Spontaneous decompression of severe RV to PA conduit pseudoaneurysm with subcutaneous pseudoaneurysm formation in the anterior chest wall in a child with repaired Tetralogy of Fallot with MAPCAs

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Background

Right ventricular outflow tract (RVOT) aneurysms and right ventricle (RV) to pulmonary artery (PA) conduit pseudoaneurysms are rare complications in patients with Tetralogy of Fallot with major aortopulmonary collateral arteries (MAPCAs) who have undergone ventriculostomy.[1,2] There is a paucity of case reports of patients with enlarging RV to PA conduit pseudoaneurysms that extended through the sternum to form an infected subcutaneous pseudoaneurysm in the anterior chest wall.[3]

Case

Our patient was an 8 year 11 month old male with a complex past medical history of DiGeorge syndrome, Tetralogy of Fallot with pulmonary atresia and MAPCAs s/p surgical repair with development of a RVOT and PA conduit pseudoaneurysm, pulmonary hypertension with right heart failure, and hypoxic ischemic encephalopathy who was admitted with fever, dehydration, and hypokalemia secondary to Salmonella species gastroenteritis with bacteremia. He was treated with IV Ceftriaxone and his symptoms resolved with clearance of the bacteremia. As he neared discharge, he developed recurrence of the fever along with erythema and cellulitis on the distal portion of the sternum overlying his previous surgical site. Doppler ultrasound of the sternum revealed the presence of a pseudoaneurysm extending off the previously known conduit pseudoaneurysm that demonstrated the characteristic "ying yang" sign (Figure 1). CT imaging supported the finding of a pseudoaneurysm of the RV to PA conduit with rupture and decompression into the subcutaneous anterior chest wall with subsequent subcutaneous pseudoaneurysm formation with active bleeding (Figure 2). Therefore, we concluded that the sternal abscess found on exam was actually an infected subcutaneous pseudoaneurysm that originated from the conduit pseudoaneurysm which likely had ruptured. Due to the patient's neurological status and the extensive surgery that would be required to definitively repair the pseudoaneurysm, the surgical team and his family concluded that surgical intervention was prohibitively dangerous and unlikely to improve his quality of life. He was treated with antibiotic therapy and symptom management. The subcutaneous pseudoaneurysm drained spontaneously to the skin without exsanguination. Culture of the wound grew P. aeruginosa susceptible to IV Piperacillin-tazobactam. He became afebrile with resolving cellulitis. He was then transferred to another facility in stable condition for further management due to his family's relocation.

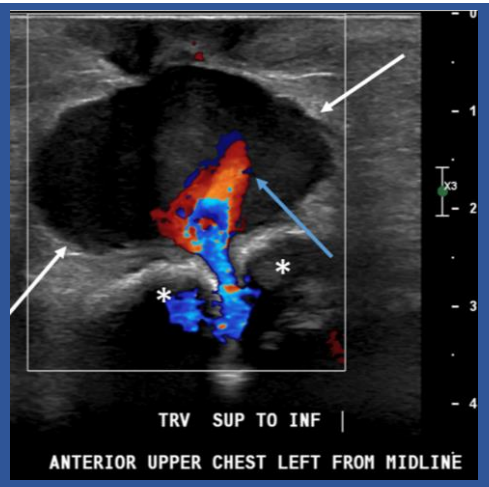

Figure 1: Transverse Doppler ultrasound image performed at the patient's site of concern labeled the upper chest to the left of midline. Round, well circumscribed hypoechoic focus (white arrows) just beneath the skin surface with pulsating Doppler flow (blue arrow) along its inferior aspect – emanating from the beneath the sternum (asterisks).

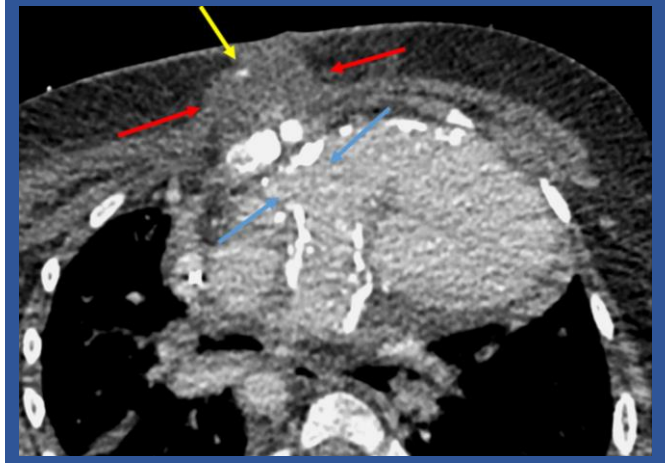

Figure 2: CT of the Chest with IV Contrast: Axial image at the level of the right ventricular outflow tract. There is a pseudoaneurysm (blue arrows) of the RV to PA conduit that has ruptured and decompressed into the subcutaneous anterior chest wall with subsequent subcutaneous pseudoaneurysm formation (red arrows). There is active bleed within this (yellow arrow).

Discussion

Surgical repair of the RVOT is rarely complicated by the development of an aneurysm or pseudoaneurysm. The diagnosis of a pseudoaneurysm after repair of Tetralogy of Fallot occurs at a mean interval of 15 months.[4] The main mechanism is thought be due to the increased RV pressure as a result of persistent obstruction of the RVOT, although pulmonary regurgitation and infection are also recognized to contribute to pseudoaneurysm formation.[4] The development of an infected pseudoaneurysm is a life threatening condition which is worsened if complicated by an extension into the subcutaneous chest wall as in this case.[5] The definitive treatment of pseudoaneurysm formation is surgical intervention.[4]

Conclusion

This case highlights a rare complication of ventriculostomy leading to RV to PA conduit pseudoaneurysm with extension into the anterior chest wall, and progression with secondary *P. aeruginosa* infection. Our patient remained hemodynamically stable with conservative treatment before transferring to another facility.

References

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