

Aortic Dissection in A Young Patient without any Predisposing Factors

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Aortic dissection is rare in the pediatric and young adult population. We hereby present a case of a 17-year-old male patient, without any predisposing factors, who developed an aortic dissection. The initial presentation was acute abdominal pain with massive retroperitoneal hematoma. His clinical condition deteriorated rapidly, did not respond to surgical hemostasis, and died within 36 hours of admission. The major autopsy finding was dissection of the descending aorta, extending from 2 cm distal from the origin of the left Subclavian artery. Microscopically, the aortic sections showed intimal thickening and tearing, medial smooth muscle loss, which was replaced by fibrous tissue, fragmentation of elastic lamellae with widening of interlamellar spaces, and cystic medial degeneration. The morphological features represented degenerative changes of the aorta, which were unusual in such a young patient. The pathophysiology, predisposing factors and relevant reports in the literature of aortic dissection in young patients are reviewed. (*Chang Gung Med J* 2006;29:419-23)

Key words: aortic dissection, young age, cystic medial degeneration.

CASE REPORT

This 17-year-old male patient suffered from sudden onset of left flank and abdominal pain and was sent to a local hospital immediately. He had no history of trauma, hypertension, or cardiovascular diseases. He was the only child in his family. The family history was unremarkable. His condition deteriorated rapidly within a few hours. After primary resuscitation, he was transferred to the emergency ward of our hospital. His coma scale was E1M4VE on arrival. The blood pressure was 140/ 79 initially and dropped to 100/ 35 later, and the pulse rate was 145/ min. He was afebrile, with clear breathing sound and regular heart beat. The abdomen was tense and distended. Hematemesis, hematuria and passage of bloody stool were noted. The laboratory data showed low hemoglobin, severe metabolic aci-

dosis, severe acute hepatic (ALT: 2136 IU/L) and renal (BUN: 18 mg/dL, Creatinine: 3.9 mg/dL) failure, and hypoglycemia. The emergency computed tomography showed massive retroperitoneal hematoma with arterial tearing at the descending aorta (Fig. 1). Emergency laparotomy was performed in order to stop the bleeding. Two tearing sites at the descending aorta were noted. One was close to the left renal artery and another was just above the inferior mesentery artery. The patient had uncontrolled hemorrhaging over the dissecting field and aortic clamping site, and died during the operation. The duration from onset of pain to death was less than 36 hours.

Autopsy findings

The patient was 180 cm in height and 80 kg in weight, and no marfanoid features were noted. Upon

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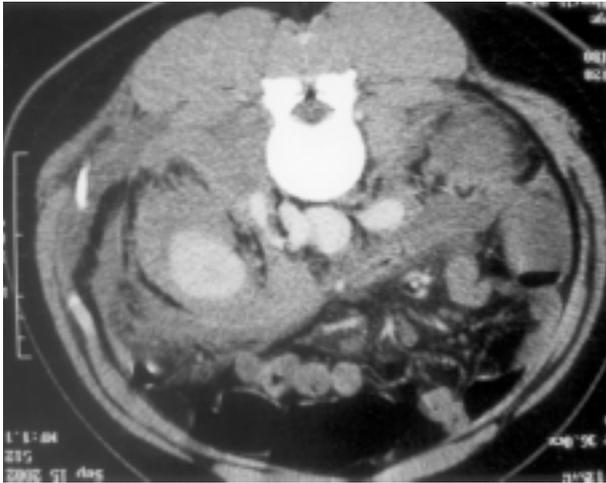


Fig. 1 Abdominal CT with contrast media showing descending aortic tear with extravasation of the contrast media.

dissection, bilateral hemothorax and massive retroperitoneal hematoma were noted. Starting from 2 cm distal to the origin of the left subclavian artery, there was extensive dissection involving the thoracic and descending aorta (Stanford Classification of type B). Only the distal portion, from the renal artery to the aortic bifurcation, was spared.

Multiple levels of the aorta had been taken. Splitting between the inner two thirds and outer one third of the media was confirmed (Fig. 2A). Microscopically, there were intimal thickening and tearing (Fig. 2B). The media layer showed loss of smooth muscle fiber, which was replaced by fibrous tissue (Fig. 2C). Disarray and fragmentation of elastic lamellae, with widening of the inter lamellar spaces, forming a cystic medial degeneration pattern, which was demonstrated using the van Gieson stain (Fig. 2D). The distal spared segment of the aorta also

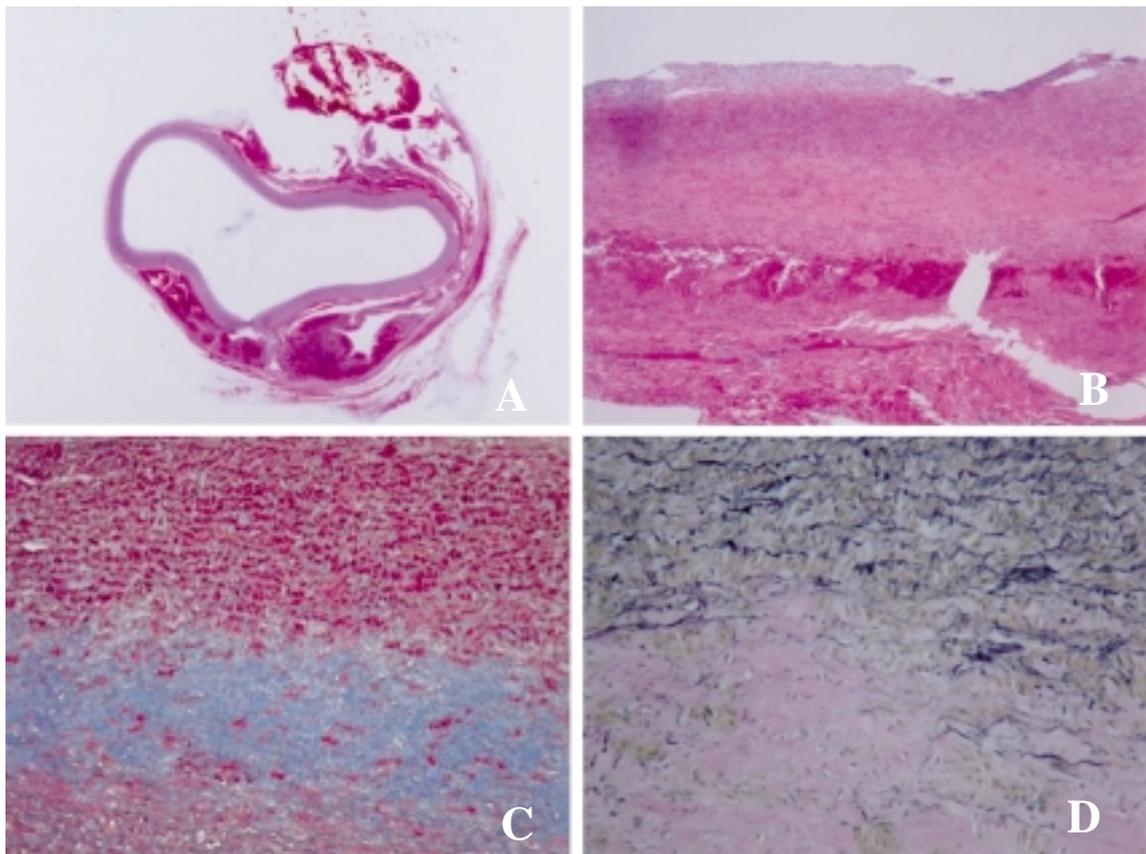


Fig. 2 (A) Splitting of the inner two thirds and outer one third of the aortic wall (H & E, original magnification). (B) Intimal thickening and splitting of media with hemorrhage (H & E, x 40). (C) The media layer shows loss of smooth muscle fiber, and is replaced by fibrous tissue (Masson Trichrome stain, x 200). (D) Disarray and fragmentation of the elastic lamellae, with widening of inter lamellar spaces, forming a cystic medial degeneration pattern (Van Gieson stain, x 400).

showed similar features. The overall histological findings were compatible with degeneration of the aorta.

Other findings included normal heart structure with 410 g in weight, bilateral pulmonary congestion and hemorrhaging, mild to moderate atherosclerosis of the left coronary artery and centrilobular necrosis of the liver, which explains the high serum ALT level. The ischemic changes of the kidney could not be recognized due to severe autolysis. The neuronal cells in the hippocampus and the purkinje cells in the cerebellum were well preserved, suggesting that there was no evidence of hypoxic encephalopathy.

DISCUSSION

Aortic dissection is an uncommon disease. It is considerably rare in young children and adolescents. Of 1085 patients reported in two earlier large series of aortic dissecting aneurysms, only 38 patients were 19 years old or younger, accounting for 3.5% of all cases.^(1,2) The first complete publication including autopsy findings was described in 1981.⁽³⁾ The researchers presented a 15-year-old male patient, without any predisposing factors, who had aortic dissection that led to rapid death. Since then, only limited cases have been documented as case reports.

Aortic dissection predominantly occurs in patients older than 40 years old. The most common predisposing factor is chronic systemic hypertension. In young children or adolescents, aortic dissection can occur without any predisposing factor, or it may be associated with congenital cardiovascular diseases and connective tissue disorders.⁽⁴⁾

Congenital cardiovascular disorder remains the most important risk factor in young patients. Aortic dissection has been reported in patients with developmental anomalies of the aorta such as aortic coarctation, aortic valvular stenosis, and in those with unicuspid/ bicuspid aortic valve.⁽⁴⁾ The intrinsic anomalies and secondary hemodynamic changes may lead to degeneration of the aortic wall. Surgical procedures might cause the occurrence of aortic dissections. A 12-year-old girl who developed aortic dissection within 14 months after a successful balloon angioplasty for native coarctation was reported in the literature.⁽⁵⁾

Marfan's syndrome is another well-established known risk. This autosomal dominant disease results

from a defect in fibrillin- I, an extracellular glycoprotein that acts as a framework during elastic fiber synthesis.⁽⁶⁾ Mutation of the Fibrillin- I gene coded on chromosome 15q21 is the cause. In large vessels, such as the aorta, elastic fibers account for more than 50% by dry weight.⁽⁴⁾ A defect of the elastic fibers led to early degeneration of the major vessel walls.

The prevalence of serious vascular malformations in young children with Marfan's syndrome is 5%, which is lower when compared with that in adults. Please state the amount in adults.⁽⁷⁾ It is important to state that mitral valve prolapse and mitral regurgitation, rather than aortic dissection, are more commonly seen. They are the leading causes of death in young children with Marfan's syndrome.⁽⁸⁾

Although our patient had left coronary artery atherosclerosis, his other major vessels including the aortic branches were unremarkable. In addition, he had no history of hypertension. We believe that the atherosclerosis was a coincidental event, most probably secondary to his diet, not a cause for the aortic dissection. A few cases of aortic dissection occurring in young patients with chronic hypertension have been reported. All of the four patients reported in the literature had sustained systemic hypertension related to chronic renal insufficiency.⁽⁹⁾

Other predisposing factors of aortic dissection at a young age reported in previously literature are summarized in Table 1. Familial cystic medial degeneration syndrome⁽¹⁰⁾ is an entity that describes those patients with more than one family member

Table 1. Summary of the Predisposing Factors of Aortic Dissection in Young Age Patients (Modified From Reference 4)

Idiopathic
Congenital cardiovascular disorders
Connective tissue diseases: Marfan's syndrome, Ehlers- Darlos syndrome ⁽¹¹⁾
Familial cystic medial degeneration syndrome ⁽¹⁰⁾
Postoperative ⁽⁵⁾
Systemic hypertension ⁽⁹⁾
Tuberous sclerosis ⁽¹²⁾
Turner syndrome ⁽¹³⁾
Cystinosis ⁽¹⁴⁾
Fibromuscular dysplasia ⁽¹⁵⁾
Pregnancy ⁽¹⁶⁾
History of weight lifting ⁽¹⁷⁾
Cocaine abuse ⁽¹⁸⁻¹⁹⁾
Trauma

developing aortic dissection or aneurysm. The aortic wall also shows similar features of degenerative changes. No specific inheritance was found. There was a study performed that indicated that first-degree relatives of probands had a higher prevalence of thoracic aortic aneurysms and sudden death, with the relative risk of 1.8, 10.9 and 1.8 in proband fathers, brothers, and sisters.

The definite pathogenesis of aortic dissection remains controversial. We support the pathophysiology of initiation and propagation.⁽⁴⁾ A patient might have underlying risk factors that induce mural degeneration. After tearing occurs, the dynamic circulatory forces induce the propagation of aortic dissection. As in our case, the patient with aortic wall degeneration developed a tear that led to circulatory collapse. Alteration of the dynamic circulatory force or possibly that of surgical manipulation during aortic clamping in order to stop the bleeding further propagated the clinical course. This theory might explain why only aortic tearing but not dissection was detected in the initial CT scan.

Despite improvement in current imaging methods, the initial radiological findings of aortic dissection might not be obvious. The histological findings of aortic dissection are characteristic. However, morphological features alone cannot disclose the underlying risk factors or pathogenesis of the aortic dissection. In conclusion, high clinical alertness remains important. Although rare, in a young patient with sudden onset of retroperitoneal hemorrhage with shock, especially in those with predisposing factors, the possibility of aortic dissection should always be listed as a differential diagnosis. The current management of aortic dissection includes open construction with conventional graft replacement, and endovascular graft method. Early recognition and management might be life saving.

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發生在年輕病人的主動脈剝離

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主動脈剝離 (aortic dissection) 在小兒及年輕病人中極為罕見，這裡我們報告一位 17 歲沒有任何危險因子而發生主動脈剝離的年輕男性病人，臨床上表現為急性腹痛以及後腹腔出血。病人情況急速變壞，並對外科止血無效，而且在 36 小時內死亡。主要解剖發現為由左鎖骨動脈以下 2 公分延伸到下主動脈出現血管剝離。病理學上的檢查顯示，主動脈呈現血管內膜增厚及撕裂，中層平滑肌消失並由纖維組織取代，內側彈性板彈性纖維斷裂 (fragmentation of internal elastic lamellae) 和間隙加大，及囊型內壁退化性病變 (cystic medial degeneration)。型態學上是退化性病變。此現象在年輕病人不常見。本人將探討年輕病人產生主動脈剝離的致病生理學及危險因子，並作相關文獻的回顧。(長庚醫誌 2006;29:419-23)

關鍵字：主動脈剝離，年輕，囊型內壁退化性病變。

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