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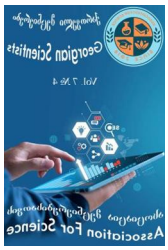

Aortic intima-media myxoid degeneration IMMD - A rare case report

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Abstract

Introduction: Aortic intima-media myxoid (IMMD) degeneration, often leading to valve insufficient by myxomatous tissue, is also known as aortic mucoid degeneration which also Leeds “cystic medial degeneration” involving the accumulation of mucin within the medial layer of the aorta, both are rare vascular disease characterized by the degenerative process in different layers of the arterial wall, which leads to formation of aneurysms at the damaged side. Patients with myxoid degeneration often have aneurysms at any location in the artery and postoperative bleedings are also common.

Case report: We describe three cases of material obtained by the cardio surgical method. Histopathological diagnosis: idiopathic aortic intima-media myxoid degeneration. Clinical cases reports are also provided, since such a pathology has not been described in the literature available to us.

Discussion: Acute decompensated heart failure may be caused by intima-media myxoid degeneration of the aorta. Aneurysms are common. Surgical treatment may be complicated by intra and postoperative bleeding, which requires caution during surgical intervention and specific postoperative management.

Key words: Cardiology; Cardio surgery; Vessels; Aneurysms; Mucoid/Myxoid degeneration of aorta: Acute decompensated heart failure.

Introduction: Aortic intima- media myxoid degeneration (IMMD), is a rare vascular disease characterized by the deposition of mucin in the arterial wall, which leads to degeneration of elastic tissue and the formation of aneurysms [1]. In Georgia, this pathological condition

is not presented as an independent nosologically entity, moreover, since the diagnosis is mainly made and are based on postoperatively obtained materials after histopathology. Due to the lack of an accurate diagnosis before surgery, intra and postoperative complications/bleeding are common [2]. Surgical treatment requires an individual approach and increased monitoring in the postoperative period. This is a pathological process in which the structure of aortic walls, especially its middle (media) layer, is disrupted due to the accumulation of mucopolysaccharides and the destruction of elastic fibers [3]. This degeneration is particularly common in young patients with hereditary connective tissue diseases, where the pathogenesis of aneurysms and aortic dissection is predominantly associated with impaired collagen synthesis [4]. The exact cause of mucopolysaccharides accumulation in elastic blood vessels, then mucoid degeneration, possible tissue rupture and aneurysms formation is still unclear. We describe three clinical cases of idiopathic aortic intima – media myxoid degeneration with histopathological findings, since such a pathology has not been described in the literature available to us. The patients had aortic dissection, aneurysmal of the arterial Wall and a ruptured thoracic aneurysm. All patients in the study underwent reconstructive vascular surgery for aortic aneurysms of various locations and aortic valve reconstruction with an artificial valve. Postoperatively, intima – media myxoid degeneration of the aorta was detected in all (three) patients. Clinical and histopathological data are discussed.

Case presentation: We are discussing patients who developed intima – media myxoid degeneration of the ascending aorta, aortic arch, descending aorta and aortic valves. All three patients are middle aged males (47,48 and 65 years). All three suffer from arterial hypertension, two of them had obesity and one hypothyroidism. The patients were admitted to the emergency department (Chapidze Emergency Cardiology Center – Tbilisi) due to acute heart failure of unspecified etiology. An echocardiogram on these patients revealed aneurysmal dilated areas in various locations of aorta (ascending aorta, aortic arch, descending aorta and aortic valves) aortic dissections, ruptured thoracic aneurysm and aortic valve insufficiency of unknown etiology. The patients underwent emergency surgery due to aortic wall dissection, aortic valve insufficiency of various localizations. In the postoperative materials obtained from the aforementioned patients: ascending aorta, aortic arch, descending aorta and aortic valves intima- media myxoid degeneration (IMMD) ICD-code171.21, with intima – media dissection, along with sclerosis and calcinosis are extremally interesting and rare conditions, because as mentioned above, mucoid/myxoid degeneration is a pathological process of independent non – inflammatory genesis, it mainly develops in the presence of genetic aberrations and is common in the av-mitral and tricuspid valves. All three cases were complicated by postoperative bleeding and one of them a 65 -years-old man inappropriate intraoperative bleeding.

Figures

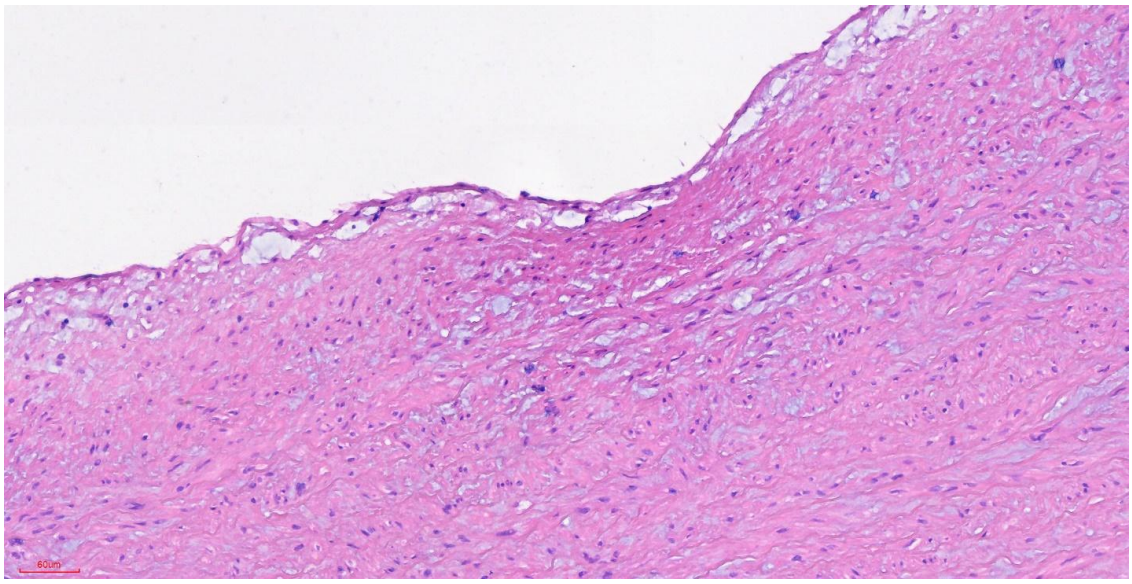

Fig. 1: Section of ascending aorta with large Myxoid intima-media degeneration. [Hematoxylin-eosin staining, original magnification 150×].

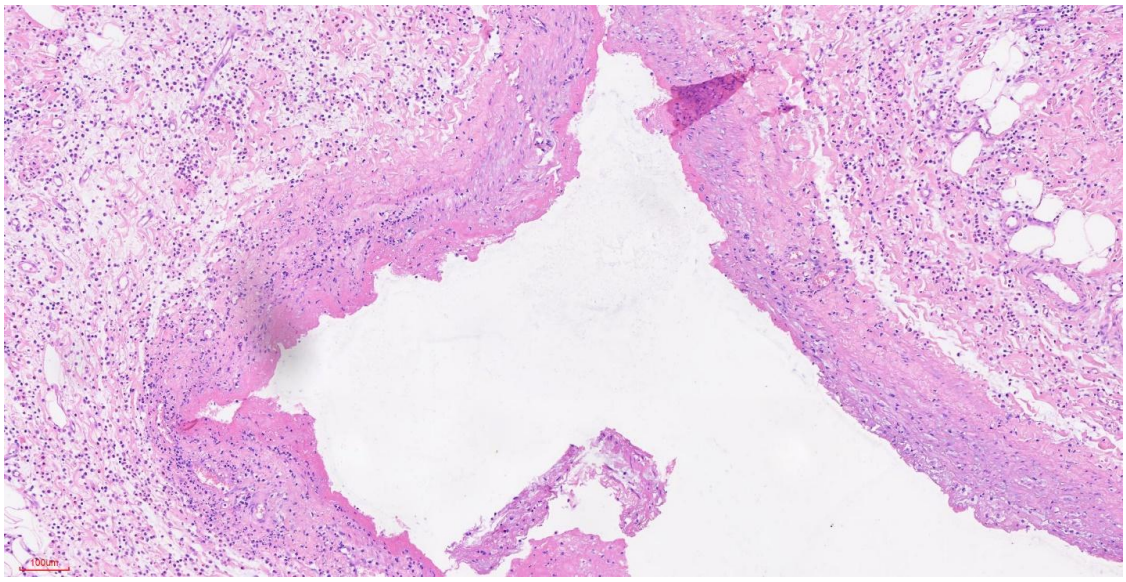

Fig.2 Section of aorta intima-media degeneration with lymphoplasmacytic infiltrate [Hematoxylin-eosin staining, original magnification 100×].

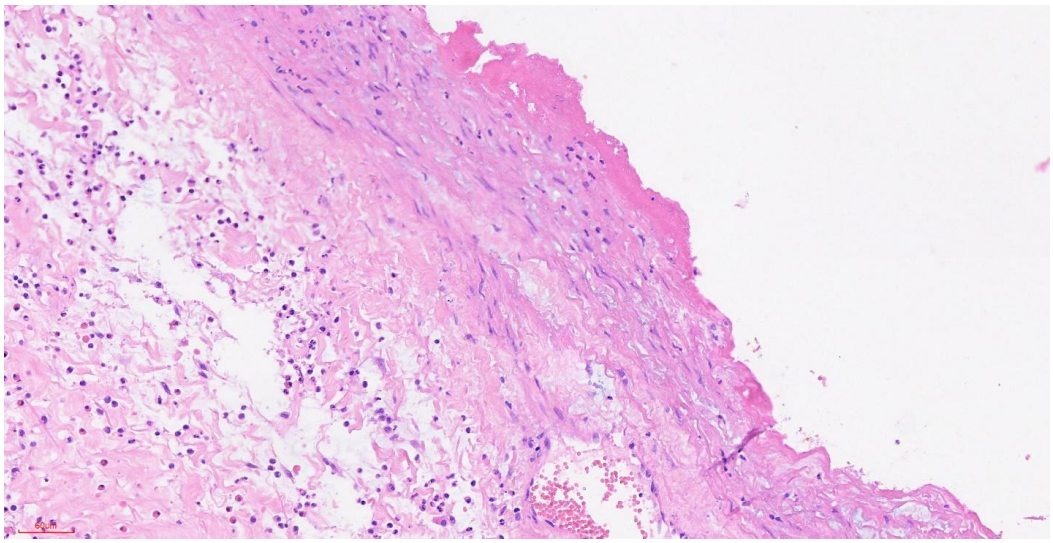

Fig.3 Section of ascending aorta with large Myxoid intima-media degeneration with lymphoplasmacytic infiltrate [Hematoxylin-eosin staining, original magnification 200×].

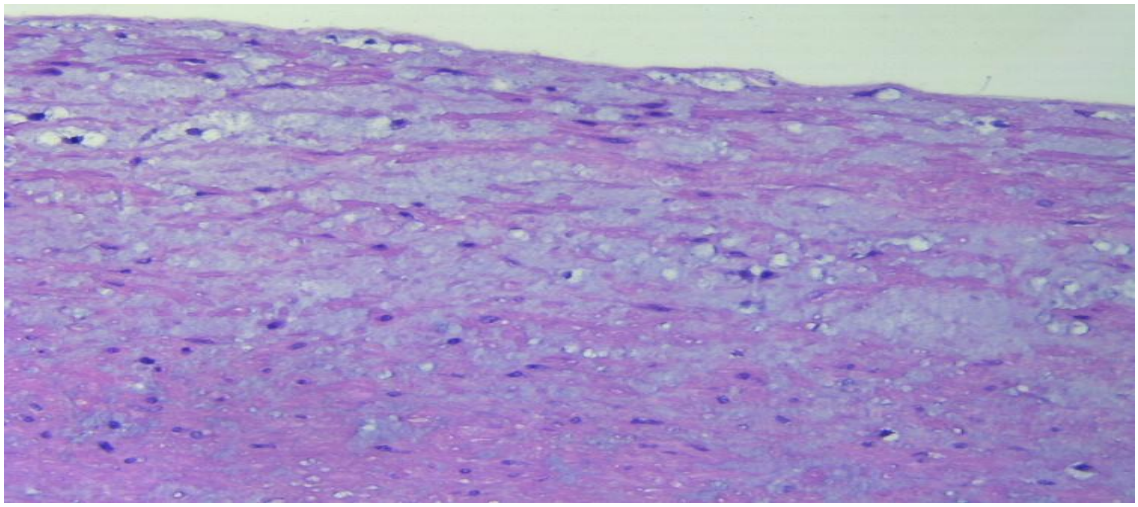

Fig. 4 Section of aorta with large Myxoid intima-media degeneration with lymphoplasmacytic infiltrate [Hematoxylin-eosin staining, original magnification 400×].

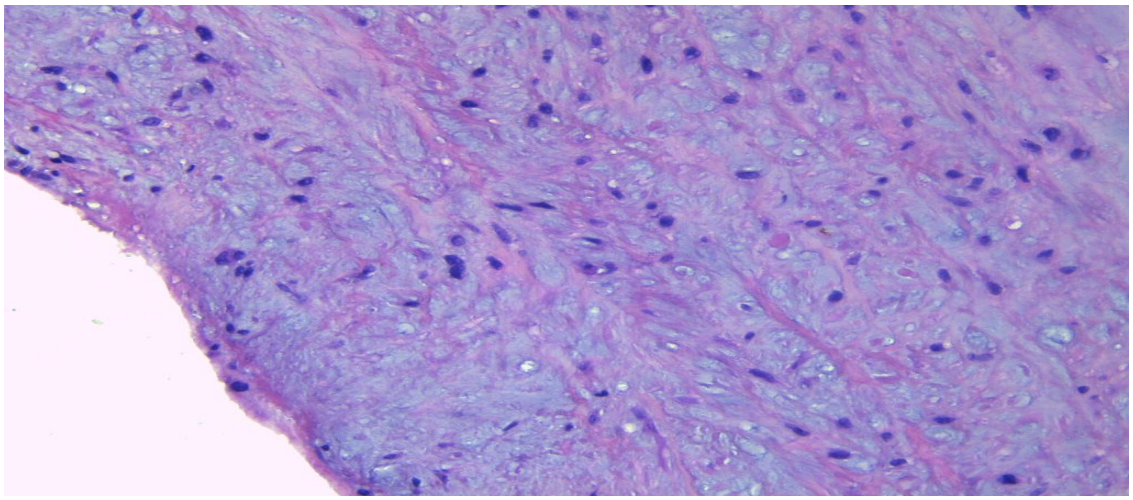

Fig.5 Section of aorta with large Myxoid intima-media degeneration [Hematoxylin-eosin staining, original magnification 400×].

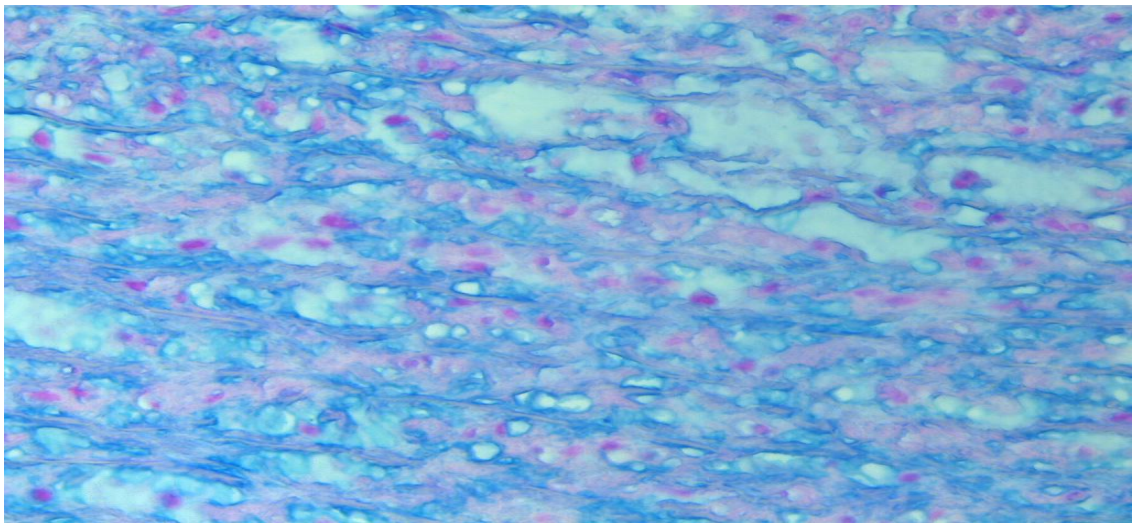

Fig.6. Section of aorta with large Myxoid degeneration [Alcian Blue staining for highlighting myxoid degeneration, original magnification 400×].

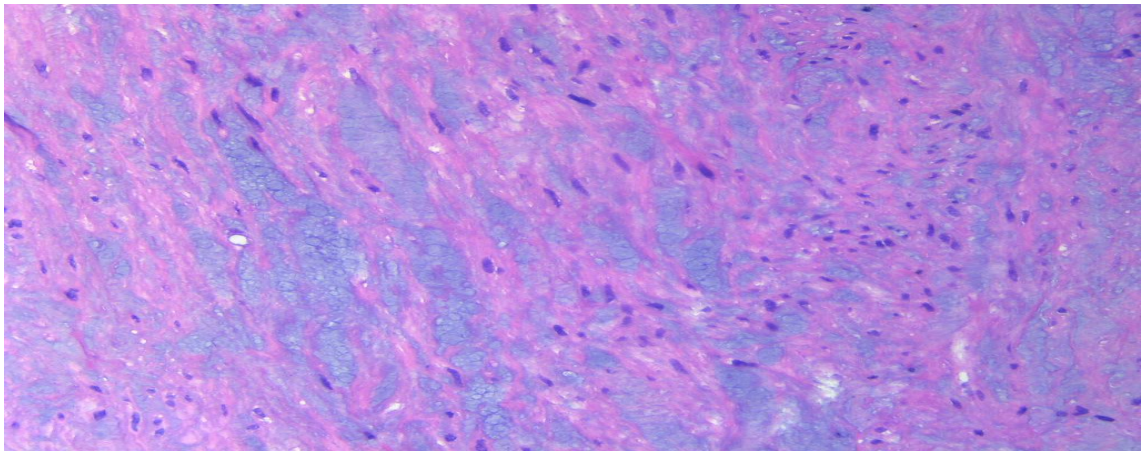

Fig.7. Section of aorta with large Myxoid degeneration. At high power, there is disruption of the organization of the wall my myxoid depositions by these elastic fibers are disorganized [Alcian Blue staining for highlighting myxoid degeneration, original magnification 400×].

Discussion: Intima – media mucoid degeneration of aorta can be mainly observed in various congenital condition associated with disorders of the connective tissue for example: Marfan syndrome have cardiovascular problems with aneurysms formation in aorta, Ehlers – Danlos syndrome (EDS) have a connective tissue defect called vascular Ehlers – Danlos syndrome is characterized by a defect in collagen, which leads to damage to the middle layer (media) of the blood vessels, mainly the artery. Vascular dissections syndrome, which can lead to aortic aneurysms and pseudoaneurysms (“false” aneurysms). Although aneurysms formation is relatively rare in dissection syndrome, aneurysm rupture/death is

unpredictable [5]. Among the four main features commonly seen in people with Lois-Dietz syndrome are aneurysms, which most often develop in the aortic root, but can also be seen in other arteries. Intima-media myxoid degeneration has been described as a distinct entity from “cystic medial degeneration/necrosis “of Erdheim. It is characterized by aneurysm formation secondary to degeneration of the elastic tissue of the arterial wall and deposition of mucin in the media. Cystic medial degeneration (CMD) and intima – media mucoid degeneration is both associated diseases of the aorta, although they differ in location and etiology. Cystic medial degeneration (CMD) primarily affects the media (middle layer) and adventitia (outer layer) of the aorta, leading to cystic degeneration and potential aortic aneurysm. Intima – media myxoid degeneration involves the intima (inner layer) and media, mucin deposition and degeneration of elastic tissue which also predisposes to aneurysm formation. IMMD mainly develops in the aorta but can also involve other large blood vessels. The exact cause of IMMD is unknown, but it is observed in young people, especially women, with a higher frequency in African populations [6]. Patients with intima-media myxoid degeneration of the aorta, dissection and aneurysms are associated with degenerative changes in its wall and, above all, with the accumulation of mucous substance along with damage to elastic fibers. Thus, the presence of aortic aneurysms in the above locations indicates morphological inferiority of its wall and should be considered when determining the tactic of surgical treatment to avoid intra and postoperative complications [7]. Based on the described cases, it can be said that patients with hypertension, diabetes mellitus, obesity and hypothyroidism who have clinically manifested heart failure from young age, regardless of congenital anomalies of vascular diseases require strict clinical monitoring due to the high probability of damage to all layers of elastic blood vessels, in addition to damage to the intima(inner layer), it is possible to simultaneously damage the aortic intima and media(inner and middle layers) with myxoid thickening/degeneration then aortic dissection or aneurysms formation in the corresponding localization of arteries , which are mainly accompanied by instability and bleeding. As, diagnosis is based on histology, due to the potential for multiple vessel damage, patients with confirmed intima – media degeneration may require full- body imaging and long – term foll-up to monitor other locations of aneurysm formation. We consider it necessary the patients who have chronic heart failure from young age to consider the expected results in accurate diagnosis based on molecular studies or magnetic resonance angiography, which help both the surgeon and the clinician in avoiding complications.

Conclusion: A rare disease of elastic blood vessels, it can occur in any location and predominantly large arteries is characterized by the deposition of “myxoid” in the intima and media layers (including young people) and leads to aneurysmal formation after myxoid degeneration of the mainly aortic wall. Aortic intima-media myxoid degeneration should be considered as an etiology of acute decompensated heart failure by causing vessels mainly in arteries: dissections, aneurysms, symptomatic aortic regurgitations and aortic valves

insufficiency. Surgical treatment can be complicated by bleeding, which requires caution during surgical intervention and specific postoperative management.

1. In the case of aneurysms of any location, the surgeon should consider the possibility of degeneration and correctly select postoperative treatment tactics to prevent bleeding.
2. Due to the presence of aneurysms at various locations, full body imaging and long-term follow-up are required to monitor the development of aneurysms at other locations.

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