Case report

Fatal Traumatic Rupture of Ascending Aortic Aneurysm Having Idiopathic Cystic Medial Necrosis: An Autopsy Case

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Abstract

This report describes an autopsy case pertaining to death due to traumatic rupture of aortic aneurysm. A 21 year old deaf and dumb male was assaulted with kicks over the chest. Autopsy revealed external injuries over the body. Internally, a fusiform aneurysmal dilatation of the lumen of the aortic root and tubular segment of the ascending aorta were observed, with tear of the anterior wall of the ascending aorta and resultant haemopericardium.

Histologically, the wall of the aneurysm revealed cystic medial necrosis, which appears to idiopathic in nature. A Common complication of aortic aneurysms is dissection, with subsequent spontaneous rupture. In this case, there was no evidence of dissection and the rupture was traumatic in nature. Death was certified as due to cardiac tamponade.

The extent of trauma to which the victim was subjected to, appears to be such as would have been insufficient to cause death in an otherwise normal individual. An account of the findings, along with a discussion of the pathology of aneurysms and cystic medial necrosis as also of the mechanism of rupture of aneurysms is provided.

Key Words: Cystic Medial Necrosis, aortic aneurysm

Introduction:

An aneurysm is a pathological dilatation of the lumen of a vessel. Degeneration of the medial layer of the aortic wall leads to weakening of the wall, resulting in progressive dilatation of the wall, leading to the formation of an aneurysm. Cystic medial degeneration of the aorta can occur due to connective tissue disorders like Marfan's syndrome or simply as an idiopathic condition due to various risk factors. An Aneurysm is at constant risk of rupture which may happen either spontaneously, or following trauma, leading to development of haemopericardium and resultant cardiac tamponade and death.

Case History:

The victim was a 21 year old mentally subnormal deaf and dumb male person. During a quarrel with his brother (who was also mentally – subnormal and congenitally deaf – mute), he was pushed from the staircase at his residence by his brother following a petty quarrel.

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When the victim landed over the cemented surface at the bottom of the flight of stairs, the accused person assaulted the victim with kicks over his chest and abdomen, following which the victim became unconscious. He was moved to the Community Health Centre of the area, where he was declared "brought dead".

Autopsy Findings:

External Injuries:

The following external injuries were observed over the body:

1. Reddish contusion, roughly circular, of 2 cms diameter and 1/4th cm deep (on cut section) over lower part of front aspect of left side of the chest.
2. Vertical grazed abrasion, measuring 13 cms x 9 cms over the back of left shoulder.
3. Reddish abrasion measuring 3 cms x 1 cms over lower part of left side of back of chest, below the angle of the scapula.
4. Reddish abrasion, measuring 4 cms x 3 cms, over the left side of lower trunk.

Internal findings:

The aortic root and tubular segment of the ascending aorta showed a fusiform aneurysmal dilatation of the lumen, with a diameter of 7 cms. There was a 9 cms tear of the entire thickness of the anterior wall of the ascending aorta. The pericardial sac contained 613 gms of clotted blood and 337 ml of fluid blood. No evidence of aortic dissection was noted in the wall. The other organs did not show any significant pathological changes. No alcohol or any

other substance was detected at chemical analysis of viscera. No fractures of the ribs or the sternum were found.

Histopathological Examination Findings:

Sections from the wall of the ascending aorta showed myxoid degeneration in the media. There was elastic tissue fragmentation and separation of the fibromuscular and elastic elements of the media by numerous cystic cleft – like spaces in the media containing basophilic amorphous extracellular matrix / ground substance. There was increased fibrosis of the media. These findings are consistent with Cystic Medial Necrosis.

Cause of Death:

Death was certified as "due to cardiac tamponade consequent to rupture of the dilated ascending aorta having cystic medial necrosis as a result of cumulative effect of the external injuries on the body, which was likely to cause death".

Discussion:

The aorta is composed of three layers: the thin inner layer or intima, a thick middle layer or media and a rather thin outer layer called the adventitia. The strength of the aorta lies in its media, which is composed of laminated but intertwining sheets of elastic tissue arranged in a spiral manner that affords maximum tensile strength. This tremendous accretion of elastic tissue gives the aorta not only tensile strength but also distensibility and elasticity.

The term aortic aneurysm refers to a pathological dilatation of the normal lumen, being defined as a permanent localised dilatation of the aorta having a diameter at least 1.5 times of the expected normal diameter of that given aortic segment. [1] The normal diameter of the ascending aorta is about 3 cms and length is about 5 cms. [2] In the instant case, the ascending aorta was dilated to a clearly aneurysmal width of 7 cms. An area of expanding investigation is the role of cellular mechanisms in the pathogenesis of aortic aneurysms. Destruction of the media and its elastic tissue is the striking histological feature of aortic aneurysms.

Experimental evidence indicates excessive activity of proteolytic enzymes in the aortas of affected patients, which may lead to deterioration of structural matrix proteins such as elastin and collagen in the aortic media and thereby promote or perpetuate the formation of aneurysms. Aneurysmal aortas contain elastolytic activity with an active elastase not present in the normal aorta, and other active proteolytic enzymes as well. The risk of rupture increases with aneurysm size. Smaller than 4 cms Aneurysm have 0-2 % risk of rupture, whereas those larger than 5 cms have 22% risk. [1]

Aneurysms of the ascending aorta most often result from the process of cystic medial degeneration/ necrosis. [1] Histologically, as in the instant case, cystic medial necrosis is characterised by elastic tissue fragmentation and separation of the fibromuscular and elastic elements of the tunica media by small left – like spaces where the normal elastic tissue is lost, and these areas are filled with the amorphous extracellular matrix of connective tissue and resemble, but are not true "cysts". Ultimately, there may be large scale loss of elastic laminae. Thus, the terminology "cystic medial necrosis", as medial degeneration is often called, is inaccurate, because neither true necrosis nor true cysts are present. Inflammation is absent. [1]

Although these changes occur most commonly in the ascending aorta, in some cases, the entire aorta may be involved. The histological changes lead to weakening of the aortic wall, which in turn results in the formation of a fusiform aneurysm. Cystic medial necrosis is found in connective tissue disorders like Marfan's Syndrome and Ehler Danlos Syndrome. However, in patients without Marfan's syndrome, it is not possible to recognise the histological diagnosis of cystic medial necrosis prospectively, i.e. without surgery or autopsy.

This fact has significantly limited understanding of cystic medial degeneration and its natural history by the scientific community, and it remains unclear as to what extent cystic medial degeneration may represent an independent disease process versus a manifestation of another disease state. It has long been suspected that patients who have proven cystic medial degeneration without the classic phenotypic manifestations of Marfan's syndrome may in fact have a variation or '*forme fruste'* of Marfan's syndrome; though this theory remains unproven. [1] In fact, Kubota J et al have reported a case of two brothers who presented with multiple visceral artery aneurysms and diffuse connective tissue fragility, but did not have any features of Marfan's syndrome. According to them, these cases belong to the heterogenous group of Marfan's syndrome in accordance to the above mentioned theory. [4]

However, on the contrary, many patients with ascending thoracic aortic aneurysms appear to have nothing more than idiopathic cystic medial degeneration, where it is unclear as to what specifically predisposes to the development of medial degeneration. It was first thought to be a degenerative process associated with old age, but subsequent reports have shown that it occurs not infrequently in young people. Hypertension is a common risk factor. Another risk factor is syphilis, though now a rare cause, in which degeneration of the aortic media occurs during the secondary phase of the disease producing a weakening of the aortic wall. Other risk

actors are Toxemia, nicotine, hyperadrenalism, infectious aortitis, great vessel arteritis. [1, 2, 5]

In the instant case, the since deceased was mentally subnormal and congenitally deaf – mute. His brother, the assailant, was also known to be mentally subnormal and congenitally deaf – mute. Otherwise, there were no definitive physical characteristics to suggest that the victim was suffering from either Ehler Danlos Syndrome or Marfan's syndrome. Hence, it appears most likely that the case is one of idiopathic cystic medial necrosis, rather than one due to Marfan'or Ehler Danlos syndrome. A genetic study in the instant case would have been helpful in arriving at a diagnosis, to prove or disprove the above mentioned theory that the victim may have been suffering from a variant of Marfan's syndrome.

A common complication of aortic aneurysms is "dissection", which begins with a tear in the intima that exposes a diseased medial layer to the systemic pressure of intraluminal blood. The blood penetrates into the media, cleaving it into two layers longitudinally and producing a blood filled false lumen within the aortic wall. This false lumen propagates distally progressively for a variable distance. Dissection is common in connective tissue disorders like Marfan'and Ehler Danlos syndrome. [2] Such a dissecting aneurysm frequently ruptures spontaneously or following trivial trauma or following a bout of hypertension, resulting in haemopericardium. In the instant case, there was no evidence of dissection. The rupture of the aneurysm was rather, secondary to trauma to which the since deceased was subjected to by the assailant.

Cardiac tamponade is a clinical syndrome caused by the accumulation of fluid in the pericardial space, resulting in reduced ventricular filling and subsequent hemodynamic compromise. In cardiac tamponade, blood accumulates in the pericardial sac faster than it can escape, either because the bleeding rate exceeds the drainage. When there is no laceration of the pericardium, there is no escape route for the blood from the pericardial sac. When sufficient blood accumulates, the pressure in the pericardial sac increases and begins to prevent the passive filling of the atria during diastole. The cardiac output falls, as does the systemic blood pressure and the venous pressure rises. If unrelieved, death follows, though the time that this takes is variable and difficult to calculate retrospectively on pathological findings. About 400-500 ml of blood is sufficient to cause death, though this seems to be a greater volume than is seen in tamponades. [6] In the instant case, the pericardial sac contained 613 gms of clotted blood and 337 ml of fluid blood.

Medico legally, culpability of the accused person would be determined by the interaction of trauma and disease. The injury sustained by the since deceased would be categorized as one that is "likely to cause death", because although it was an injury that posed a threat to the life of the victim and death as a consequence was not surprising owing to the structures involved, yet it was one where the death was caused due to superimposed trauma in an already diseased person. The same degree of external trauma to which the since deceased was subjected would not have resulted in death in a normal and healthy individual. Hence, the accused would be held guilty vide S. 299 IPC and punishable vide S. 304 PC for culpable homicide not amounting to murder. [9]

The ascending aorta is one of the four common sites where the aorta may rupture following trauma, the others being the aortic isthmus distal to the ligamentum arteriosum, the lower part of the aorta above the diaphragm, and at the junction of the innominate artery with the aortic arch. [8]

Rupture is more likely at sites of aneurysms, because of the reduced thickness and hence greater weakness of the wall, and because of the "La Place's law", according to which tension acting on the wall of a vessel is greater where the luminal diameter is greater. Hence, a much lesser degree of trauma would be required to rupture an aneurysmal aorta as compared to a normal one, as in the instant case.

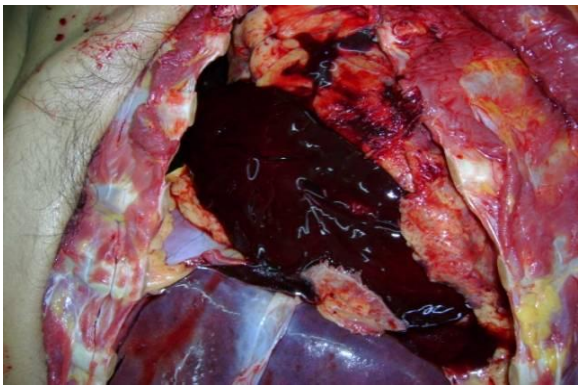
According to the osseous pinch mechanism / the aorta is crushed or pinched between the vertebral column and the inner surface of the manubrium, first rib and the clavicles during antero–posterior thoracic compressive deformation. [9]

Sudden deceleration also contributes to trauma to the aorta. During sudden deceleration, traction and torsion forces are placed on the aorta at points of anatomic fixation, i.e. isthmus and supravalvular ascending aorta. This is called the mechanism of "differential deceleration". During falls, the primary stress is longitudinal traction, with the weight of the heart producing greatest traction on the aortic root. Also, a sudden increase in intraluminal aortic pressure occurs with the force of impact, which is called the "water hammer effect." [10]

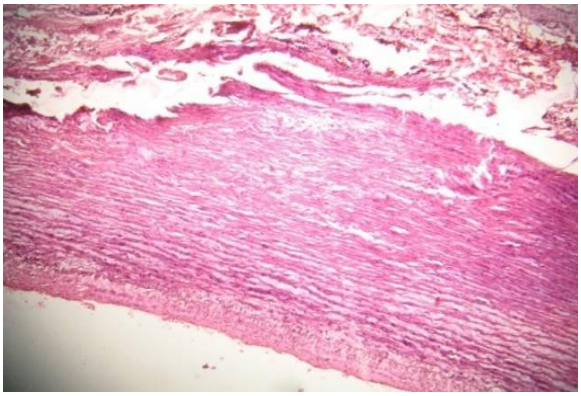
In the instant case, all of the above mechanisms may have acted together to produce laceration of the wall of the aorta. There was history of direct trauma to the chest by kicks from the assailant. The victim was seen to have been lying on the floor at the time he was being kicked, indicating that the chest would have been fixed, the back being supported by the floor. Presence of a bruise over the precordium as mentioned above makes it highly likely that kicks may have been perpendicularly directed with respect to the chest wall in a stamping – like action. This would cause antero posterior compression of the chest, traumatizing the ascending aortic aneurysm in accordance with the osseous pinch

mechanism, acting along with the "water hammer effect" mentioned above.

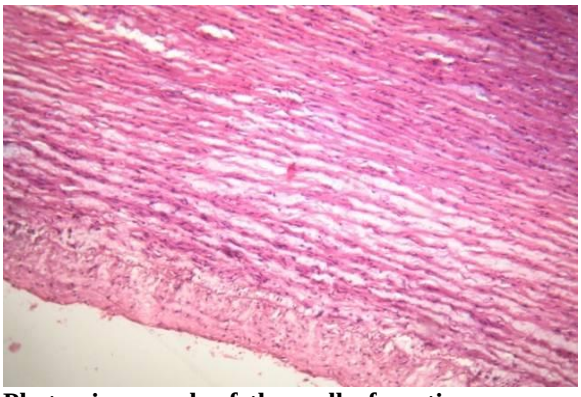
As the victim was pushed down a flight of stairs, sudden deceleration of the heart would have occurred, which would give rise to traction forces acting on the aorta, resulting in its rupture in accordance with the mechanism of differential deceleration.



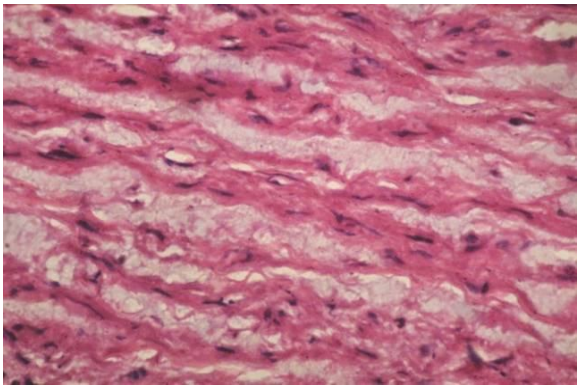
Photograph of the thorax showing Haemopericardium consequent to rupture of aortic aneurysm



Photomicrograph of the wall of aortic aneurysm showing cystic medial necrosis (Stained with H & E, x 5 magnification)



Photomicrograph of the wall of aortic aneurysm showing cystic medial necrosis (Stained with H & E, x 10 magnification)



Photomicrograph of the wall of aortic aneurysm showing cystic medial necrosis (Stained with H & E, x 40 magnification)

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