From the Eastern Vascular Society

Intimomedial mucoid degeneration of the peripheral arteries

Georgios Tzavellas, MD,[a] Edvard Skripochnik, MD,[a] David Landau, MD,[a] Reese A. Wain, MD,[b] *and* Apostolos K. Tassiopoulos, MD,[a] *Stony Brook and Mineola, NY*

ABSTRACT

Intimomedial mucoid degeneration, a rare vascular disorder characterized by mucinous deposition in the intima and media layers, causes aneurysmal degeneration of the vessel wall in young patients. Because of the potential for involvement of multiple vessels, these patients may require full body imaging and long-term follow-up. We describe three patients with intimomedial mucoid degeneration and variable clinical presentations. One patient presented emergently with a spontaneously ruptured nonaneurysmal subclavian artery; one patient presented with a known posterior tibial artery aneurysm and new onset of focal pain and paresthesias over the aneurysm; and one patient presented with a self-discovered dorsalis pedis artery aneurysm. (J Vasc Surg Cases and Innovative Techniques 2019;5:452-5.)

Keywords: Intimomedial mucoid degeneration; Aneurysm; Peripheral arteries; Arterial disease

Intimomedial mucoid degeneration (IMD) was first described in 1977 by Decker et al[1] in a case series of nine black patients from South Africa with aortoiliac aneurysms. Since then, only a few case reports and series have been published in the literature describing this pathologic process. Vessels involved with IMD display specific histopathologic findings that include mucoid deposition within the intima and media along with elastic fiber degeneration. IMD should not be confused with other genetic syndromes that cause arterial degeneration, such as Ehlers-Danlos and Marfan syndromes, or with cystic medial necrosis (Erdheim mucoid degeneration). We describe three patients with IMD who were treated surgically. We obtained the patients' consent for the submission and publication of this report.

CASE REPORTS

Case 1. The first patient, a 28-year-old Hispanic landscaper, experienced the sudden onset of right upper chest pain and right neck swelling while using a power leaf blower over his right shoulder. On presentation to the emergency department, he was hemodynamically stable and underwent computed tomography angiography, which revealed a self-contained rupture of the right proximal subclavian artery just proximal to the right vertebral artery origin (Fig 1, *A* and *B*). Through a ministernotomy with right supraclavicular extension, open proximal and distal control of the subclavian artery was obtained. Interestingly, on evacuation of the hematoma, the subclavian artery was found severed with almost clean-cut edges (Fig 1, *C*). A 7-mm ringed polytetrafluoroethylene interposition graft was placed (Fig 1, *D*). Histopathologic examination of the subclavian artery showed mucoid deposition of the intima and the media (Fig 2). After a short, uncomplicated postoperative course, the patient was discharged home on postoperative day 5. The patient was seen at 6-month follow-up with no deficits and underwent arterial duplex ultrasound examination, which showed a patent bypass.

Case 2. The second patient, a 28-year-old white man, had a history of a right ankle fracture sustained at the age of 10 years. He self-discovered a pulsatile mass posterior to his right medial malleolus. After experiencing pain in that area for approximately 1 year, he sought medical attention and was ultimately referred for examination. Arterial duplex ultrasound scanning revealed a focal posterior tibial artery aneurysm surrounded by a plexus of veins. Catheter-based angiography was performed to evaluate the full extent of the aneurysm and to delineate the outflow into the foot (Fig 3). Aortoiliac angiography did not reveal any other aneurysms. An elective aneurysmectomy was performed and an interposition reverse saphenous vein graft was placed. During the dissection, an arteriovenous fistula was identified between the aneurysm and an adjacent vein. The resected aneurysm specimen was sent for histopathologic evaluation, which showed mucoid deposition of the intima and the media (Fig 4). On completion, the patient had a palpable distal posterior tibial pulse and had an uncomplicated postoperative course.

Case 3. The third patient, a 36-year-old woman, presented with a symptomatic right dorsalis pedis artery aneurysm. She underwent elective ligation and excision after noninvasive testing revealed adequate forefoot perfusion from the posterior

From the Vascular and Endovascular Surgery Division, Stony Brook Medicine, Stony Brook[a]; and the Vascular Surgery Division, NYU Winthrop University Hospital, Mineola.[b]

Author conflict of interest: none.

Presented as a rapid fire presentation at the Thirty-second Annual Meeting of the Eastern Vascular Society, Washington, D.C., September 6-8, 2018.

Correspondence: Georgios Tzavellas, MD, Department of Surgery, Stony Brook Medicine, Health Sciences Center, T-19, Rm 090, 101 Nicolls Rd, Stony Brook, NY 11794-8191 (e-mail: giorgostzav@gmail.com).

The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

https://doi.org/10.1016/j.jvscit.2019.04.005

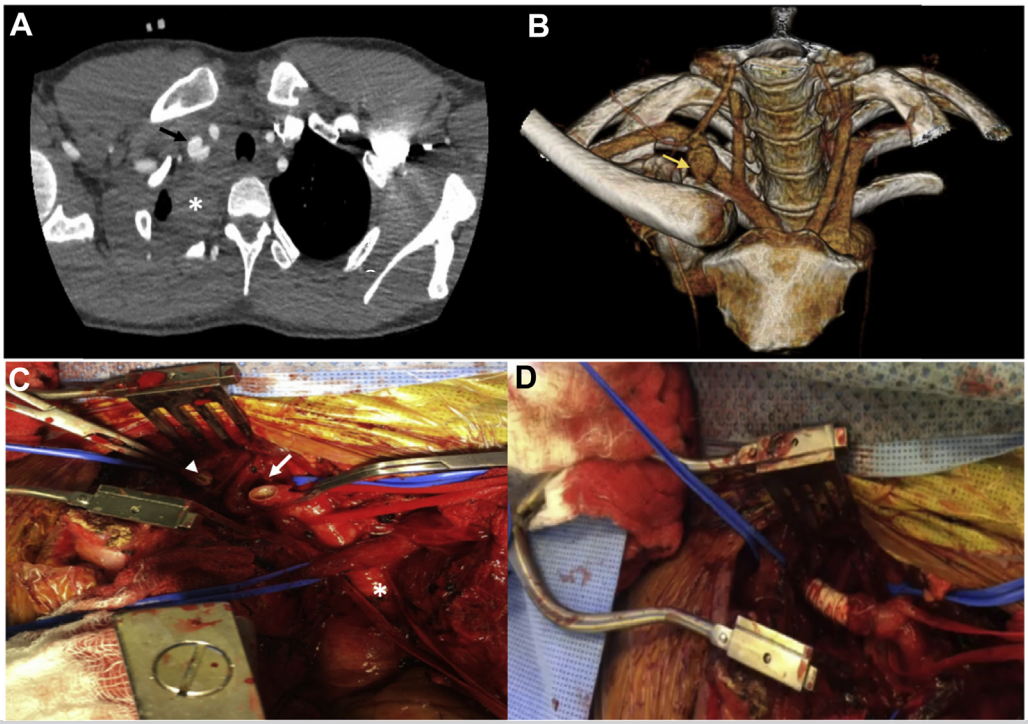

Fig 1. A, Computed tomography angiography (axial image) demonstrating the right subclavian artery pseudoaneurysm (*arrow*) with surrounding mediastinal hematoma (*asterisk*). **B,** Three-dimensional reconstruction of computed tomography angiography image demonstrating the right subclavian pseudoaneurysm (*arrow*). **C,** Ruptured right subclavian artery, proximal edge (*arrow*), distal edge (*arrowhead*), and innominate artery (*asterisk*). **D,** Right subclavian artery repair with interposition ringed polytetrafluoroethylene graft.

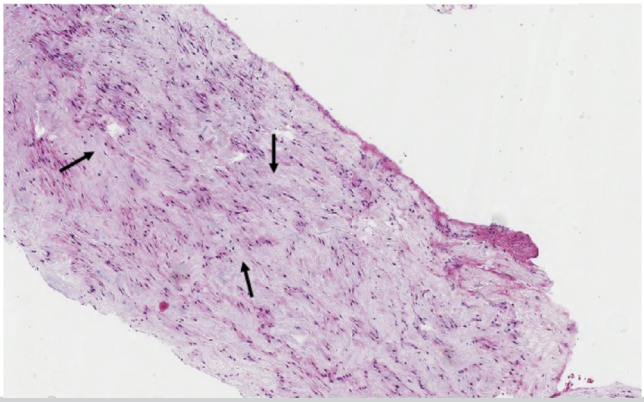

Fig 2. Histopathology image (hematoxylin and eosin stain, magnification ×100) of the subclavian artery showing mucoid deposition of the intima and media (*arrows*).

tibial artery (Fig 5). Histopathologic evaluation of the excised aneurysm revealed mucoid deposition within the intima and the media. The patient was also diagnosed with a superior gluteal artery aneurysm that was embolized through an endovascular approach (previously reported in the literature[2]). Family history of all three patients did not reveal any information about aneurysmal or connective tissue disease. Additional work up included inflammatory and infectious testing markers (CRP, ESR) which were negative for all three patients.

DISCUSSION

Following the first report in the literature by Decker et al[1] in 1977, only a few case series and reports regarding IMD have been published. Costa and Robbs[3] published a series of abdominal aortic aneurysms in the black South African population that included two cases with histopathologic findings of IMD. The first extra-aortic manifestation of IMD was reported in 1993 in a series of six black patients by Cooper.[4] In this study, one patient was diagnosed with a subclavian artery aneurysm, one with a common iliac aneurysm, two with carotid aneurysms, and one with a superior mesenteric artery aneurysm. Interestingly, in the same study, a subclavian artery occlusion without evidence of aneurysm was described in a patient with upper extremity claudication that was treated surgically. Two other subsequent case reports described two patients with brachial artery aneurysms and IMD.[5,6] Collation of the results from all previously reported studies revealed that the abdominal aorta is the most commonly involved artery.

The majority of patients with documented IMD have been black (87%) and tend to be female (74%). However, IMD has been described in men and in other races.[2,6] In our study, which is the largest series in a nonblack population, two of the three patients were male, two were white, and one was of Hispanic descent, supporting the

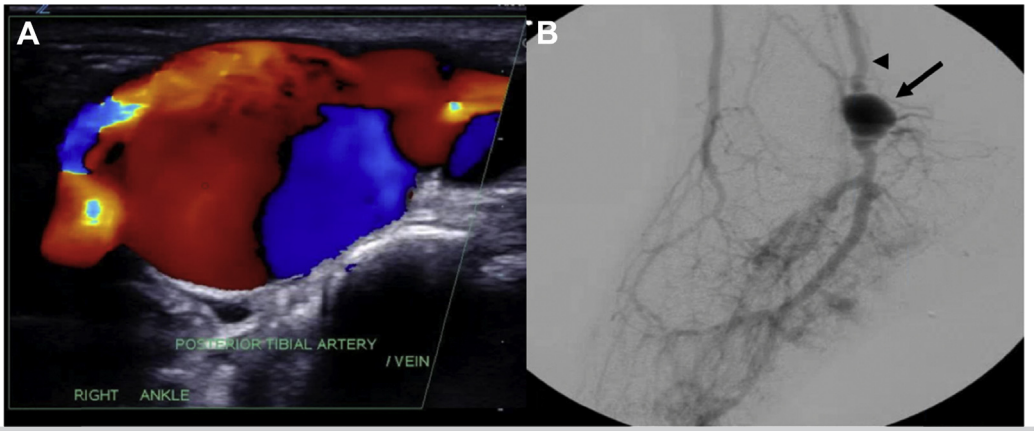

Fig 3. A, Arterial duplex ultrasound image showing aneurysmal dilation of the posterior tibial artery. **B,** Angiogram demonstrating patent posterior tibial artery (*arrowhead*) and the posterior tibial aneurysm (*arrow*).

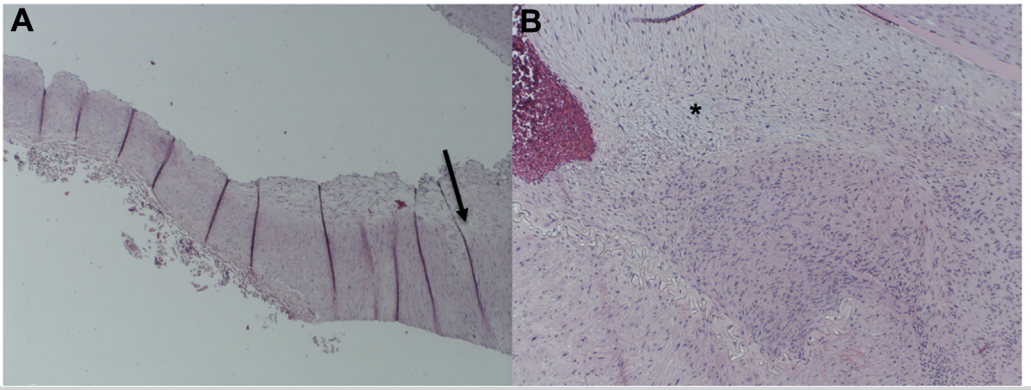

Fig 4. A, Histopathology image (hematoxylin and eosin stain, magnification ×40) of the posterior tibial artery aneurysm showing the mucoid degeneration of the tunica intima and tunica media (*arrow*). **B,** Magnification (hematoxylin and eosin stain, magnification ×200) showing the mucoid deposits (*asterisk*).

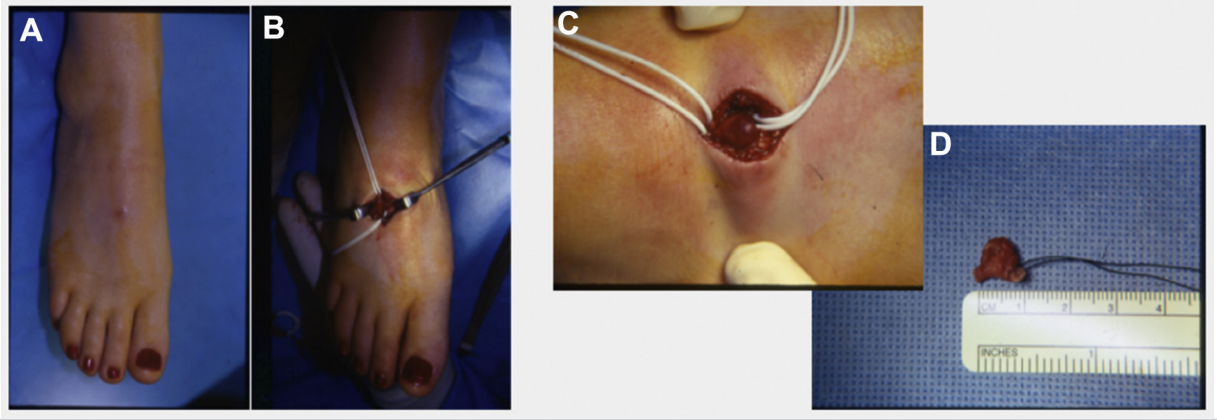

Fig 5. A, Pulsatile mass on the dorsal right foot. **B** and **C,** Dissected right dorsalis pedis artery aneurysm. **D,** Resected right dorsalis pedis artery aneurysm.

belief that the disease is not confined to the black African population. In addition, IMD seems to affect younger patients,[7] which is also supported by our study. The youngest patient ever described with this pathologic process was an infant delivered during gestational week 31 who died shortly after the delivery. An autopsy revealed partial occlusion of the right pulmonary artery with wall abnormalities and histologic features similar to IMD.[8] The same

findings were identified within the superior mesenteric artery.

Patients with IMD and aneurysms may have symptoms dependent on the location of the aneurysm. Patients with abdominal aortic and iliac artery aneurysms have presented with pulsatile abdominal or pelvic masses, abdominal or back pain, and rupture. Local symptoms due to compression of surrounding structures have also been described, such as dysphagia from carotid artery aneurysms.[4] The largest series published by Abdool-Carrim et al[7] included 22 black patients from South Africa with aortic and extra-aortic involvement. Four patients presented with aortic rupture and three with lower extremity ischemia due to dissection, indicating that a significant percentage of patients present with serious and often life-threatening complications. Of all aortic aneurysms contained in the reviewed case series and reports, 18% were ruptured. Rupture can occur in all ages. The youngest patient reported to have a rupture was 17 years old and had concomitant abdominal aortic and common iliac artery aneurysms with rupture of the iliac artery aneurysm.[9] The size of the aneurysm (aortic) at the time of repair has been >5 cm. The largest one reported was 15 cm.[10] Distal embolization of ulnar and radial arteries has been reported in a patient who presented with intermittent upper extremity claudication and a brachial artery aneurysm.[5] Renal function impairment has also been described in a young patient with a complex type II thoracoabdominal aortic aneurysm with multiple dissection flaps and involvement of both renal arteries.[11] Mortality is significant in emergent cases of ruptured abdominal aortic and iliac artery aneurysms.[7,9]

The majority of patients were treated with open surgical repair. During open aortic repair, Abdool-Carrim et al[7] noticed significant bleeding diatheses in three patients with decreased levels of platelets, fibrinogen, and factors V and VIII and absence of intraluminal clot, all of which suggests acute fibrinolytic activity. Only one patient with proven IMD has been treated through an endovascular approach (embolization), which was for a superior gluteal artery aneurysm[2] (patient 3 in our series). Because the aneurysm was left in situ, IMD was not confirmed histologically but was assumed, given the patient's dorsalis pedis artery aneurysm with findings of IMD.

In the largest series by Abdool-Carrim et al,[7] 10 of 22 patients had 11-year follow-up without development of any new aneurysms. In two case reports[9,11] with involvement of two different arteries, the diagnosis was made during the initial workup. The young age of these patients dictates the necessity of surveillance and long-term follow-up with computed tomography scan for the detection of new aneurysmal sites.

The presence of associated arteriovenous fistulas has previously been reported in the literature, similar to our patient with the posterior tibial artery aneurysm.[6] In that case, because of the absence of any trauma, it was thought that the fistulas were congenital and that increased flow with IMD contributed to the formation of the aneurysm. Our patient did indeed experience prior local trauma that might have contributed to the formation of the arteriovenous fistula and then subsequent development of the posterior tibial aneurysm.

CONCLUSIONS

IMD is a histopathologic entity that plays an important role in the development of arterial aneurysms in specific patients. The abdominal aorta is most commonly affected with a significant rate of rupture (18%).[7] Our results indicate that IMD is not confined exclusively to black women; it affects men and different races. Young age is the common denominator in both older and more recent studies. Because of the potential for involvement of multiple vessels, patients with confirmed IMD should undergo full body imaging and long-term follow-up to monitor for the development of aneurysms at other sites.

REFERENCES

1. Decker GA, Samson ID, Schmaman A. Abdominal aneurysm in South African Negroes due to intimomedial mucoid degeneration. Br J Surg 1977;64:513-6.
2. Katz JR, West DL, Bui JT, Knuttinen G, Chejfec G, Owens CA. Endovascular treatment of intimomedial mucoid degeneration. J Vasc Interv Radiol 2008;19:1765-8.
3. Costa M, Robbs JV. Abdominal aneurysms in a black population: clinicopathological study. Br J Surg 1986;73:554-8.
4. Cooper K. Extraaortic intimomedial mucoid degeneration: a clinicopathological study. Angiology 1993;44:477-82.
5. Wali MA, Dewan M, Renno WM, Ezzeddin M. Mucoid degeneration of the brachial artery: case report and a review of literature. J R Coll Surg Edinb 1999;44:126-9.
6. Raber M, Meerwaldt R, van Det R. A young man with intimomedial mucoid degeneration of the brachial artery. J Vasc Surg 2011;53:822-4.
7. Abdool-Carrim AT, Robbs JV, Kadwa AM, Kenoyer G, Cooper K. Aneurysms due to intimomedial mucoid degeneration. Eur J Vasc Endovasc Surg 1996;11:324-9.
8. Zaleski M, Chu Y, Frauenhoffer E, Malysz J, Rassaei N, Abendroth C, et al. A case of vasculopathy of unknown etiology associated with fatal hydrops fetalis and review of the literature on intimomedial mucoid degeneration. Cardiovasc Pathol 2018;33:39-44.
9. Gajjar H, Beningfield S, Naidoo N, Motala A. Multiple aneurysms due to intimomedial mucoid degeneration: a short presentation. S Afr J Radiol 2009;13:98-101.
10. Woolgar JD, Robbs JV. Giant abdominal aortic aneurysm secondary to intimomedial mucoid degeneration. Postgrad Med J 2001;77:782.
11. Viljoen C, Szymanski P, Osman N, Henning KL, Scholtz P, Rayner B, et al. Intimomedial mucoid degeneration causing aortic and renal artery aneurysms in a young adult. Cardiovasc J Afr 2016;27:49-52.

Submitted Jan 9, 2019; accepted Apr 12, 2019.