

## What a Headache

### Rare Neuroendocrine Indication for Cardiopulmonary Bypass for Severe Left Ventricular Dysfunction and Shock

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A 45-year-old male presented with 2 days of nausea, sweating, and abdominal pain. Examination revealed tachycardia, hypertension, diaphoresis, widespread crepitations, and diffuse abdominal tenderness. Profound hypotension developed, despite intravenous fluids, and was treated with noradrenaline and dobutamine; hypoxia required endotracheal intubation, followed by chest radiograph, which demonstrated extensive pulmonary edema. Echocardiography revealed severe global left ventricular systolic impairment, with an estimated ejection fraction of only 10% (Data Supplement Movies I and II). The patient was transferred to our hospital for the consideration of intraaortic balloon counterpulsation or left ventricular assist device support or both. An intraaortic balloon pump was inserted, and inotropic support was changed to adrenaline with modest improvement. Examination revealed a large mobile nonpulsatile mass in the left paraumbilical region, confirmed as a paraganglionoma on computed tomography (Figure 1).  $\alpha$ -Blockade with phentolamine was commenced because an irreversible agent, such as phenoxybenzamine, was unsuitable in an unstable patient. Unopposed  $\alpha$ -blockade resulted in severe reflex tachycardia, and  $\beta$ -blockade with intravenous esmolol precipitated critical hypotension. Recurrent atrial fibrillation and worsening pulmonary edema indicated that the patient was unlikely to survive with conservative treatment; yet, the risks of conventional surgical resection without autonomic blockade were considered prohibitive. The only option for a successful outcome was to provide the patient with an artificial circulation and remove the tumor. Femoro/femoral bypass was rejected as this form of support would be unable to provide for the potential need for very high flow rates. The patient underwent cardiopulmonary bypass following a midline sternotomy. The heart was left beating, and moderate hypothermia was achieved. Laparotomy confirmed a large tumor adherent to a left sided inferior vena cava. It was removed without complication while the patient was maintained on bypass by using a phentolamine and esmolol infusion throughout (Figure 2). The patient was weaned from cardiopulmonary bypass with adrenaline support without complica-

tion at the first attempt, and following rapid improvement, he was discharged from hospital 10 days after surgery. Echocardiography before discharge showed an estimated ejection fraction of 40% (Data Supplement Movies III and IV). Urine analysis collected preoperatively showed markedly elevated normetadrenaline and metadrenaline levels. The patient subsequently confirmed a history of several months of severe headaches lasting 5 minutes, along with intermittent palpitations, and the feeling his head was going to explode, which usually occurred when bending over to lace his boots in the morning. Histology of the tumor confirmed a paraganglionoma with extensive ischemic necrosis.

#### Discussion

Phaeochromocytoma are rare catecholamine-producing tumors, typically presenting with headache, sweating, palpitation, and hypertension. Eighty-five percent arise from the adrenals, and when they arise outside the adrenals, they are termed paraganglionomas. Recognized cardiovascular complications include sudden death, myocardial infarction, heart failure, hypertensive encephalopathy, and cardiogenic shock<sup>1</sup> including fatal cardiomyopathy.<sup>2</sup> The mechanism underlying the impaired ventricular function is unclear and may be because of a tachycardia-related cardiomyopathy, ventricular hypertrophy as a result of systemic hypertension, or a direct effect of catecholamines on cardiac myocytes. Following diagnosis, the challenge is to stabilize the patient with autonomic blockade to allow safe surgical removal of the tumor. Given the presenting symptom of abdominal pain with hypertension and diaphoresis on arrival followed by cardiogenic shock, it is likely that the acute event was infarction of part of the phaeochromocytoma, as suggested on histology with a surge in catecholamine levels and acute left ventricular stunning.

Phaeochromocytoma should be considered in patients presenting with heart failure and cardiogenic shock and no other obvious diagnosis.<sup>3</sup> Removal of the tumor may lead to rapid reversal of catecholamine-induced cardiomyopathy. Extracorporeal membrane oxygenation or left ventricular assist

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The online-only Data Supplement can be found at <http://circheartfailure.ahajournals.org/cgi/content/full/1/2/143/DC1>.

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(*Circ Heart Fail.* 2008;1:143-145)

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DOI: 10.1161/CIRCHEARTFAILURE.108.766865

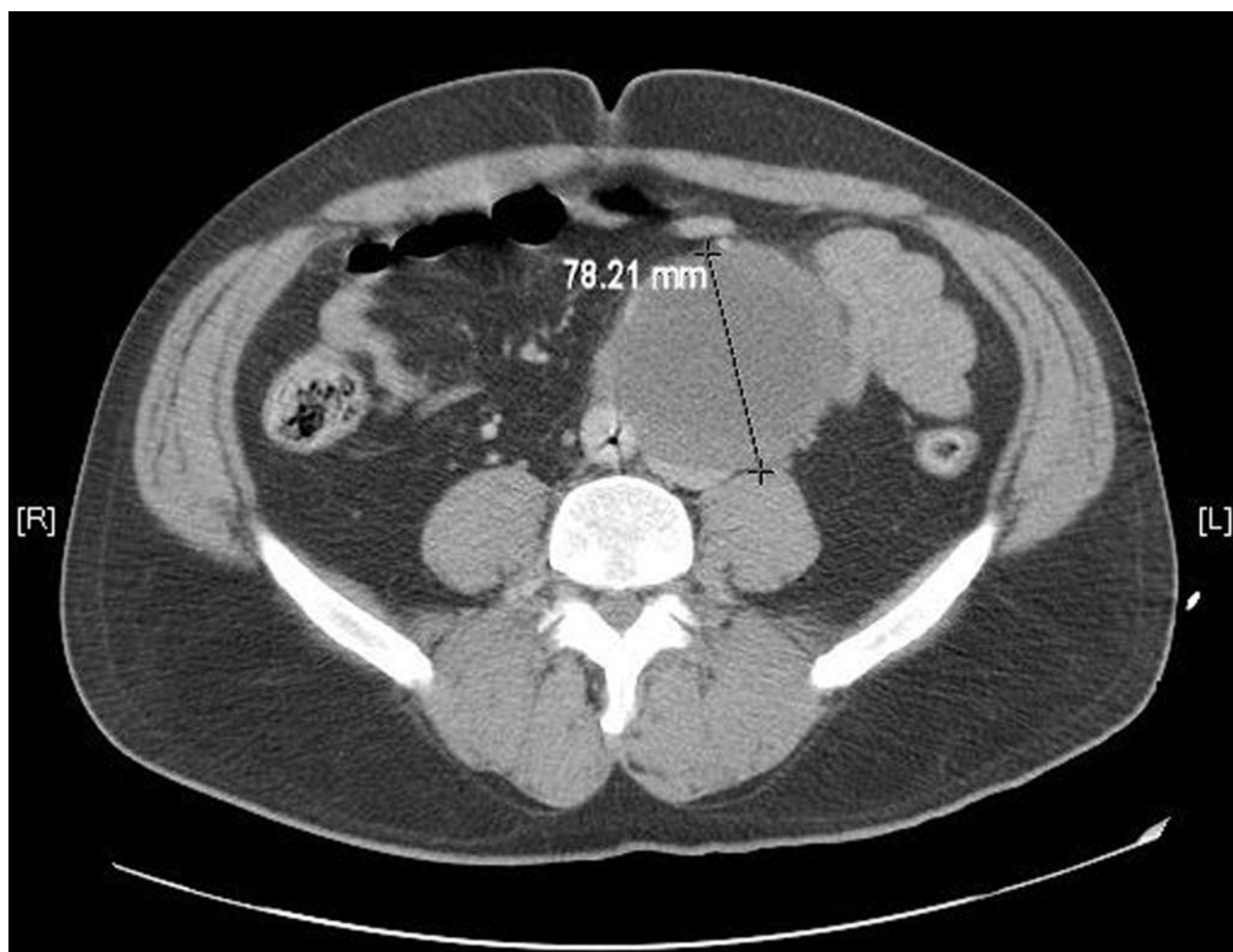
devices<sup>4</sup> have been used to support surgery. We report the use of cardiopulmonary bypass to facilitate surgical removal in a critically ill patient with severe cardiac dysfunction.

### Disclosures

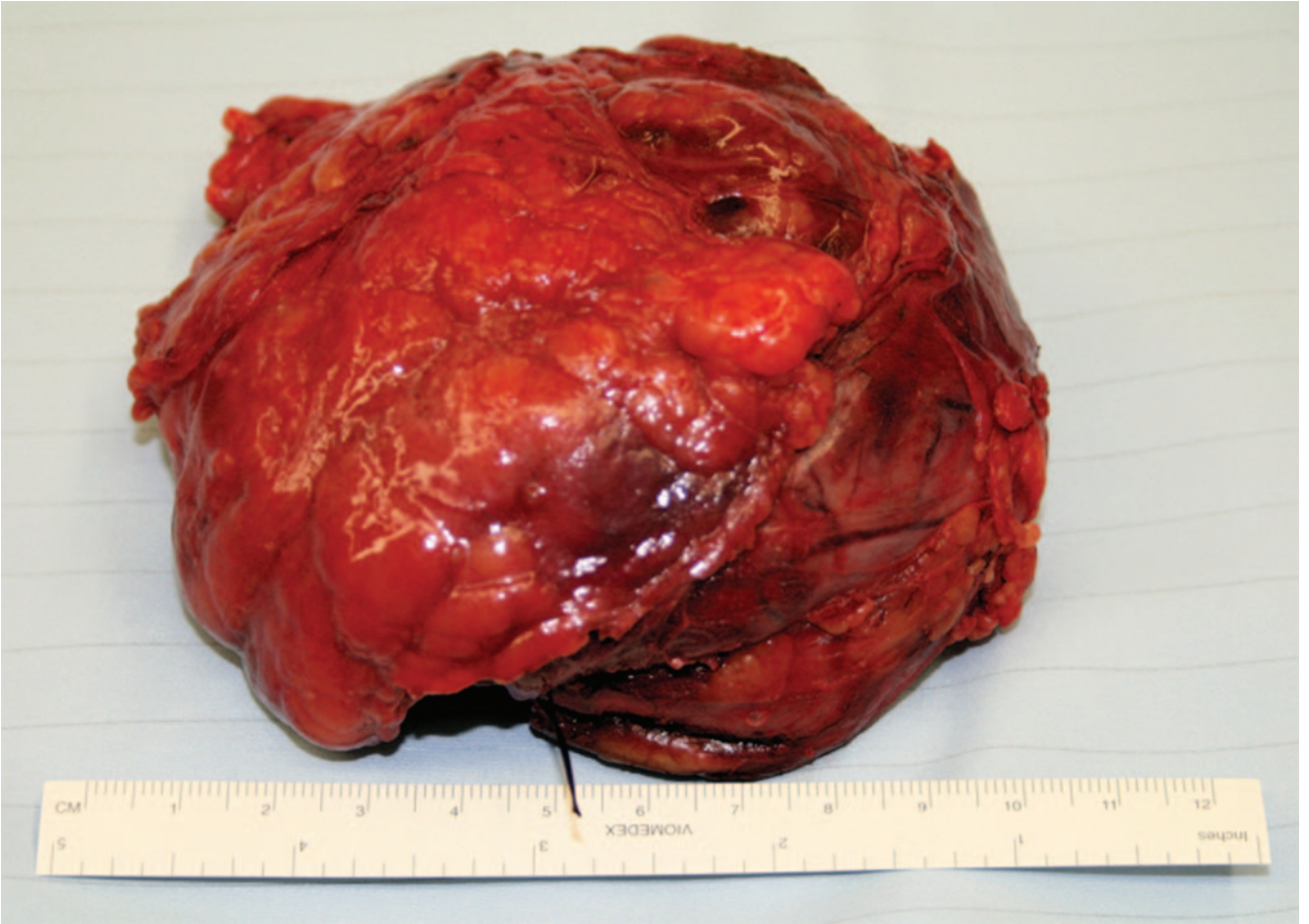
None.

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**Figure 1.** Computed tomography of the abdomen showing a mass overlying and compressing a left-sided inferior vena cava measuring 7.8 cm in diameter. An artifact from the intraaortic balloon pump in the aorta was noted.



**Figure 2.** Operative specimen.

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doi: 10.1161/CIRCHEARTFAILURE.108.766865

*Circulation: Heart Failure* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

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Print ISSN: 1941-3289. Online ISSN: 1941-3297

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