



Pheochromocytoma Presenting as Acute Cardiomyopathy and Multiorgan Failure in a Young Lady-Think Twice before Sending a Patient for Heart-Transplantation

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Abstract

We illustrate a case of pheochromocytoma-related cardiomyopathy in a young lady who presented with acute decompensated cardiomyopathy. Initially it was thought to be caused by mumps infection and heart-transplantation was planned. Through a collaborative effort among different specialists, the correct diagnosis was eventually reached. A novel management pathway lining up pre-operative embolization of tumor followed by adrenalectomy with ECMO was proven to be successful.

Introduction

Pheochromocytoma is a rare neuroendocrine tumor arising from the adrenal medulla chromaffin tissue, which secretes catecholamines. The annual incidence of this tumor is ~0.8 per 100,000 person-years [1], although a significant number of cases are only identified during autopsies [2]. It often poses as a diagnostic challenge to physicians since clinical manifestations range widely from asymptomatic to paroxysmal hypertension, headache, palpitation or even cardiomyopathy and multiorgan failure. We describe a case of a young lady who presented with severe acute cardiomyopathy and multiorgan failure. The patient was initially planned for heart-transplantation and later regained near-normal myocardial function when a pheochromocytoma was correctly identified, embolized and resected.

Case Report

A 29-year-old woman presented with headache, palpitation and vomiting with an ECG showing supraventricular tachycardia with a heart rate of 149 beats per minutes was admitted directly from the emergency department to the intensive care unit and was started on Extracorporeal Membrane Oxygenation (ECMO) as life sustaining measure for cardiogenic shock with multiorgan failure. Significant past medical history was an episode of myocarditis a few years ago labeled to be caused by a viral infection, from which she regained full cardiac function. Extensive workup for this second episode of

acute cardiomyopathy included a myocardial biopsy, which only revealed prominent fat cell infiltrate resulting from the previous myocarditis with focal cardiomyocyte injury and a positive mumps Immunoglobulin M (IgM) antibodies in serology test. Echocardiogram showed a left ventricular ejection fraction of only 10% with global hypokinesia, mild mitral regurgitation and no vegetation. Therefore, an initial diagnosis of “mumps-related myocarditis” was made. The patient was therefore planned to be put on the heart transplantation waiting list.

Upon enrollment with the heart-transplantation team, a specific request to rule-out a pheochromocytoma was made based on her young age and recurrent cardiomyopathy history. Endocrinologists were consulted for this pursuit. Since the patient was anuric with acute renal failure, the usual screening test for pheochromocytoma employed in our institute, 24-hour-urine catecholamines, was not feasible. Plasma metanephrines

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were assessed instead. The results showed sky-high readings (Metanephrine 12932 pmol/L, reference-range 40-450 pmol/L, Normetanephrine 4327 pmol/L, reference-range 110-740 pmol/L). Serum Chromogranin A, a neuroendocrine tumor marker, came back to be very high as well (2961 ng/mL, reference-range 27-94 ng/mL). A subsequent abdominal Computer-Tomography (CT) with contrast revealed a well-defined 7 cm right adrenal mass with heterogeneous density with mild contrast enhancement. The diagnosis of a right “pheochromocytoma-related cardiomyopathy” was made based on the above investigation results. The patient was started on phenoxybenzamine as alpha-blockade and later, on esmolol as beta-blockade.

The overall plan was to remove the tumor as soon as possible in view of the tremendous adverse effects resulting from excess catecholamines. However, compared with our other patients with pheochromocytomas, this patient was extremely frail, relying on ECMO for life maintenance. Thus, she would unlikely survive a hypertensive crisis during the adrenalectomy, when there runs a risk of catecholamines/blood pressure surges during manual manipulation of the pheochromocytoma. A joint meeting involving endocrinologists, surgeons, interventional-radiologist, anesthetists and intensivists was held. The collective decision was to first reduce the blood supply to the very vascular right pheochromocytoma by embolizing the supplying vessels. This would reduce the risk of hemodynamic instability intraoperatively. An uneventful right adrenal and accessory right renal arteries embolization with 100 micron microspheres was completed via a right femoral approach with near complete occlusion achieved. Serum creatinine was 127 umol/l prior to the embolization procedure and subsequently elevated to 149 umol/l immediately after the procedure. A follow-up CT abdomen with contrast showed a wedge-shaped hypo enhancing areas in the right kidney probably representing ischemia and/or infarcts. A successfully right adrenalectomy was performed one day later. There was no hypertensive crisis during the operation.

The patient then gradually regained near-normal cardiac and renal function and ECMO was weaned off. Histology revealed an encapsulated tumor measuring 7 cm in diameter, with most of the tumor being necrotic, which was compatible with the previous embolization. Immunostains confirmed the diagnosis of a pheochromocytoma. Post-operatively, there was a substantial reduction in plasma metanephrines (metanephrine 397 pmol/L, normetanephrine 1013 pmol/L). Post-op radioisotope scan had been arranged as follow-up investigations which did not show any residual pheochromocytoma. Gratitude had been expressed to the heart-trans-

plantation team for preventing this lady from having an unnecessary heart-transplantation with the smart prerequisite request.

Discussion

Pheochromocytomas are often only identified upon autopsy of sudden death cases. An Australasian group reviewed more than 40000 coronial autopsy records over a 16-year period. They reported that clinicians fail to diagnose a significant number of pheochromocytoma (incidence of 0.05%) during life and this tumor may have contributed to up to 50% of the patient deaths [2]. Our case illustrates that high clinical suspicion in the setting of unusual recurrent cardiomyopathy in a young patient, coupled with sharp clinical acumen in working through the list of differential diagnoses meticulously, resisting to take the most obvious “mumps myocarditis as supported by positive serology result” to be the final answer, were invaluable attributes required of clinicians in saving this patient from an unnecessary heart-transplantation. Particularly, mumps IgM antibodies are non-specific and can be false-positive in up to ~6% of serum from normal subjects and patients recovering from other viral infections [3].

Acute cardiomyopathy in patients with pheochromocytoma is rare. Only 10% of the patient population presents with catecholamine-induced cardiomyopathy [4]. Upon autopsy, the heart weight is found to be increased in 95% of the patients [2]. Upon blockade of the catecholamine actions either medically, with alpha and beta-blockers or surgically, with tumor removal, cardiac function is usually reversible [5,6]. In addition to pheochromocytoma-related cardiomyopathy, Takotsubo cardiomyopathy and myocarditis are two other common differential diagnoses in patients presenting with acute decompensated cardiomyopathy. It has been suggested that intravascular volume of patients with pheochromocytoma-related cardiomyopathy is depleted when compared with patients with Takotsubo cardiomyopathy and myocarditis [7]. Patients with Takotsubo cardiomyopathy usually have recent stressful emotional or physical events, while those with myocarditis commonly have a viral prodrome prior to the onset of heart failure symptoms [7].

Pre-operative embolization has been employed to lower circulating catecholamine levels and help wean patients off ECMO before proceeding to adrenalectomies [8,9]. Our case is the first reported case to have post-embolization adrenalectomy done with ECMO on board due to the very ill status of our patient. Successful operation and normalization of her cardiac function prove that this is a feasible management pathway in critically ill patients with pheochromocytoma-related cardiomyopathy.

Although no radioisotope scan was done prior to the adrenalectomy due to the lack of immediate availability of radioactive tracer, an iodine-123 metaiodobenzylguanidine scan had been scheduled post-operatively to look for any residual disease which showed no residual pheochromocytoma. Genetic tests had been offered to the patient in view of pheochromocytoma diagnosed in young age and would be performed once the patient gives consent to those.

We have illustrated a case of pheochromocytoma-related cardiomyopathy in a young lady presented with acute decompensated cardiomyopathy, initially thought to be caused by mumps infection and was planned for a heart-transplantation. Though high clinical suspicion, sharp clinical acumen and a collaborative effort among different specialists, the correct diagnosis was reached eventually. A novel management pathway lining up pre-operative embolization of tumor followed by adrenalectomy with ECMO on board was proven to be successful.

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