Pulmonary Edema in Pheochromocytoma

Key words: acute respiratory distress syndrome, bronchoalveolar lavage, catecholamine, neutrophil

Pheochromocytoma is a rare condition and can be fatal if left undiagnosed. Diagnosis is notoriously difficult, as pheochromocytoma may present a broad spectrum of clinical manifestations. Typically, patients present with sustained or paroxysmal hypertension and frequently with the triad of headaches, palpitations and sweating. However, there are many reports of unusual presentations of benign or malignant pheochromocytomas that require emergency intervention. A high index of suspicion of its existence and its early diagnosis must be the key to a successful outcome.

Variable endocrine emergencies associated with pheochromocytoma cause variable symptoms. Marked catecholamine release from the tumor may cause cardiovascular emergencies, such as hypertensive crisis, hypotension, arrhythmia, myocarditis, cardiomyopathy, myocardial ischemia, and peripheral ischemia.

Hypertension can be sustained and paroxysmal, as a result of episodic secretion of catecholamines by the tumor. Blood pressure can reach very high values and such a situation is termed a hypertensive crisis when it is life threatening or compromises vital organ function. In 75% of patients, paroxysms with severe hypertension occur at least weekly (1). Severe hypotension is seen infrequently in pheochromocytoma patients and may be preceded by a paroxysmal hypotension. In less than 2% of pheochromocytoma patients profound shock is the presenting manifestation (2). Although sinus tachycardia is most commonly noted, a wide variety of arrhythmias can be detected in pheochromocytomas (3–6). Arrhythmias are common in patients with other diseases. Thus, in a situation where arrhythmia is accompanied with sweating, hypertension, and anxiety and it is paroxysmal, clinicians should consider pheochromocytoma.

As for other emergencies, pulmonary edema, abdominal bleeding, paralytic ileus, hemiplegia, acute renal failure, diabetic ketoacidosis, and so on may occur. Pulmonary edema as the first presentation of pheochromocytoma is uncommon and usually rapidly fatal (7, 8). It can be seen not only during the course of the disease, but also after tumor resection (9). In most pheochromocytoma patients pulmonary edema is cardiogenic in origin. Nonetheless, noncardiogenic pulmonary edema is also seen (7). It is thought to occur as a result of catecholamine-induced transient increase in pulmonary capillary pressure due to pulmonary venoconstriction and altered pulmonary capillary permeability (10).

Sukoh et al reported a case with noncardiogenic pulmonary edema associated with pheochromocytoma, and found neutrophil accumulation in the lung in this case (11).

Although there are some previous reports of noncardiogenic pulmonary edema associated with pheochromocytoma, findings of bronchoalveolar lavage fluid (BALF) had not been reported. Acute respiratory distress syndrome (ARDS) or acute lung injury (ALI) is known to involve neutrophil accumulation in the lung, and these neutrophils have a pivotal role for respiratory dysfunction. The report by Sukoh et al suggested that the neutrophil accumulation in the lung was caused by catecholamine drive. Further examinations, such as BALF from pheochromocytoma patients without pulmonary edema, will be necessary to clarify this point. We hope that collecting these cases will make the mechanism of noncardiogenic pulmonary edema clear in patients with pheochromocytoma.

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