H119 A Peculiar Autopsy Case of Infrarenal Aortic Hypoplasia (AH) and Premature Atherosclerosis in a 13-Year-Old Male

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Learning Overview: After attending this presentation, attendees will be aware of the case of a young male subject affected by infrarenal AH and premature atherosclerosis with no symptoms of hypertension or lower extremities hypoperfusion, who died from a cardiac tamponade consequent to acute type A aortic dissection with intrapericardial rupture.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by raising awareness of this peculiar and rare pathologic condition.

AH is defined as a narrowing of the aortic lumen with consequent hemodynamic obstruction of the blood flow. It is different from coarctation, where the luminal narrowing involves a short tract of the aorta and primary involves the upper descending aorta adjacent to the site of attachment of the ductus arteriosus. While coarctation of the abdominal aorta is extremely rare, AH is an even a rarer condition and has been described both in the thoracic and abdominal aorta.[1-4] The precise etiology of AH is unknown; however, both congenital and acquired (inflammatory and infectious) causes have been proposed.[5] The clinical manifestations are related to the localization of the hypoplastic segment because, when proximal to the renal arteries, it is responsible for the activation of the renin-angiotensin system.[2,6] The infrarenal hypoplasia can result in hypoperfusion of the lower extremities, causing symptoms such as claudication and lower levels of blood pressure relative to the upper limbs.[5]

This presentation reports the illustrative case of a 13-year-old child who died soon after a sudden illness at school. The child's previous medical history was notable for only mild aortic valvular failure along with an ectatic aortic bulb. At autopsy, the main findings were: (1) type A aortic dissection with intrapericardial rupture and hemopericardium (450ml of blood in the pericardial cavity); (2) hypoplastic infrarenal aorta (1.5cm in circumference extending for more than a length of 3cm) with a calcific atherosclerotic plaque deposition; (3) cardiomegaly (heart weight: 480g) with hypertrophy of the left ventricle (left ventricular thickness: 1.8cm; interventricular septum thickness: 2cm); (4) atherosclerosis of the descending thoracic and infrarenal abdominal aorta; (5) pulmonary congestion, and (6) hepatosplenomegaly. Histopathologic examination showed separation of the tunica media layers of the ascending aortic tract with acute hemorrhagic extension and fragmentation of the elastic fibers, as well as atherosclerosis of the infrarenal aortic tract. It also revealed myocardial hypertrophy, hepatic steatosis, and thickening of the renal arteriolar walls.

In conclusion, the described case shows the association between AH of the infrarenal tract and premature atherosclerosis, consistent with the published literature.[5,7] The peculiarity of the case lies in the extreme rarity of this pathological condition in a male subject of such a young age, without antecedent signs or symptoms. Only in the immediate peri-mortem period was there a crushing onset of clinical manifestations, including dyspnea and cold sweating with cardiac arrest shortly thereafter; this was suspected to be due to cardiac tamponade consequent to acute type A aortic dissection with intrapericardial rupture. Ultimately, the dissection (as well as the left ventricular hypertrophy) was the result of prolonged hypertensive stimulation of the cardiovascular system, despite the localization of the aortic hypoplasia to the infrarenal aorta.

Reference(s):

1. Roques X., Bourdeaud'hui A., Choussat A., Riviere J., Laborde N., Hafez A., et al. Coarctation of the abdominal aorta. *Ann Vasc Surg* 1988; 2: 138-44.
2. Graham L.M., Zelenock G.B., Erlandson E.E., Coran A.G., Lindenauer S.M., Stanley J.C. Abdominal aortic coarctation and segmental hypoplasia. *Surgery* 1979;86:519-29.
3. Di Eusanio G., Astolfi D., Piccoli G.P., Clementi G., Palminiello A. Diffuse hypoplasia of the thoracic aorta: surgical treatment with ascending aorta-supracaeliac abdominal aorta bypass graft. *J Cardiovasc Surg (Torino)* 1982;23:344-7.
4. Palmaz J.C., Carson S.N., Hunter G., Weinshelbaum A. Male hypoplastic infrarenal aorta and premature atherosclerosis. *Surgery* 1983;94:91-4.
5. Terramani T.T., Salim A., Hood D.B., Rowe V.L., Weaver F.A. Hypoplasia of the descending thoracic and abdominal aorta: A report of two cases and review of the literature. *J Vasc Surg.* 2002 Oct;36(4):844-8.
6. Parker F.B., Farrell B., Streeten D.H., Blackman M.S., Sondheimer H.M., Anderson G.H. Hypertensive mechanisms in coarctation of the aorta. Further studies of the renin-angiotensin system. *J Thorac Cardiovasc Surg* 1980;80:568-73.
7. Rossi M.A. Infrarenal aortic coarctation and diffuse hypoplasia of the aortoiliac-femoral system. *Acta Cardiol* 1997;52:373-9.

Infrarenal Aortic Hypoplasia, Premature Atherosclerosis, Aortic Dissection

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