

VENTRICULAR TACHYCARDIA AND SPONTANEOUS RUPTURE OF AN EXTRA-ADRENAL PHEOCHROMOCYTOMA CASE

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To the Editor:

In their article, "Extra-adrenal Pheochromocytoma Presenting with Life-threatening Ventricular Tachycardia: A Case Report", Li et al discussed an unusual case of pheochromocytoma with the presentation of life-threatening ventricular tachycardia (VT) [1]. We have some concerns about this case report.

This patient was first admitted to our cardiology ward due to syncope and had undergone detailed diagnostic examinations and intensive preoperative treatment before transfer to the urology ward for surgery. The following clinical presentations need to be addressed further.

First, this 64-year-old female had had hypertension for 2–3 years. When syncope occurred during admission, the associated ventricular arrhythmia was monomorphic rapid VT rather than Torsade de pointes. Electrophysiologic study revealed normal sinus node and arteriovenous nodal function and could not induce any significant VT. In addition, there was no significant organic heart disease, which may indicate that VT was not due to a reentry mechanism. Diaphoresis, headache, and high blood pressure are usually noted before a VT attack. Surface electrocardiography (ECG) showed a normal sinus rhythm with diffuse T-wave inversion from V2 to V6. The QT interval was prolonged with QTc of 503 ms (Figure 1A). All clues were highly suggestive of catecholamine-dependent VT. Twenty-four-hour urinary vanillylmandelic

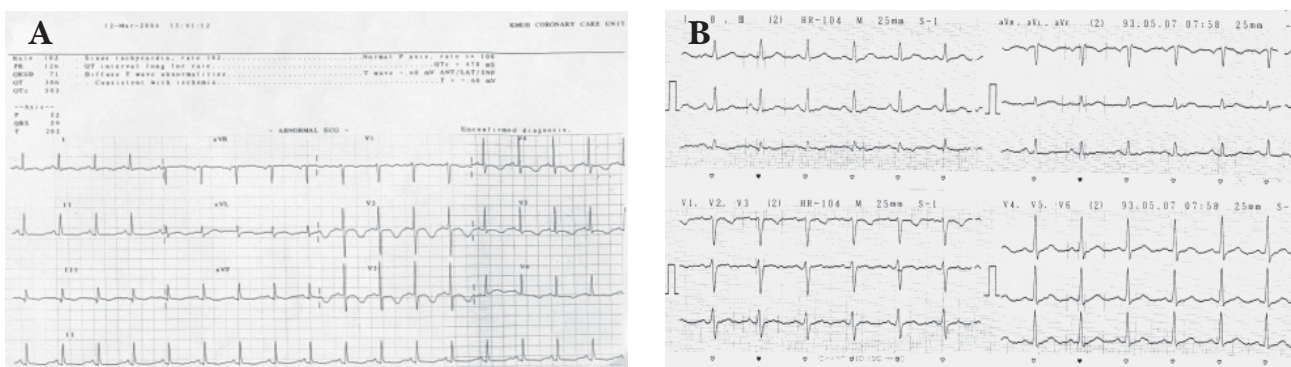


Figure 1. (A) 12-lead electrocardiogram showing diffuse T-wave inversion and prolongation of QT interval. (B) These findings disappeared after removal of the paraganglioma.

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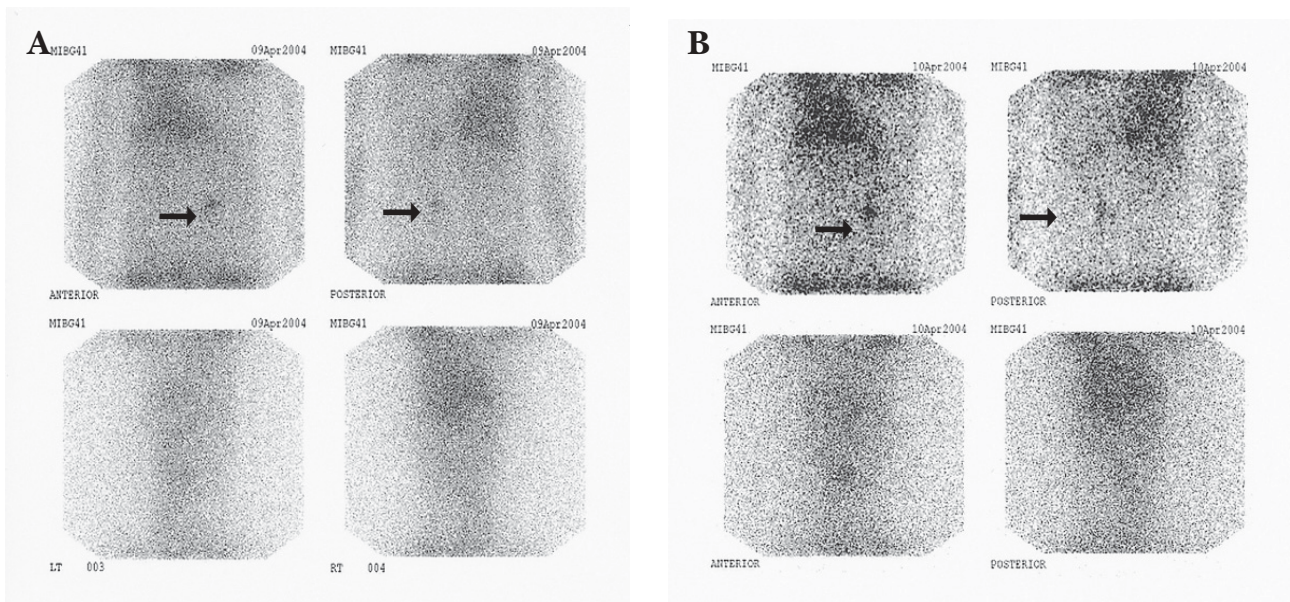


Figure 2. ^{131}I -meta-iodobenzylguanidine scan showing accumulation of radioiodine in the left upper quadrant and left middle portion of the abdomen throughout the serial 24-, 48-, and 72-hour images. Arrows indicate the region of the paraganglioma.

acid was greater than 35.8 mg/day (normal, 1–10 mg/day). ^{131}I -meta-iodobenzylguanidine scan revealed radioiodine accumulation in the left upper quadrant and left middle portion of the abdomen, which confirmed the diagnosis of paraganglioma (Figure 2). The paraganglioma was located at a relatively rare site (the inferior para-aortic region) at surgery, which was compatible with the imaging findings described. The diagnosis was further substantiated by the postoperative pathology. Brilakis et al reported a 51-year-old woman with pheochromocytoma who presented with QT prolongation and Torsade de pointes [2]. In our case, after removal of the paraganglioma, blood pressure returned to normal without any antihypertensive treatment. VT and syncope disappeared during follow-up. The QT prolongation and diffuse T-wave change on the ECG also became normal 1 month after surgery (Figure 1B).

Second, the internal bleeding episode due to spontaneous rupture of the paraganglioma confirmed by surgical findings was another rare presentation. Sapienza et al reported a case of retroperitoneal hemorrhage due to spontaneous rupture of a right adrenal pheochromocytoma, presenting as an acute abdominal emergency with symptoms of peripheral vasoconstriction [3]. Forty and Dale presented a case of a pheochromocytoma with hemorrhagic necrosis resulting in signs of acute abdomen with shock [4]. The classical signs of ruptured pheochromocytoma are intense vasoconstriction with pale extremities, tachycardia, and

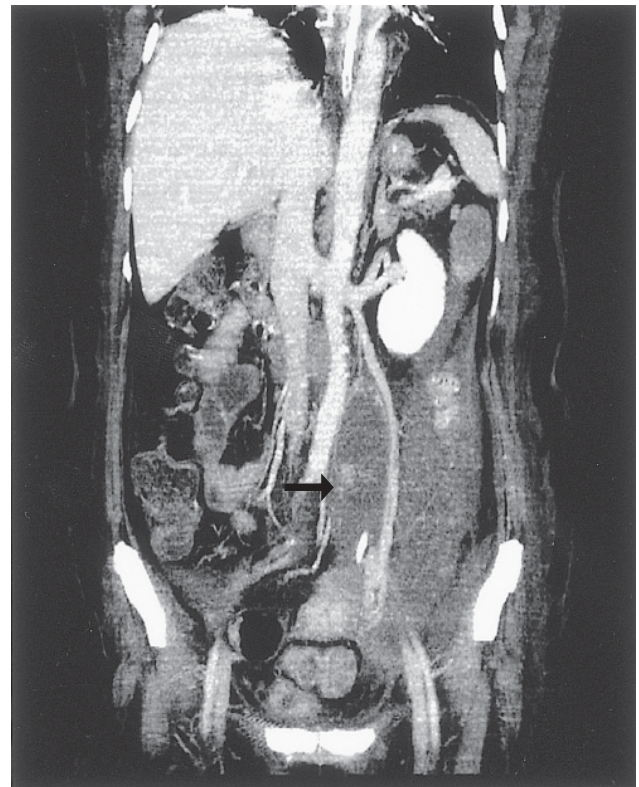


Figure 3. Abdominal computed tomography after reconstruction. A cystic paraganglioma in the left anterior pararenal space with focal contrast extravasation at the left L5–S1 level and a hematoma below the left L4–5 level are present. The arrow indicates the region of the ruptured cystic paraganglioma with contrast extravasation.

high blood pressure, which were present in our patient in the coronary care unit. The major differential diagnosis was leaking abdominal aortic aneurysm. The key feature of labile blood pressure can differentiate pheochromocytoma from leaking abdominal aortic aneurysm. However, abdominal computed tomography was the best examination for accurate diagnosis in this case (Figure 3).

In conclusion, in a hypertensive patient with the typical triad of palpitation, headache and sweating, and specific ECG findings of QTc prolongation and diffuse inverted T-wave changes, a chromaffin-cell tumor should be taken into consideration in the differential diagnosis.

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