CASE REPORT

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Fatal Aortic Tumor Embolism Presenting as Acute Paraplegia

Sung-Chul Jin, M.D.,[1] Do-Sang Cho, M.D.,[1] Jun-Hyeok Song, M.D.[2]

Department of Neurosurgery,[1] College of Medicine, Ewha Womans University, Seoul, Korea

Department of Neurosurgery,[2] Wooshinhyang Hospital, Seoul, Korea

We report a case of fatal aortic tumor embolism presenting as acute paraplegia. A four-year-old girl was referred from a local hospital with sudden paraplegia and a poor medical condition. A neighbor had noticed her fall from a bike, and she could not walk. She had no previous illness. Emergency spine MRI revealed no remarkable findings. During the process of evaluation, her general condition deteriorated progressively. Chest and abdominal CT showed a large mass in the left lung field, and a diagnosis of aortic occlusion was made. An emergency transfemoral embolectomy was attempted. However, the patency of the aorta was not recovered. On pathological examination of tissues taken from the embolectomy, a pleuro-pulmonary blastoma was found. The patient died 22 hours after the onset of her symptoms. We describe a possible mechanism for the tumor embolism. To the best of our knowledge, this is the first case report of aortic occlusion caused by an embolic malignancy, presenting as acute paraplegia.

KEY WORDS : Paraplegia · Pleuro-pulmonary blastoma · Aortic embolism.

Introduction

Acute paraplegia is a common presentation following aortic occlusion[3,5,6,8]. However, aortic occlusion itself is very rare[6]. The common causes of aortic occlusion include thrombosis of the atherosclerotic aorta, an embolus that originates in the heart, thrombosis of a thoracic aneurysm, and aortic dissection[7]. We report a case of acute aortic occlusion caused by an embolic pleuro-pulmonary blastoma, a very rare childhood malignancy. To the best of our knowledge, this is the first case report of acute paraplegia resulting from an aortic occlusion caused by a malignant tumor embolism.

Case Report

A four-year-old girl was referred from a local hospital for sudden paraplegia. According to her parents, a neighbor had noticed her fall from a three-wheeled bicycle, and she could no longer stand. In the local hospital, she complained of increasing pains in her legs, and her medical condition progressively deteriorated. She had no remarkable history of illness. On presentation to our emergency department, she was drowsy and exhibited paraplegia. Vital signs were : pulse, 140/min; blood pressure, 110/80mmHg; and respiratory rate, 30/min. Both lower extremities were cool, and no pulses were palpable there. Her abdomen was slightly distended and no bowel sound was audible. Emergency brain computed tomography(CT) was negative. Thoraco-lumbar magnetic resonance imaging (MRI) data were also unremarkable. Laboratory investigation showed severe metabolic acidosis (pH 7.243) and leukocytosis (28,000/dl). A simple chest X-ray revealed a large round mass in the left lung field. Chest CT showed a huge cystic mass adjacent to the thoracic aorta. Abdominal CT revealed aortic occlusion, multiple infarctions of the kidney and liver, and a distended bowel (Fig. 1). During the diagnostic assessment, the condition of the patient deteriorated. Emergency embolectomy was attempted soon after the diagnosis of aortic embolic occlusion was made. A longitudinal skin incision was made below the right inguinal ligament, and the femoral artery was exposed. The femoral artery showed no pulse. After clamping the femoral artery, we opened the artery and inserted a Fogarty catheter (# 3, 5 Fr.) upward, which was pulled down

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• Address for reprints : Do-Sang Cho, M.D., Department of Neurosurgery, College of Medicine, Ewha Womans University, 70 Jongno-6-ga, Jongno-gu, Seoul 110-783I, Korea Tel : +82-2-760-5132, Fax : +82-2-744-2489, E-mail : drcho@ewha.ac.kr

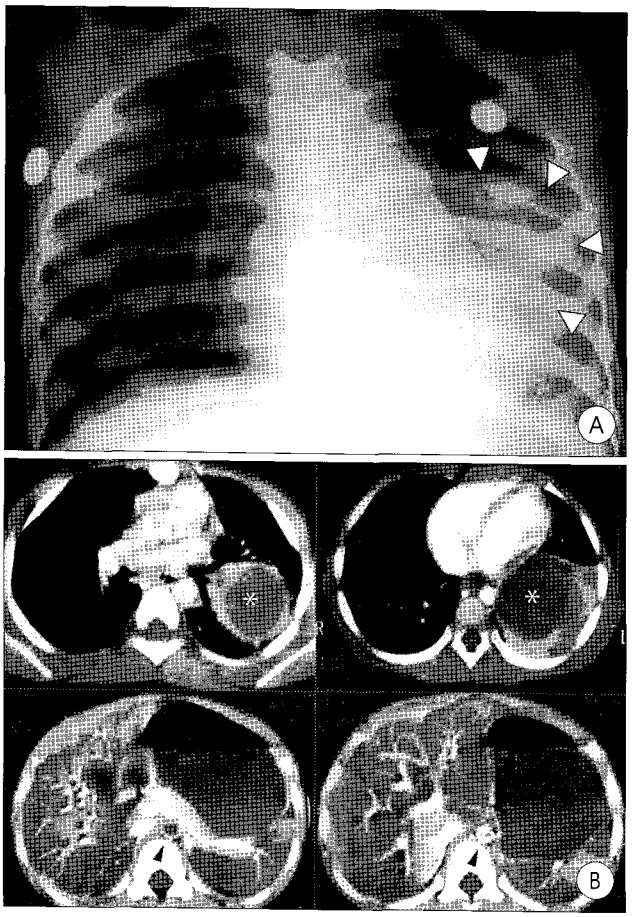

Fig. 1. A : Chest X-ray showing a large round mass (arrow head) in the a lung. B : Chest computed tomography showing a cystic tumor in the a lower lobe of the lung (asterisk). The mass and the aorta are in contact. Contrast enhanced computed tomography showing a filling defect in the descending aorta (arrow head), and multiple hypodense areas of liver infarction.

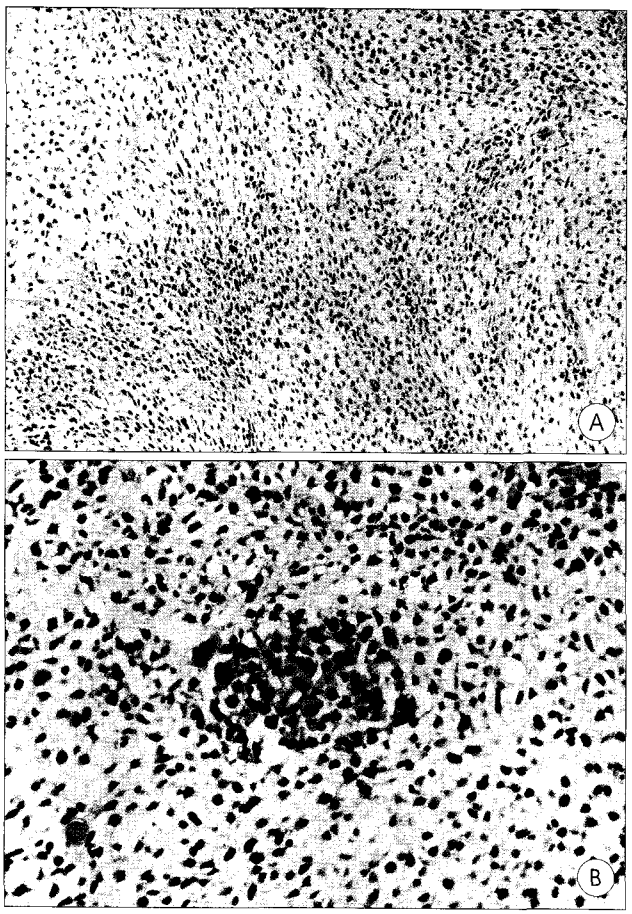

Fig. 2. A : Alternating bands of compact cells and loose cells in a myxoid matrix, characteristic features of pleuro-pulmonary blastoma (H&E, X100). B : A focus of anaplastic and pleomorphic mesenchymal cells (H&E, X200).

after ballooning. A large amount of gray gelatinous material was expelled through the opening. Confident of a tumor embolism, we closed the wound after discussion with the patient's parents. The patency of the aorta was not recovered. Despite intensive care, the patient died 22 hours after the onset of her symptoms. After pathological evaluation, a pleuro-pulmonary blastoma was diagnosed (Fig. 2). The child's parents refused an autopsy.

Discussion

The clinical syndrome of acute paraplegia is caused by traumatic spinal cord compression, by ischemic spinal cord injury resulting from occlusion of the aorta or supplying arteries, or by spinal cord compression caused by a hematoma or empyema. Aortic occlusion has been known to occur among patients with heart and/or atherosclerotic aortoiliac disease[1,3,7]. This can cause either ischemic spinal cord damage or ischemia of the cauda equina and peripheral nerves. A malignant tumor in our patient caused an embolic aortic occlusion, which has not been reported previously. Pleuro-pulmonary blastoma is a very rare childhood malignancy[4,9]. The entity was described as a variant of the classical blastoma that occurs in adults[2]. The concomitance of mesenchymal tumor and cystic lesions in the lung, as seen in our patient, has been reported in the literature[2]. This tumor has a poor prognosis[9]. Because no autopsy was performed, we could not identify the exact mechanism behind the aortic tumor embolism. Because chest CT revealed a cystic tumor mass in contact with the aorta, we assumed that the malignancy had progressively invaded the aortic wall, finally causing a defect in the aortic wall. A sudden blow to the chest or abdomen may have initiated the fatal embolic event. Since the tumor tissue was gelatinous and soft, it may easily have migrated through the defective aortic opening.

We believe that paraplegia following aortic occlusion is an extremely rare condition, and it may be difficult for neurosurgeons to make a prompt diagnosis. Spontaneous aortic occlusion typically presents as a catastrophic event with intense ischemic pain and a profound systemic response, including

tachycardia, diaphoresis, and a mottling of the extremities[7], as observed in our patient. The classic five P's – pain, pallor, pulselessness, paralysis, and paresthesias – can be diagnostic clues[7]. In our patient, the diagnosis of aortic occlusion was made 12 hours after the onset of her symptoms. The patient was too young to describe her symptoms precisely and traumatic spinal cord injury was the first assumption, considering the patient's history. This may have delayed an earlier diagnosis. In their clinical study of 18 cases of aortic occlusion, Littooy and Baker[7] reported a mean interval of about 18 hours from the onset of symptoms to definitive treatment. They reported a high perioperative mortality rate (40~62.5%). Various complications, including renal failure, compartment syndrome, adult respiratory distress syndrome, myocardial infarction, and disseminated intravascular coagulation, were also reported.

The operative approach will vary, depending on the origin of the acute aortic occlusion and the status of the patient. Bilateral transfemoral catheter embolectomy under local anesthesia is the usual choice when an embolus is suspected. The so-called golden period (6 to 12 hours) is relative and will vary among patients. If untreated, an aortic saddle embolism results in a mortality rate of at least 75%[7].

Conclusion

Prompt recognition and treatment may mitigate irreversible spinal cord injury or a fatal outcome. The possibility of an acute aortic occlusion should be considered when patients present with acute paraplegia and systemic deterioration.

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