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A Case of Pheochromocytoma Manifested as Noncardiogenic Pulmonary Edema

Received: March 10.1998

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Abstract: Pheochromocytoma is known as chromaffin cell tumor frequently originated from adrenal medulla and results in hypersecretion of catecholamines (1). Typical clinical findings include headache, palpitation, excessive sweating and paroxysmal hypertension, which persistently maintains in 60% of the cases. Generally, paroxysm or attacks are often, but they can appear with intervals of weeks or months (1, 2). Additionally, tachycardia, bradycardia and/or

fatal arrhythmia, acute myocard infarcts, spasms in coroner arteries, carcinoid-like manifestations, hyperamilasemi, acute abdomen, chest pain, intolerance to carbohydrates, nephritic syndrome (very rare), cardiac pulmonary edema, cardiomyopathie caused by catecholamines and very rarely noncardiogenic pulmonary edema may develop (3, 4, 5, 6).

Key Words: Pheochromocytoma, Noncardio-

A 16 year-old male patient was hospitalised because of his gruntly breath, dyspnea, nausea, vomiting and palpitation compliances. For the last 18-24 months, the patient applying to some health organisations with gruntly breath, dyspnea, dizziness and palpitation had been treated by several medication with the intention of managing cardiac rhythm irregularities.

Initial physical findings: Blood pressure: Systolic 150 and diastolic 90 mm Hg, pulsation: 90-100 beat /min rhythmic, body temperature: 36.7°C, general condition anxious and tired, skin pale and sweaty, conscious open. At respiratory system examination, difficult ventilation, long expiration, moderate and crude ralles in upper and middle zones in addition to disseminated biphasic roncus in bilateral lung regions in auscultation were found. At the examination of the heart, there was no pathological finding apart from increased intensity in the 2nd voice in pulmonary focus. At the abdominal examination, widespreedly, there was only mild sensitivity without rebound to palpation.

Initial laboratory findings: White blood cell: 7500 /mm³, Hb: 15 gr/dl, Htc: 49%, erythrocytes sedimentation rate: 2 mm/hr. There was no important finding at routine biochemical and urine tests. The values of arterial gases were PaO₂ 64 mm Hg, PaCO₂ 38.3 mm Hg, HCO₃ 18.2 mmol/L, pH 7.4. Central venous pressure was 5 cm H₂O. At chest x-ray fylms, there were soft infiltration zones with unclear borders that they were more obvious in the upper zones and heterogeneous in upper and middle zones (Figure-1). At teleradiography, cardiomegaly was not observed. No feature was present at tests ran microbiologically and rheumatologically.

Electrocardiographic and ecocardiographic tests, which were applied to clarify the ethiological reason for pulmonary edema, and consultation by cardiologists indicated that there was no cardiological pathology to cause to edema. The patient was started to be treated with nonspecific double antibiotic with wide spectrum for a possible pneumonia and pulmonary edema and managed.

Clinical progress: Respiratory difficulty was partly

decreased by O₂ inhalation, and Pa O₂ was escalated 77 mm Hg. Hypotension was seen for a short period after taking the patient under management could not be taken for a period, although fluid perfusion and positional applications were done. Arterial tension was tried to be kept over critical values by infusing vasopressor agents. Because vital organ perfusion was tried to be obtained. However, whenever we tried to cut administration of the vasopressor agents, hypotension developed immediately. There were considerable positional hypotension, vertigo and tiredness in the patient. Respiratory difficulty and infiltrations at lung radiographs recovered dramatically in a few days after hospitalisation (Figure 1 and 2). As the patient's symptoms partly stabilised approximately 60 hours after hospitalisation, vasopressor agents were cut because of finding systolic pressure 210 mm Hg and diastolic pressure 140 mm Hg. We observed immediate increase in TA once more at control done a few days later. During hypertensive attack and after that, the patient's respiration and lung examination findings did not get worse, and tachycardia was not found to be serious (90-110/min, rhythmic). In ultrasonography and CCT, a mass,

diameters of 5.4 cm x 4.6 cm x 5.0 cm, was detected in the left surrenal region. In order to investigate paroxysmic hypertensive disease, tests for vanil mandelic acid level in urine in 24 hours were ran, and it was found to be 17.0 mg. The patient was transferred to surgery department with the diagnosis of pheochromocytoma underwent an operation, and the mass excised from the left surrenal gland. Biopsy specimens confirmed the diagnosis. No pathology developed in patient in the postoperative period.

In pheochromocytoma, cardiogenic and non cardiogenic pulmonary edema may be present. Cardiogenic pulmonary edema resulted from pheochromocytoma is a well-known phenomena. This finding develops as consequence of late diastolic pressure increase of the left ventricle due to paroxysmal elevations in arterial blood pressure. The same finding may also be caused by myocarditis due to the high levels of catecholamines (2). Echocardiographic findings in cardiomyopathie caused by the elevated levels of catecholamines include either dilated or hypertrophic



Figure 1. Initial chest x-ray film showing nonhomogeneous-soft infiltrates in upper and middle zones.



Figure 2. Chest x-ray film in a few days after initial chest x-ray film

Table 1. Pheochromocytoma cases with noncardiogenic pulmonary edema reported

Author(s) and Year	Clinical findings	Hemodynamic finding	Urine levels of Catecholamine	Result
Scully et al, 1975	Vomiting and midepigastic pain	PCWP 5 mm Hg	ND	Death by pulmonary edema
Munk et al, 1977	Episodic tiredness palpitation	ND	VMA 30-116 mg/day CR (0-7) E 61-82 mg/day (0-15) NE 126-210 mg/day(0-50)	Recovery after a successful operation
Naejie et al, 1978	Respiratory failure, dyspnea	ND CVP 2 cm H ₂ O	NE 118-432 mg/day (20-120) E 13-36 mg/day (6-36)	Recovery after a successful operation
De Leeuw et al, 1986	Acute respiratory failure, dyspnea	PCWP 2 mm Hg, PAP 21/3 mm Hg COP 3.45 L/min Normal SVR	VMA 2.7 mmol/ CR mmol NE 20.2 nmol/L (2.5) E 3.8 nmol/L (0.8)	Recovery after a successful operation
O'Hickey et al, 1987	Vomiting and acute respiratory failure	Mean PAP 35 mm Hg PCWP 5 mm-Hg	MN 30.7 nmol/24hr (<5.5)	Death by DIC, diagnosed by autopsy
Joshi et al, 1993	Respiratory failure, vomiting, abdominal pain	PCWP 11 mm Hg COP 5.42 L/min CVP 9 cm H ₂ O SVR 878 dyne/sec/cm ²	NE 1043 mg/day E 202.8 mg/day NMN 13760 mg/day VMA 40.29 mg/day	Recovery after a successful operation
Our case 1997	Acute respiratory distress, nausea, vomiting, tiredness	ND CVP 5 cm H ₂ O	VMA 17.10 mg/day	Recovery after a successful operation

PCWP=pulmonary capillary wedge pressure, PAP=pulmonary artery pressure, COP=cardiac output, SVR=systemic vascular resistance, CVP=central venous pressure, VMA=vanillylmandelic acid, CR=creatinine, NE=norepinephrine, E=epinephrine, MN=metanephrine, NMN=normetanephrine. DIC=disseminated intravascular coagulation, ND=not done

cardiomyopathie-sometimes obstructive type-findings (5).

Noncardiogenic pulmonary edema is very rare. In the literature, only 6 cases have been reported since 1966. From them as only three well-substantiated cases and three possible cases were revealed. Scull et al. reported a case indicating that although the case had hemodynamic characters of noncardiogenic pulmonary edema, these factors might lead to lung findings because of presence of the left ventricular enlargement in the chest graphs and ascendant aorta dilatation findings (2) (Table-1).

The mechanism of the development of noncardiogenic pulmonary edema in pheochromocytoma cases is not clearly understood yet. An immediate beginning without

cardiac dysfunction findings implicates a pathogenesis alike neurogenic pulmonary edema. Theoretical mechanism explaining the appearance of neurogenic pulmonary edema is a formation of immediate and transient vasoconstriction resulted from intensive α -adrenergic stimulation due to symphatetic activity. As this condition affects the extravascular fluid clearance and causes to;

- shift of blood from the systemic circulation to lung circulation,
- vasoconstriction in the lung,
- lymphatic obstruction.

These factors result in edema due to the increase in hydrostatic pressure. Additionally, pulmonary

hypertension may lead to capillary permeability alterations and pulmonary haemorrhage. Neurogenic pulmonary edema may be prevented by early treatment with adrenergic blockers (2).

Diagnosis can be a problem in pheochromocytoma, since it has a great numbers of variations in clinical findings and biological activities. Paroxysmal hypertension is not a specific finding and not present, generally. With

the intention of diagnosis, catecholamines and their metabolites can be measured in the blood. Basal plasma catecholamine concentrations may have often and slightly been increased during normotensive periods, nevertheless, samples taken during hypertensive crisis present more accuracy and significance. When basal measurements are not sufficient for the accurate diagnosis, inhibitory and stimulatory tests may be ran (7).

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