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Case Report

Aortic Dissection in an 11-Year-Old Boy: Case Report

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ABSTRACT

We describe a young boy with epigastric pains and transiently altered consciousness, admitted for surgery.

After cardiology consultation and transthoracic echocardiography, a distal type aortic dissection was diagnosed. Computed tomography angiography for the evaluation of the aortic dissection was performed. Given the patient's hemodynamically stable state and the distal type of the aortic dissection, he was discharged with medical follow-up. ***(Iranian Heart Journal 2019; 20(3): 95-100)***

KEYWORDS: Aortic dissection, Young age

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Aortic dissection is very rare among children and young adults, with a prevalence rate of between 0.37% and 3.5%. [1, 2] It is a fatal condition characterized by the separation of the muscle layers of the aortic wall and the formation of a false lumen due to intimal tears. In some cases, these luminal tears may extend and cause rupture and sudden death. [3] Based on the literature, the mortality rate of aortic dissection exceeds 1% per hour for the first 48 hours and approaches 80% during the first month. Considering this extremely high mortality rate, aortic dissection is deemed a potential cardiovascular emergency. Therefore, early diagnosis and management are key factors in reducing patients' morbidity and mortality. [3, 4]

The clinical manifestations of aortic dissection differ noticeably depending on the involved portion of the aorta and also the organs affected by ischemia. [5] However, cases with aortic dissection are individuals older than 40 years [6] and usually present with complaints of sudden onset severe and ripping chest or back pains; syncope; dyspnea; weakness; absent or asymmetric pulses; and a cool, clammy, shocked appearance. [7, 8] Still, not all patients present with common signs and symptoms, and the diagnosis may be missed. [9]

We describe an 11-year-old boy with an unusual presentation in that he was referred to us with epigastric pains with radiation to the back, rendering the diagnosis extremely difficult. In this case report, we present the diagnostic evaluation of this patient.

Case Report

An 11-year-old boy with no significant past medical history presented to the Emergency

Department of Shahid Mohammadi Hospital for moderate epigastric pains radiating to his back. The pains had started 4 hours prior to admission, after transient altered mental status during exercise at school. The pains were continuous and non-reproducible and were not associated with dyspnea and aggravating or relieving factors. There were no retrosternal chest pains, orthopnea, paroxysmal nocturnal dyspnea, and palpitations.

In the initial evaluation, the patient was conscious and alert but appeared anxious. He had blood pressure of 100/70 mm Hg in the right arm and 95/60 mm Hg in the left arm, a body temperature of 36.9 ^{o}C, a respiratory rate of 14 breaths per minute, and a pulse rate of 80 beats/minute. The pulses were strong and symmetric in the carotid, brachial, and radial arteries; however, they were minimally weaker in the right femoral, dorsalis pedis, and posterior tibialis arteries than those in the left arteries.

In the head and neck examination, no bruits were heard and the jugular venous pressure was flat. Normal S1-S2 without cardiac murmurs was heard. The lungs were clear, and the bowel sounds were present. The abdomen was not tender, and there were no pulsatile masses. There was a normal rectal sphincter tone, and the stool was hemoccult-negative. Mental status and cranial nerve examinations were also normal. In addition, strength and deep tendon reflexes were normal in both upper and lower extremities.

The patient's white cell count had elevated to 11.8×10^{9}/L, but the other blood chemistry and hematologic studies were normal. In addition, abdominopelvic sonography was normal. His ECG indicated normal sinus rhythm without ischemic changes; however, he developed inverted T waves in the V_1-V_3 leads of his ECG.

Due to the difference in the force of pulses in the lower extremities, the suspicion of aortic dissection was created. Thus, bedside echocardiography was carried out for the evaluation of aortic dissection, which revealed normal left ventricular size, left ventricular ejection fraction, right ventricular size and function, no mitral regurgitation, aortic insufficiency, aortic stenosis, and mild tricuspid regurgitation. Furthermore, a dissection flap and a false lumen with a turbulent flow in the true lumen were seen in the descending Aorta, distal to the left subclavian artery (Stanford classification type B) (Fig. 1 & 2).

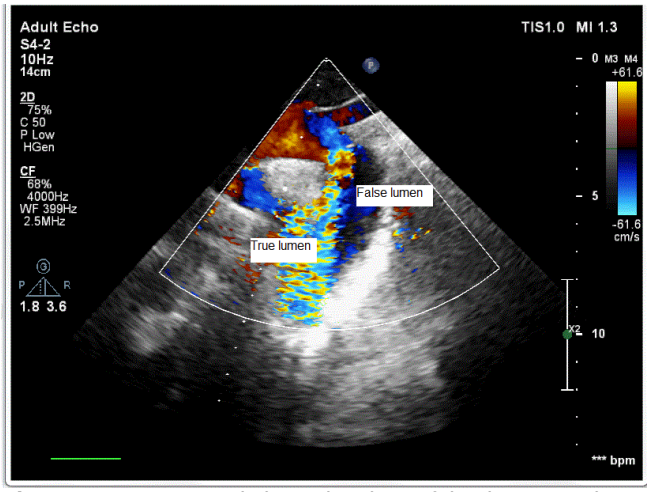

Figure 1. Suprasternal view, showing a false lumen and a turbulent flow in the true lumen

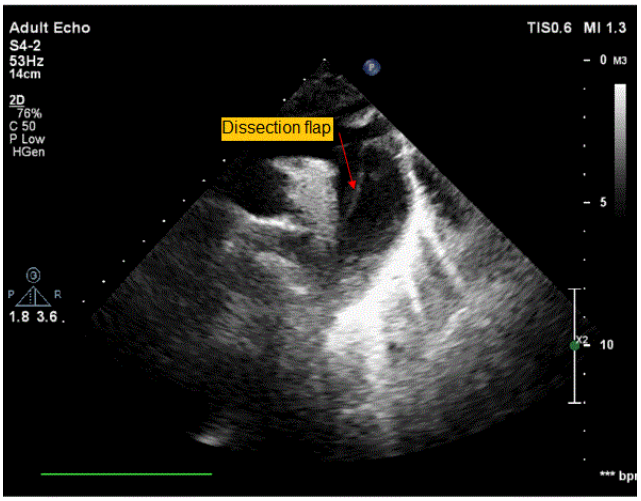

Figure 2. Suprasternal view, revealing a dissection flap just below the left subclavian artery

Additionally, Doppler study of the origin of the dissection showed a coarctation appearance, in favor of pseudocoarctation (Fig. 3).

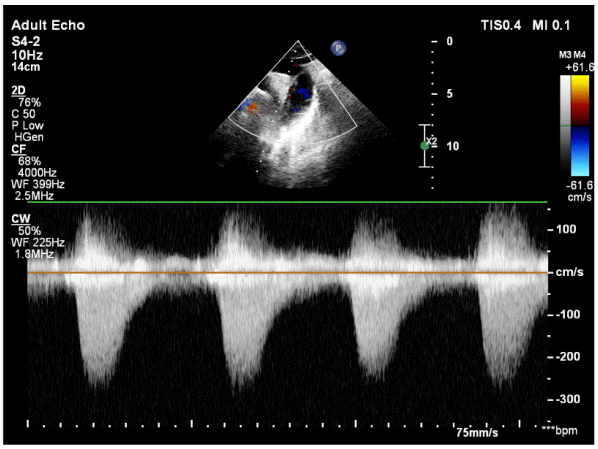

Figure 3. Doppler study in the proximal descending aorta, showing a coarctation pattern

In addition, a computed tomography angiography was performed, and it confirmed the diagnosis of the distal aortic dissection as far as the origin of the right renal artery. Consequently, the patient was admitted to the cardiology service.

Close monitoring of the patient's vital signs and clinical status was performed, and metoral (25 mg twice a day) was started. He was asymptomatic and stable.

Subsequently, the patient was sent to a referral center, where he was diagnosed with a type B aortic dissection and stable vital signs. He was hospitalized in the vascular surgery service. A 48-hour repeated computed tomography angiography showed no evidence of the expansion of the dissection. He was candidated for medical treatment and discharged on metoral (25 mg twice a day).

After 1 month, follow-up echocardiography illustrated no changes in the findings. In addition, the patient was stable hemodynamically and had no complaints.

During the patient's hospital admission, he received genetic counseling. Genetic studies were recommended to examine the possibility of Ehlers–Danlos syndrome and Marfan syndrome, but the patient did not consent to do it.

DISCUSSION

Aortic dissection is the most fatal condition of the aorta, with an annual incidence rate of 2.9 in 100 000 patients in the United States of America. [3] However, it is very rare in children and adolescents. Based on the literature, of 1085 cases reported in 2 studies, only 38 patients were 19 years old or younger, accounting for 3.5% of all the cases. [1] Hirt et al [10] reviewed 505 patients with aortic dissection and reported that only 7 (1.4%) patients were younger than 20 years and none was under the age of 14 years. In another survey, only 45 out of a total of 12 142 (0.37%) cases of aortic dissection occurred in persons 21 years old or younger and no patient was younger than 15 years of age. [2]

In untreated patients, 60% of individuals will die within 24 hours of presentation, 80% within 15 days, and 90% die within 3 months. However, survival may be considerably increased by the timely beginning of appropriate medical and surgical therapy. [11]

Although there are several types of classifications for aortic dissection, the Stanford and DeBakey classifications are the most commonly used formats. The Stanford classification divides the dissections into 2 types: type A, which involves the ascending aorta, and type B, which is any dissection that does not involve the ascending aorta. [4] On the other hand, DeBakey classifies the dissections into 3 types: 1) dissection of the ascending aorta and a variable portion of the descending aorta; 2) involvement of the ascending aorta; and 3) involvement of the descending aorta with (IIIb) or without (IIIa) the dissection of the abdominal aorta. [12] Therefore, our patient clearly had a type III (DeBakey classification)/type B (Stanford classification) dissection. The majority of aortic dissections (up to 90% of cases) arise from the proximal portion of the aorta, while only 10% originate distal to the left subclavian artery. In fact, the

dissection of the descending aorta is relatively rare in comparison with the ascending part. [13] Similar to the present report, Schor et al [6] documented a case of a descending dissection (type B or type III) in a young weightlifter.

The intimal tear and its propagation classically begin by the forces of blood pressure, turbulence, ejection velocity, and the steepness of the pulse wave. There are variable predisposing factors for the degeneration of the collagen, elastin or muscles in the intima media, which lead to the thinning of the media. In fact, the weakness of the intima predisposes it to tear. [4, 6]

In adults, atherosclerosis and chronic hypertension are the most prevalent predisposing factors, while inherited disorders of the connective tissues—especially Marfan syndrome—congenital cardiac diseases, and traumas are the most common risk factors in children. [14] Surgical procedures might also lead to the occurrence of aortic dissections. For instance, a 12-year-old girl who developed aortic dissection within 14 months after successful balloon angioplasty for native coarctation was reported by Beitzke et al. [15] Marfan syndrome has been reported as the most common factor in aortic dissection in children and adolescents. [14] In contrast, Fikar [2] reported that the most common associated condition was trauma (42%), with Marfan syndrome the second most common (24%), while 10 out of 45 (22%) adolescent and young adult patients had no apparent risk factors. Other well-established factors include coarctation of the aorta, aortitis and arch hypoplasia, Turner syndrome, Takayasu arteritis, tuberous sclerosis, giant cell arteritis, crack cocaine use, Noonan syndrome, strenuous resistance training, cystinosis, and pregnancy. [3, 4]

Another case-series study performed by Zalzstein et al [14] reported 13 children and young adults with aortic dissection between 1970 and 2000 in 4 medical centers. In 6 patients, there was an acute onset of chest pains, while 2 had abdominal pains or signs of bowel ischemia and 1 patient had neck swelling and signs of the superior caval venous syndrome. As was seen in our patient, the femoral pulses were absent or weak in 2 patients. One patient with chronic dissection due to Marfan syndrome was asymptomatic. A congenital cardiac defect was present in 5 patients, with the tetralogy of Fallot present in 2, patency of the arterial duct in 1, aortic valvar stenosis in 1, and aortic coarctation in the other. Marfan syndrome had been diagnosed in 4 patients. In 3 patients, the dissection followed blunt traumas to the chest during road accidents, while the final patient had no previous known risk factor. In 3 patients with congenital cardiac defects, aortic dissection was a complication of a medical procedure. Finally, only 3 of these cases had the descending aorta dissection.

Based on previous studies, the typical presenting symptom is the sudden onset of intense chest or back pains within the interscapular region. The pains are characteristically stabbing, tearing, or ripping. [4] In addition, sometimes dissection presents with symptoms of ischemia in cerebral and visceral organs or extremities, which were absent in our patient. [6] For instance, Morita et al [9] reported completely painless acute aortic dissection in a patient who presented with transient left hemiparesis in association with the dissection of the aortic wall. Syed and Fiad [16] also reported a case of aortic dissection in a 32-year-old man who presented with mild central chest pains and transient paraplegia.

Some clinical features suggest the possibility of aortic dissection; these features include the presence of chest pains and differences in pulses and blood pressure between the arms in hypertensive middle-aged or elderly men (5th – 7th decades). On the other hand, the absence of a history of hypertension, young age, and the subsequent absence of significant blood pressure difference between the arms can lead to diagnostic difficulties. [7, 16] In this regard, up to one-fifth of patients with acute aortic

dissection may present with syncope without a history of chest pains, as was seen in the present report. [14]

Considering the literature, our patient differs from others in several respects. The uniqueness of our patient is the site and severity of pain, the presence of dissection in the descending aorta, very young age, and the lack of significant difference in blood pressure between both upper and lower limbs. However, the diagnosis of aortic dissection was suspected as a result of the presentation of a difference in the pulses in this young boy. The patient's transient altered consciousness and the difference in the pulses of the lower extremities supported the possibility of systemic embolization. Chiming in with the present report, Klompas [17] highlighted the importance of a complete physical examination noting that 31% of cases had evidence of a pulse deficit.

Given that the diagnosis needs to be confirmed by imaging tests, we performed echocardiography and computed tomography angiography. Other diagnostic modalities include chest radiography, contrast-enhanced computed tomography, aortography, and magnetic resonance imaging. [4]

Although the mode of presentation was uncommon, this was not the first such case. A patient reported by Lee et al [18] presented with dysphagia and hoarseness associated with painless aortic dissection, which was a rare case of cardio-vocal syndrome. Therefore, considering the variable manifestations of aortic dissection is a clue, which will facilitate the diagnosis.

CONCLUSIONS

No definite cause of aortic dissection could be established in our patient. Aortic dissection in individuals younger than 40 years of age is rare unless there is a familial predisposition such as Marfan syndrome. The main challenge in managing acute aortic dissection is to suspect and diagnose the disease as early as possible. Because of its rarity in the younger population, prompt diagnosis is often delayed. One also needs to be aware of less common presentations such as the absence of chest pains. In conclusion, this report highlights the importance of considering the diagnosis of aortic dissection and performing a complete clinical examination in all patients with epigastric pains.

REFERENCES

1. Ngan, K.-W., et al., Aortic dissection in a young patient without any predisposing factors. Chang Gung medical journal, 2006. 29(4): p. 419.
2. Fikar, C.R. and R. Fikar, Aortic dissection in childhood and adolescence: an analysis of occurrences over a 10-year interval in New York State. Clinical cardiology, 2009. 32(6).
3. Deshmukh, A. and G. Deshmukh, Aortic Dissection-A Case Report. Indian Journal of Public Health Research & Development, 2013. 4(1): p. 66.
4. Aziz, F., et al., Peripartum acute aortic dissection: a case report & review of literature. Journal of thoracic disease, 2011. 3(1): p. 65.
5. Inamasu, J., et al., Paraplegia caused by painless acute aortic dissection. Spinal cord, 2000. 38(11): p. 702.
6. Schor, J.S., M.D. Horowitz, and A.S. Livingstone, Recreational weight lifting and aortic dissection: case report. Journal of vascular surgery, 1993. 17(4): p. 774-776.
7. Donovan, E.M., G.K. Seidel, and A. Cohen, Painless aortic dissection presenting as high paraplegia: a case report. Archives of physical medicine and rehabilitation, 2000. 81(10): p. 1436-1438.
8. Rosen, S.A., Painless aortic dissection presenting as spinal cord ischemia. Annals of emergency medicine, 1988. 17(8): p. 840-842.
9. Morita, S., et al., Painless acute aortic dissection with a left hemiparesis: a case report. Journal of Stroke and Cerebrovascular Diseases, 2005. 14(1): p. 36-38.

10. HIRST JR, A.E., J. VARNER J JOHNS, and S.W. KIME JR, Dissecting aneurysm of the aorta: a review of 505 cases. Medicine, 1958. 37(3): p. 217.

11. Lijoi, A., et al., Circumferential dissection of the ascending aorta with intimal intussusception. Case report and review of the literature. Texas Heart Institute Journal, 1994. 21(2): p. 166.

12. Morris-Stiff, G., et al., Post coital aortic dissection: a case report. Journal of medical case reports, 2008. 2(1): p. 6.

13. Nusair, M., et al., A case of aortic dissection in a cocaine abuser: a case report and review of literature. Cases journal, 2008. 1(1): p. 369.

14. Zalzstein, E., et al., Aortic dissection in children and young adults: diagnosis, patients at risk, and outcomes. Cardiology in the young, 2003. 13(4): p. 341-344.

15. Beitzke, A., et al., Dissection of the descending aorta after balloon angioplasty of native coarctation. Pediatric cardiology, 1997. 18(3): p. 222-225.

16. Syed, M. and T. Fiad, Transient paraplegia as a presenting feature of aortic dissection in a young man. Emergency medicine journal, 2002. 19(2): p. 174-175.

17. Klompas, M., Does this patient have an acute thoracic aortic dissection? Jama, 2002. 287(17): p. 2262-2272.

18. Lee, S.-I., S.-B. Pyun, and D.-H. Jang, Dysphagia and hoarseness associated with painless aortic dissection: a rare case of cardiovocal syndrome. Dysphagia, 2006. 21(2): p. 129-132.