Dissection of aorta: a pediatric case report

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We present a 15-year-old boy who developed sudden walking disability and sensory loss. He could not stand up on his feet and had no feeling following a sudden fall while playing basketball. He had been referred to a local hospital with these symptoms. In his physical examination absence of deep tendon reflexes and sensory loss were noted. His arterial blood pressure was 210/160 mmHg. He was transferred to our hospital with these findings and diagnosis of Guillain-Barré syndrome and hypertensive encephalopathy. There was sudden onset of sensory loss, walking disability and history of trauma. In the following hours hematuria, back pain and lower extremity ischemia developed. We suspected spinal artery injury based on the findings. Dissection of descending aorta was established with the help of magnetic resonance imaging of spinal region and contrasted aortography. The patient went to surgery immediately. He was lost on the second day after operation because of malperfusion. We report this case because dissecting aorta is very rare in the pediatric age group. High index of suspicion and early aortography are needed to diagnose aorta dissection.

Key words: aorta dissection, childhood, hypertension.

Dissection of the aorta is characterized by separation of the layers of the media by a column of circulating blood. This acute event is not associated with the presence of an aneurysm. The incidence is approximately 5.2 per million per year. It is seen in all age groups but it is rare in the extremes of life[1]. Although the etiology of aortic dissections is not well defined, hypertension, connective tissue disorders, aortic stenosis, coarctation of the aorta, iatrogenic trauma and pregnancy are the common causes. Clinically dissections seen within the first two weeks following onset of symptoms are considered acute and beyond this period chronic. The acute mortality in the critical first two weeks varies from 57% to 89%[2]. De Bakey[3] classification simplified the dissections into three basic types: Type 1 is a dissection starting in the ascending aorta and involving the entire length of the aorta; type 2 is limited to the ascending aorta; and type 3 starts distal to the left subclavian artery, but spares the ascending aorta and the arch. Male/female ratio has been reported as 2:1 or 3:1. Average age is 50-70 years. Acute mortality is 60-90% in type 1 and type 2, but 15-40% in type 3 dissections.

Case Report

15-year-old boy was referred to the hospital with the symptoms of walking disability and sensory loss. His symptoms had started when he was playing basketball. He had jumped up and fallen down suddenly, and immediately thereafter noticed that he had no feeling his feet and could not walk. He was referred to a local hospital: his arterial blood pressure was 210/160 mmHg. Absence of deep tendon reflexes and sensory loss of distal part of extremities were noticed. He was transferred to our hospital with these findings and a diagnosis of Guillain-Barré syndrome and hypertensive encephalopathy. His past and familial history revealed no remarkable findings. In his physical examination, weight was 90 kg ($<97^{th}$ percentile), height 180 cm ($>97^{th}$ percentile), and body mass index was 27.7 kg/m^2 (overweight). Cardiovascular and respiratory system examination was normal. Arterial blood pressure was 150/90 mmHg (90^{th} percentile).

Anti-hypertensive agent was given at the first hospital to which he was referred in the following hours his blood pressure was difficult to control. In neurologic examination he was conscious and cooperative, cranial nerves were intact, and muscle strength and tonus were normal in upper extremities, but significantly depressed in distal. Deep tendon reflexes were normal in upper extremities but absent in distal. Sensory loss was bilateral significant up to 10 cm over his knees. The other system findings were normal. Laboratory investigations included white blood cell count 22,300/mm^2, red blood cell count 4.58×10^6/mm^3, hemoglobin 10.9 g/dl, hematocrit 33.6%, platelets 295,000/mm^3 and normal urine examination in the first hour. At the sixth hour macroscopic hematuria started and significant erythrocyturia was found. Liver and kidney function tests, blood glucose, serum ions and prothrombin time were normal. At the sixth hour blood chemistry revealed: SGOT: 932 IU/L, SGPT: 341 IU/L, CPK: 1318 mg/dl, CPK-MB: 45 mg/dl, and LDH: 624 IU/L. Cholesterol and lipid values were normal. He was transferred to our hospital with initial diagnosis of Guillain-Barré syndrome and hypertensive encephalopathy. We thought this diagnosis was inlikely because symptoms of our patient appeared suddenly, did not show progression and there was no prodromal period. We were also doubtful of hypertensive encephalopathy, because he did not have clinical findings of encephalopathy. He had a history of sudden trauma and had serious hypertension. Lumbosacral X-ray and cranial tomography were normal. Chest X-ray showed widening of mediastinum (Fig. 1). His extremities were pulseless and became cold at about the seventh hour and he started to complain of back pain. We suspected spinal cord and artery injury with these clinical signs. Magnetic resonance imaging (MRI) of spinal region was done and showed pathology of the aorta, and type 3 dissection was defined (Fig. 2). The patient went to operation immediately and graft disposition was performed; he was last at the 48th hour after operation because of malperfusion.

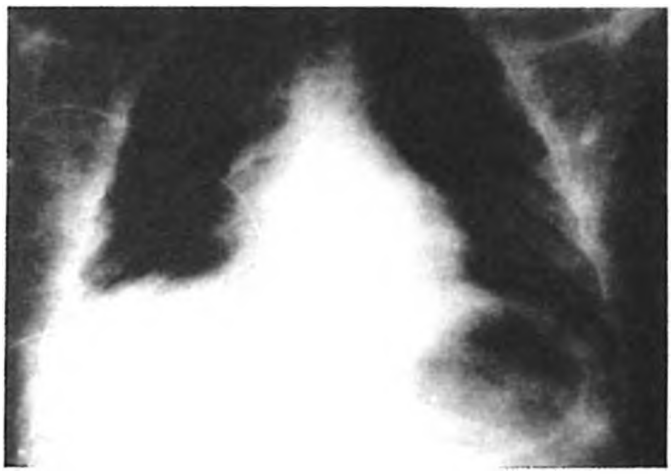

Fig. 1. Antero-posterior chest X-ray showed mediastinal broadening.

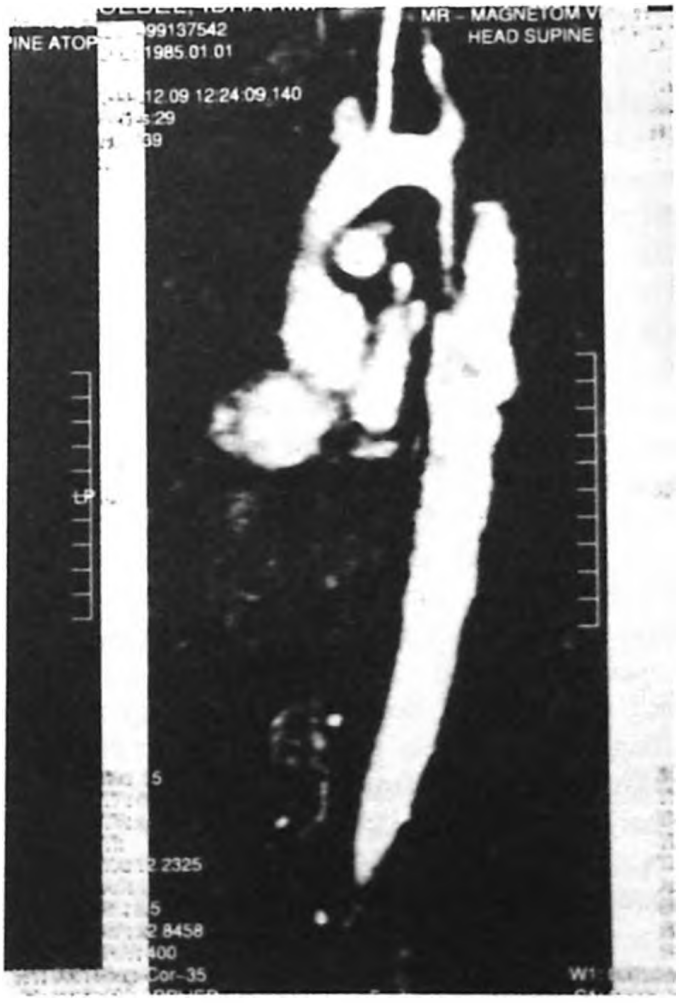

Fig. 2. 3D contrast magnetic resonance aortography showed dissection located at the proximal part of the descending aorta starting distally to the left subclavian artery.

Discussion

Acute dissection of the aorta is the most frequent catastrophic disease involving the aorta and remains the leading cause of death from aortic pathology. This pathology is very rare in pediatric age groups especially in children who do not have any predisposing risk factors. Etiology of aortic dissections is not well defined, but hypertension, heredity, connective tissue disorders, Marfan's syndrome, aortic stenosis and coarctation of the aorta are the predisposing

conditions[4]. Our patient was not evaluated before, so he and his family did not know whether he had hypertension or not. He was overweight and had serious hypertension when he was referred to the hospital. Sudden hypertensive attack might have occurred at the time of trauma during his sudden fall. Incidence of hypertension in acute dissections is about 75%[2]. The presence of hypertension is more common among patients with type 3 dissections[2]. Vogt et al.[5] reported four patients aged 14 to 21 years who developed acute aortic dissection. All four patients had systemic hypertension related to chronic renal insufficiency. The role of heredity in aortic dissections in the general population is poorly defined except for patients with connective tissue disorders like Ehlers-Danlos syndrome and Marfan's syndrome. Familial occurrence of dissections and annuloaortic ectasia in association with a mutation in the gene for type III procollagen has been reported[6]. Aorta-related complications, especially acute dissections, are the leading cause of death in Marfan's syndrome. Dilatation of the ascending aorta and a family history of acute dissection are associated with an increased risk of dissection in this syndrome[7,8]. There was no history of aortic dissection or sudden death in our patient's family. Aortic stenosis and coarctation, which are other etiologies of aortic dissection, were not determined during operation in our case. A review of the literature revealed many reported many of aortic aneurysms in childhood but very few of cases with aortic dissection. Dissection of aorta was reported in two cases with Turner's syndrome. They were 10- and 9-year-old girls and both had aorta coarctation repaired previously[9]. Teien et al.[10] reported a case of spontaneous dissection in a 12-year-old boy whose half brother had an idiopathic dilated aorta and whose mother had also required surgery for dissection of a dilated aorta. No features of connective tissue disorder were presented in any family member. Panja et al.[11] presented a nine-year-old girl with chest pain and dyspnea of sudden onset. She also did not have Marfanoid features and had normal aortic valve on echocardiography. The diagnosis of dissecting aneurysm of ascending aorta was established with the help of aortagraphy. Nitsuya et al.[12] reported that aortic wall dissection in young patients might be etiologically associated with increased acid mucopolysaccharide accumulation in the aortic media. Clinical features of aorta dissection change due to the type of dissection. Sudden or accelerated death may be the presentation and the diagnosis can be made only at autopsy. Pain is the most dramatic symptom. Back pain started in our case in the follow-up period. Neurologic manifestation such as temporary blindness, degrees of hemi- or paraparesis or plegia, or deep coma may be seen on presentation[1]. Our case presented with paraparesis and lower extremity ischemia. The blood pressure on admission is normal or high in over 80% of the patients. The blood pressure was difficult to control in our case. The severe hypertensive response may have been related to renal ischemia primary hypertension may have been present have in our case. Limb ischemia signs are present in about 40% patients at admission[2]. Hematuria and oliguria are signs of renal involvement. Hematuria started at the sixth hour in our case but urine volume as adequate (3 ml/kg/h). Abnormalities seen in the serum chemistries are related to severity of the accompanying organ dysfunction. Chest X-rays show widening of the mediastinum and blurring of the aortic knob in over 80% of the patients. Because diagnosis can be made only with computed tomography, magnetic resonance imaging or aortography[11], pediatric cases may be undiagnosed. Suspicion is essential to diagnose the disorder. We must consider dissection in children, especially with predisposing risk factors, in the presence of sudden onset of pain, dyspnea, syncope, hypertension, limb ischemia symptoms and hemiparesis.

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