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Case report (includes case series that include 5 or fewer patients)

Title: Undiagnosed pulmonary artery hypertension leading to spontaneous Pulmonary Artery Dissection

Description: "Pulmonary artery dissection (PAD) is a life-threatening complication of pulmonary artery hypertension (PAH). Unlike aortic dissections, false lumen in pulmonary artery dissection tends to rupture rather than to develop re-entry site. Given the high mortality of PAD and non-specific presentation symptoms, high index of suspicion and utilizing multimodal imaging is warranted. Early diagnosis of PAH and treatment may avoid this fatal complication."

Undiagnosed pulmonary artery hypertension leading to spontaneous Pulmonary Artery Dissection:

Mohan Gudiwada, Mansi Oberoi, Adam Burdorf

Background:

Pulmonary artery dissection (PAD) is a life-threatening complication of pulmonary artery hypertension (PAH). Unlike aortic dissections, false lumen in pulmonary artery dissection tends to rupture rather than to develop re-entry site.

Case:

23 yrs old male with history of PDA closure at the age of 3 presented to hospital secondary to syncope, dyspnea, and chest pain. CTA revealed PAD. Patient was electively placed on VA ECMO and transferred to our institute. He was emergently taken to surgery and underwent PA dissection repair with Dacron graft and a PFO was created to relieve pulmonary pressures. His post op course was complicated by thrombosis of RVOT and PA as well as thrombosis in the aortic cusp. After a multidisciplinary discussion, his condition was deemed futile and was managed conservatively with anticoagulation. His condition continued to worsen, and family decided to go comfort care.

Discussion:

Though his PDA was repaired at the age of 3, he was lost to follow up until 5 yrs ago when he was evaluated for a syncopal episode. An ECG done at that point suggested RVH but a follow up TTE was reported to be normal. Review of his current CT and echo showed RV hypertrophy, suggestive of chronic PH. Degeneration of tunica media and increased wall stress secondary to PAH is likely the cause of PAD in our patient. Though thought to be rare, review of case reports by Fernando DMG et al. in 2019, showed the incidence of PAD to be on a rising trend, likely secondary to better diagnostic modalities and better care for the patients with congenital heart diseases.

Conclusion:

Given the high mortality of PAD and non-specific presentation symptoms, high index of suspicion and utilizing multimodal imaging is warranted. Early diagnosis of PAH and treatment may avoid this fatal complication.

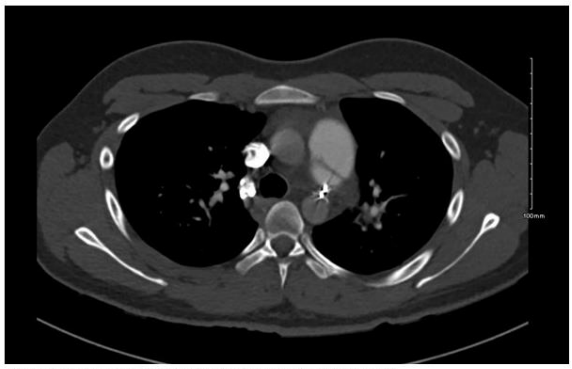

Image 1: CT scan demonstrating PAD and Amplatzer ductal occluding device.

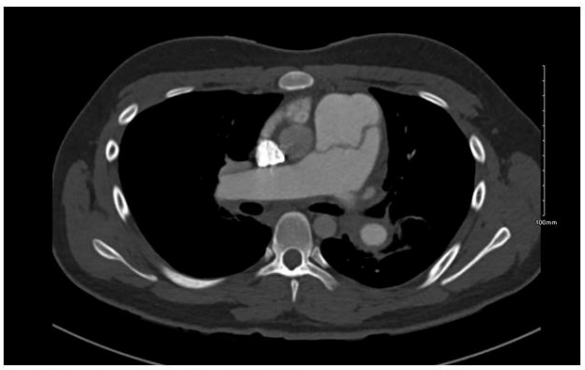

Image 2: CT scan demonstrating maximum extent of PAD

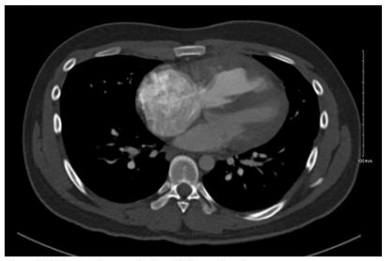

Image 3: CT scan demonstrating RV hypertrophy

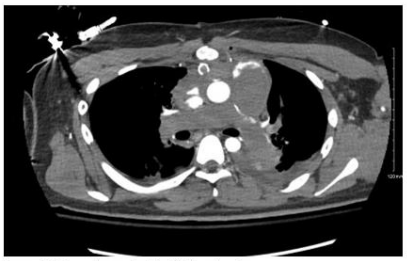

Image 4: CT scan demonstrating PA thrombosis

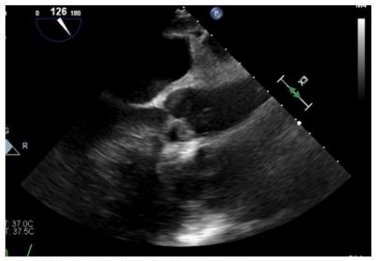

Image 5: TEE showing thrombus in coronary cusp

References:

1. Fernando DMG, Thilakarathne SMNK, Wickramasinghe CU. Pulmonary artery dissection-A review of 150 cases. *Heart Lung*. 2019;48(5):428-435. doi:10.1016/j.hrtlng.2019.02.007