

Aneurysm caused by intimomedial mucoid degeneration: A case report

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Intimomedial mucoid degeneration (IMMD) is characterised by aneurysm formation following mucin deposition in the intima and media, with elastic tissue degeneration of the arterial wall. IMMD aneurysms have been sporadically reported, with a series of 22 cases by Abdool-Carrim *et al.* emanating from the University of Natal, Durban, South Africa. We report on a patient who presented with a ruptured abdominal aortic aneurysm. IMMD was confirmed on histological examination of a biopsy specimen taken during open repair of the aneurysm.

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Aneurysms due to intimomedial mucoid degeneration (IMMD) are rare. They were first described in South Africa (SA) by Decker *et al.* in 1977.^[1] Subsequently, a case series from the University of Natal was reported by Abdool-Carrim *et al.*^[2] Katz *et al.*^[3] stated that to their knowledge, only 24 cases had been reported in the medical literature at the time of writing. IMMD is characterised by aneurysm formation following mucin deposition in the intima and media, with subsequent degeneration of elastic tissue in the arterial wall. We report on a patient with IMMD operated on at Pietersburg Hospital in Polokwane, Limpopo Province, SA.

Case report

A 49-year-old woman was referred from Tshilidzini Hospital, Shayandima, Limpopo, with a ruptured abdominal aortic aneurysm. On arrival at Pietersburg Hospital, she was hypotensive and had a low haemoglobin concentration and renal impairment. The hospital's massive blood transfusion protocol was initiated and activated. A computed tomography scan performed prior to referral to the Department of General Surgery demonstrated a ruptured infrarenal aortic aneurysm. The patient was prepared for open surgical repair. Intraoperatively, the diagnosis of IMMD was suspected because no intraluminal thrombus was identified, while

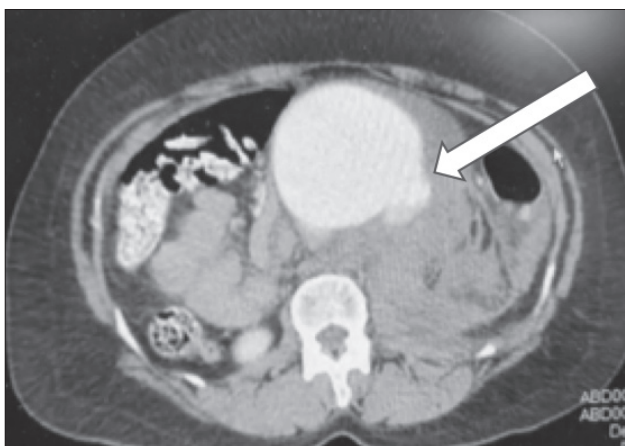


Fig. 1. Axial abdominal computed tomography scan with intravenous contrast, demonstrating the abdominal aneurysm with the retroperitoneal contained leak (arrow).

the aortic wall was found to be thin and frail. The surgical repair was difficult owing to copious oozing from the suture line despite all the necessary precautions. Fresh-frozen plasma and platelets were given intraoperatively, and postoperatively the patient was admitted to the intensive care unit on high doses of inotropes for ongoing resuscitation. During the postoperative period, she developed acute renal failure requiring dialysis. She recovered gradually, was transferred to the normal surgical ward, and was discharged after 3 weeks in a stable condition. She is being followed up and monitored, in particular because of her renal function, which was recovering at the time of writing.

Discussion

IMMD was first described in SA in 1977,^[1] and since then 87% of cases have been reported in black hypertensive females who were relatively younger than patients with atherosclerotic aneurysms. The patients identified by Abdool-Carrim *et al.*^[2] were mostly black women (average age 52 years for patients with IMMD, with a range of 20 - 71 years, v. 65 years for those with atherosclerotic aneurysms), with an unknown aetiology. Our patient, a black woman aged 49 years, fits the previously reported profile. In the medical literature, there have been some reports of IMMD in the non-black population, but very few compared with cases in blacks.

The exact cause of IMMD remains unknown. It has mostly been reported in the large and medium vessels, but Georgios *et al.*^[4] reported a case in the peripheral vessels, indicating that IMMD can affect other vessels, and questioning its earlier description as a disease of large and medium vessels only. IMMD does not have distinctive clinical features, and the diagnosis can only be confirmed on postoperative histological examination of the resected specimen. However, cases of IMMD are rare, and can be missed if the pathologist is not specifically looking for this condition.^[5] If there are clinical grounds for suspecting IMMD, we suggest that this should be mentioned to the pathologist.

Conclusion

IMMD remains a rare disease, with unknown aetiology. It should be suspected in younger patients, particular females, who present with large aneurysms with no other underlying causes. Care should be taken to avoid complications related to this rare disease.

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