/min for 18 hours). However, the VT storm began to be controlled by the addition of a β-blocker (metoprolol 50 mg twice daily) and unilateral adrenalectomy. Histopathological examination finally confirmed a malignant pheochromocytoma (Figure 4). Although the oral β-blocker was skipped because of fasting on the day after the unilateral adrenalectomy, pulseless sustained VT recurred (Figure 1C). The patient underwent cardioverter-defibrillator implantation and was discharged in stable condition with amiodarone (200 mg once daily), metoprolol (50 mg twice daily), and low-dose phenoxybenzamine (10 mg twice daily).

On postoperative day 28, we reduced the amount of metoprolol to 50 mg once daily because of patient’s frequent dizziness and low blood pressure, although his urinary catecholamine levels were still above the normal range. However, after the dose of metoprolol was reduced, he was readmitted because of repetitive implantable cardioverter-defibrillator shocks triggered by recurrent VT storm (postoperative day 38; Figure 1D). Another region of focal MIBG uptake in the right anterior lung field was detected (Figure 2F, arrow) on follow-up scanning. We increased the dose of metoprolol (up to 75 mg twice daily) and added iodine-131-MIBG therapy for metastatic lesions. About 7 weeks after the iodine-131-MIBG therapy was started (postoperative day 156), the patient’s urinary catecholamine levels had normalized (Figure 3) and the VT recurrence had resolved. Pheochromocytoma frequently presents as various cardiovascular manifestations, such as paroxysmal or persistent hypertension, sinus tachycardia, paroxysmal supraventricular tachycardia, atrial fibrillation, and ventricular premature contractions. However, the presence of sustained VT has rarely been reported.1–3 Several mechanisms of VT associated with pheochromocytoma have been proposed, including catecholamine release from the pheochromocytoma, QT prolongation, and neuropeptide Y.2–4 However, our case suggests the role of catecholamine release, especially β-adrenergic hyperstimulation, in the development of VT in pheochromocytoma more clearly than the previous reports; frequent episodes of VT were almost always preceded by a period of vital sign fluctuation (elevated heart rate and blood pressure). The VTs were not controlled by α blockers or amiodarone alone, but were suppressed by adrenalectomy with the addition of a β-blocker. The VT events recurred when the β-blocker was stopped or reduced. Finally, it is unlikely that the patient had a preexisting arrhythmogenic substrate because his heart was structurally normal.

Unlike previous reports,1,2 our patient experienced VT recurrence even after the tumor was removed, and this recurrence could have been due to regrowth of the pulmonary metastatic nodule. This being the case, we were reluctant to discharge the patient without implantation of a defibrillator.

Disclosures
None.

References

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Figure 1. A, The initial ECG showed very rapid monomorphic ventricular tachycardia (VT) with right bundle-branch block morphology and left-axis deviation. B, Nonsustained and sustained ventricular tachycardia recurred although the patient was treated with an \( \beta \)-blocker. C, Pulseless sustained VT accompanied by syncope developed when the oral \( \beta \)-blocker was skipped immediately after the adrenalectomy. D, Implantable cardioverter-defibrillator interrogation showed successful termination of fast VT by appropriate implantable cardioverter-defibrillator shock.
Figure 2. A and B, Adrenal computed tomography scans show a hypervascular mass with central necrotic change involving the left adrenal gland (arrowhead) with lymph node metastasis in the left para-aortic space (arrow). C and D, Computed tomography scans show multiple metastatic pulmonary nodules in both lungs. E, The I-123 metaiodobenzylguanidine scan shows several areas of abnormal uptake in the left upper posterior abdomen (arrowhead) and in the right lung field (arrow). F, Another region of focal uptake is visible between the previous lung lesions on the follow-up scan.

Figure 3. Changes in the levels of urinary catecholamine and its metabolites. Before the adrenalectomy, urinary concentrations of normetanephrine (>2000 \( \mu \)g/24 h; reference range, 82–500 \( \mu \)g), vanillylmandelic acid (VMA; 25.9 mg/24 h; reference range, \( \leq 9 \) mg), and norepinephrine (>2000 \( \mu \)g/24 h; reference range, 15–80 \( \mu \)g) were significantly elevated. They began to decrease after removal of the mass and returned to within the normal ranges ~6 weeks after I-131 metaiodobenzylguanidine (MIBG) therapy. POD indicates postoperative day.
Figure 4. A, The specimen consists of the 12×7×3-cm left kidney (arrow) and a well-capsulated 8×8×3-cm tumor (arrowhead). The cut surface of the tumor shows a bright yellowish to tan sclerotic lesion encompassing the entire adrenal gland. B, The tumor cells have a finely granular basophilic cytoplasm with round to ovoid nucleoli (hematoxylin and eosin stain, ×400). C and D, Immunohistochemical staining shows strong cytoplasmic immunoreactivity to synaptophysin (×200) and chromogranin (×200), respectively.
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