A Case With Massive Pulmonary Embolism Related To Essential Thrombocythemia

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ABSTRACT

Essential thrombocythemia (ET) is a disorder that causes persistent increase in the platelet count. Thrombotic and haemorrhagic complications are the main causes of morbidity in ET. We present a case with massive pulmonary embolism related to essential thrombocythemia and treated with streptokynase (STK).

A 41 year-old woman presented complaining of progressive dyspnea, cough. The blood pressure 127/70 mmHg, pulse rate 104/minute, respiration rate 30/minute. The heart examination was normal. Pulmonary breath sounds were lowered in the basal of portion both hemitorax. Leukocyte count 16700/mm^3 with left shift, hemoglobin 11.4 g/dl, hematocrit 34.8%, platelet count 3384000/mm^3, pO_2 43 mmHg, pCO_2 25 mmHg, sO_2 81%, PH 7.48, D-dimer 1100 ug/ml. The chest X-ray findings were normal. The diagnosis of pulmonary embolism was made on the basis of clinical, laboratory, computorize tomography (CT) findings (filling defects which are thought as embolus in the both right-left pulmonary artery and their segmental branches). Heparin perfusion and aspirin were started. Because of clinical worsening; clopidogrel, diltiazem and STK perfusion were added to therapy. The patient was discharged from the hospital with aspirin, warfarin, clopidogrel, hydroxyurea, allopurinol.

We presented a case with massive pulmonary embolism related to ET. In this case, we had to decide whether we would make agressive treatment (expecially thrombolytic therapy) or not. For fatal bleeding complications could occur in ET as well as thrombosis. We presented this massive pulmonary embolism related to ET, as it was successfully treated with thrombolytic agent.

• ***Key Words:*** *Pulmonary embolism, essential thrombocythemia, thrombolytic.* ***Nobel Med 2008; 4(3): 42-44***

ÖZET

ESANSİYEL TROMBOSİTEMİ İLE İLİŞKİLİ MASİF PULMONER EMBOLİ OLGUSU

Esansiyel trombositemi (ET), trombosit sayısında persistan artışa neden olan bir hastalıktır. Trombotik ve hemorajik komplikasyonlar, ET'de mortalitenin ana nedenleridir. Biz, ET'ye bağlı olan ve trombolitik tedavi uygulanan bir masif pulmoner emboli vakasını sunuyoruz.

41 yaşında kadın hasta progresif dispne ve öksürük şikayetiyle başvurdu. Kan basıncı 127/70 mmHg, nabız 104/dakika, solunum sayısı 30/dakikaydı.

Kardiyak muayene taşikardi dışında normaldi. Akciğer sesleri her iki hemitoraksın bazallerinde azalmıştı. Lökosit sayısı 16.700/mm³ sola kaymayla birlikte, hemoglobin 11,4 gr/dl, hematokrit %34,8, trombosit sayısı 3.384.000/mm³, pO_2 43 mmHg, $pC0_2$ 25 mmHg, sO_2 %81, PH 7,48, D-dimer: 1.100 ug/ml. Akciğer grafisi bulguları normaldi. Pulmoner emboli tanısı klinik, laboratuvar ve bilgisayarlı tomografi bulguları (sağ ve sol pulmoner arter ve onların segmental dallarında emboli olduğu düşünülen dolum defekti) temelinde konuldu. Heparin perfüzyonu ve aspirin başlandı. Klinik kötüleşme nedeniyle tedaviye klopidogrel, diltiazem ve streptokinaz eklendi. Hasta aspirin, varfarin, klopidogrel, hidroksiüre ve allopurinol tedavisi ile taburcu edildi.

Biz bu vakada, agresif tedavi uygulayıp uygulamayacağımıza karar vermek zorundaydık. Zira, ET'de tromboz kadar fatal hemorajik komplikasyonlar da görülür. ET'ye bağlı masif bir pulmoner emboli vakasında başarıyla uygulanan trombolitik tedaviyi sunduk.

• **Anahtar Kelimeler:** Pulmoner emboli, esansiyel trombositemi, trombolitik. **Nobel Med 2008; 4(3): 42-44**

INTRODUCTION

Essential thrombocythemia (ET) results from a clonal proliferation of megacaryocytes within the bone marrow, leading to an absolute elevation of platelets that can cause both haemorrhagic and thrombotic complications.[1, 2] ET, occurs comparatively more often in young people and women.[3] We present a 41 year old woman suffered from massive pulmonary embolism related to ET and succesfully treated with thrombolytic therapy.

CASE REPORT

A 41 year-old woman presented to the emergency department complaining of progressive dyspnea cough for three days. There was nothing special on her own and family history. On examination, the blood pressure was 127/70 mmHg, pulse rate 104/minute, respiration rate 30/minute, temperature 37.1^{0}C. The heart examination was normal except tachycardia. Pulmonary breath sounds were lowered and there were minimal rales in the basal portions of both hemitorax. There was no organomegaly.

The laboratory tests on admission revealed a white blood count of 16700/mm³ with left shift, hemoglobin of 11.4 g/dl, hematocrit of 34.8%, platelet count of 3384000/mm³, pO_2 43 mmHg, pCO_2 25 mmHg, SO_2 of 81%, pH of 7.48, D-dimer of 1100 ug/ml, prothrombin time of 16.3 second, INR of 1.45, active partial thromboplastine time of 42.5 second. The renal and the liver function tests were normal. The chest X-ray findings were in the normal range. The electrocardiogram revealed sinusal tachycardia and $S_1Q_3T_3$ pattern. The spiral computerized tomography (CT) of the lungs revealed alveolar opacity and linear atellectasis in the right basal area. There were filling defects which are thought as embolus in the both right pulmonay artery and its segmental branches, and the left pulmonary artery's segmental branches. Portal doppler ultrasonography (USG), bilaterally lower extremity arteriel and venous doppler USG were normal. The scintigraphic examination of lungs was planned. But the patient's dyspnea progressed and her oxygen saturation decreased rapidly.

The diagnosis of pulmonary embolism was made on the basis of clinical, laboratory and CT findings. 1000 u/hour heparin perfusion and 150 mg/day aspirin were started. But, the clinical status failed. Thus, 75 mg/day clopidogrel, 180 mg/day diltiazem, were added to therapy, it was decided to start streptokynase (STK) perfusion. It was planned to give 100.000 unit/hours. Four hours later after the treatment oral mucosal bleeding developed. The STK perfusion was stopped until the coagulation tests became within the normal limits. When this therapy was interrupted, oxygen saturation of the →

patient started to decrease again. So, STK perfusion was restarted and completed in 28 hours totally. On the following hours, patient's clinical status was stabilized. She did not complain of dyspnea or bleeding and the oxygen saturation was normal. The therapy continued with aspirin, clopidogrel, and warfarin. The patient was eveluated by the hematologist and bone marrow aspiration and biopsy were done. The diagnosis of essential thrombocytosis was made of rulling out all other possible causes of secondary thrombocythemia. The patient was discharged from the hospital with aspirin, warfarin, clopidogrel, hydroxyurea, allopurinol calling for control.

DISCUSSION

ET is largely a diagnosis of exclusion. According to Polycythemia Vera Study Group,[4, 5] a diagnosis of ET requires a platelet count >600000/ ml, megakaryocytic hyperplasia on bone marrow aspiration and biopsy, and absence of conditions associated with reactive thrombocytosis or other myeloproliferative disorders such as polycthemia vera, chronic myelogenous leukemia or myelofibrosis with myeloid metaplasia on clinical, laboratory and bone marrow examination.

Although ET was first described with bleeding complications, thrombotic complications predominate.[1,6-8] Manifestations of these complications can range from myocardial infarction and stroke to acute extremitiy ischemia or erythromelalgia. Large artery thromboembolism such as is relatively uncommon. Deep vein thrombosis and/or pulmonary embolism as well as portal vein thrombosis predominate in the venous circulation. Hemorrhagic complications present most commonly as bleeding from the upper gastrointestinal tract and mucosal sites.[9] An elevated platelet count in an asymptomatic patient requires no therapy. Platelet pheresis has not been proven efficacious and can not be recommended. If platelet reduction is deemed necessary on the basis of neurologic symptoms refractory to salicylates. IFN-alfa or anagrelide, aquinazoline derivative can reduce the platelet count, but neither is uniformly effective nor without significant side effects. Hydroxyurea should be considered only if these agents are not effective or tolarable.[10]

In conclusion, we presented a case with massive pulmonary embolism related to ET. Because of the fastly worsening of the patient's clinical condition, we could not do the other diagnostic steps for the pulmonary embolism. We diagnosed with CT, laboratory and clinical findings. We had to decide that we would make agressive treatment (expecially thrombolitic therapy) or not. For fatal bleeding complications could occur in ET as well as thrombosis. But early and extensive usage of thrombolytic therapy in selected thromboembolic patients, lower mortality and morbidity rates.[11] Finally we completed STK perfusion with success in the therapy of massive pulmonary embolism.

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