Case reports

Late-onset Blalock-Taussig shunt occlusion due to a subclavian artery pseudoaneurysm

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A 3-month-old infant with tetralogy of Fallot presented with progressive severe cyanosis and intractable acidosis about 2 months after a successful modified right-sided Blalock-Taussig shunt. At cardiac catheterization, the suspected shunt malfunction was confirmed. It was due to a bulky, pear-like mass arising from the right subclavian artery and compressing the polytetrafluoroethylene conduit. Any attempt to recanalize the shunt by percutaneous techniques proved unsuccessful. At surgery, a huge dilation of the anterior wall of the right subclavian artery, that sharply bent the prosthetic conduit, was found. Pathologic examination revealed that the compressing mass was pseudoaneurysmal in nature. Despite a second successful shunt operation with a dramatic clinical improvement, the patient died due to multiorgan failure 72 hours following surgery. Extrinsic compression by a false aneurysm is a rare cause of shunt occlusion that should always be suspected in patients presenting with a rapidly progressive shunt malfunction late after a successful shunt procedure.

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Over time, the Blalock-Taussig shunt has established itself as the most cost-effective palliative approach to patients considered unsuitable for the primary repair of congenital cardiac malformations associated with a reduced pulmonary blood flow. The early mortality is acceptable and the long-term morbidity negligible[1-3] and both are mainly due to progressive narrowing or, more infrequently, sudden occlusion. Surgical flaws, resulting in stenosis of the shunt-feeding artery[4] or in kinking of the prosthetic conduit[1,3,5,6], very often cause shunt malfunction. Less frequently, hypercoagulable states[7], local infection[8,9] or extrinsic compression of the prosthesis by a seroma[10,11] or pseudoaneurysm[9,12] may cause shunt narrowing or occlusion. This paper reports on a rare case of late-onset sudden Blalock-Taussig shunt occlusion due to a subclavian artery pseudoaneurysm and reviews the current literature on this topic.

Case report

A 3-month-old infant with tetralogy of Fallot, submitted to a 5 mm modified right Blalock-Taussig shunt at 2 weeks of age, was admitted to our department in critical conditions due to progressive cyanosis lasting a few days. At clinical examination, the child was found to be tachypnoic, hypotonic, severely cyanosed (percutaneous oxygen saturation about 20%), vasoconstricted and hypothermic and with severe mixed acidosis (arterial pH 7.05, pCO_2 86 mmHg, pO_2 21 mmHg, HCO_3 26 mmol/l, base excess -10 mmol/l). His clinical conditions steadily improved after mechanical ventilation, volume replacement, inotropic support and metabolic acidosis correction. At cardiac auscultation, no murmur was audible at the right subclavian area and for this reason shunt malfunction was suspected. Echocardiography did not image any flow through the shunt, but failed to explain the cause of this late-onset shunt malfunction. Then, cardiac catheterization was performed in order to confirm the diagnosis and possibly restore the shunt flow by local thrombolysis or balloon dilation. At right ventricular angiography, the presence of a critical stenosis of the right ventricular outflow tract was confirmed (Fig. 1). Selective angiography of the innominate artery revealed a bulky, pear-like mass arising from the right subclavian artery and compressing the polytetrafluoroethylene (PTFE) conduit (Fig. 2). Any attempt to enter the shunt with a floppy hydrophilic guidewire proved

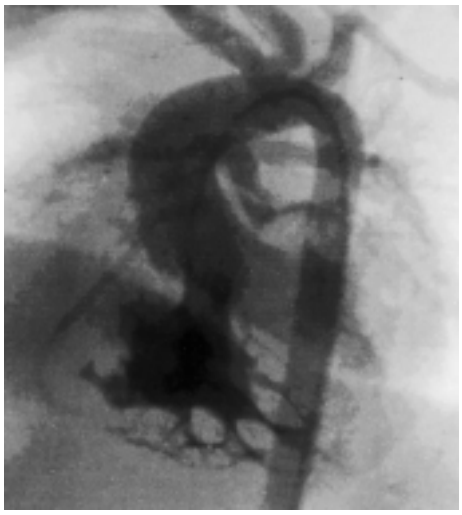

Figure 1. *Right ventricular angiography (sitting-up view) showing a severe right ventricular outflow tract obstruction due to subvalvular stenosis and pulmonary annulus hypoplasia. Even the pulmonary artery trunk and its main branches appear markedly hypoplastic.*

Figure 3. *False aneurysm of the subclavian artery excised together with the compressed and distorted polytetrafluoroethylene conduit.*

to be unsuccessful. Thus, the patient underwent surgery to perform a second shunt on the opposite side and/or to remove the compressing mass. At surgery, performed through a median sternotomy access, a huge aneurysmal dilation of the anterior wall of the right subclavian artery, just proximal to the shunt implantation site, was found. This mass compressed both the distal subclavian artery and the prosthetic conduit, which appeared sharply bent (Fig. 3). The surgical procedure consisted in a 5 mm modified Blalock-Taussig shunt on the left side and in the complete removal of the compressing mass with repair of the subclavian artery. At pathologic examination, the excised mass was found to be pseudoaneurysmal in nature, with a small slit-like feeding entry at the proximal site of the prosthesis implantation. An organized clot extensively coated the inner part of the pseudoaneurysmal cavity and completely occluded the lumen of the PTFE conduit. Despite a sudden dramatic improvement in the child's clinical conditions and oxygen saturation, the patient died 72 hours later of multiorgan failure due to the long-standing hypoxia and acidosis.

Discussion

The Blalock-Taussig systemic-to-pulmonary shunt is almost universally considered as the best surgical palliation for cardiac malformations associated with a reduced pulmonary blood flow in patients who are considered unsuitable for primary repair. Early shunt occlusion or significant narrowing has been reported in up to 7% of patients, accounting for most of the in-hospital mortality and morbidity[1-3,5]. In large series, an unsatisfactory palliation due to shunt malfunction has been reported in up to 40% of patients over the mid-term follow-up[5,6], requiring reoperation in up to 3-20% of patients. Shunt thrombosis is mainly related to sur-

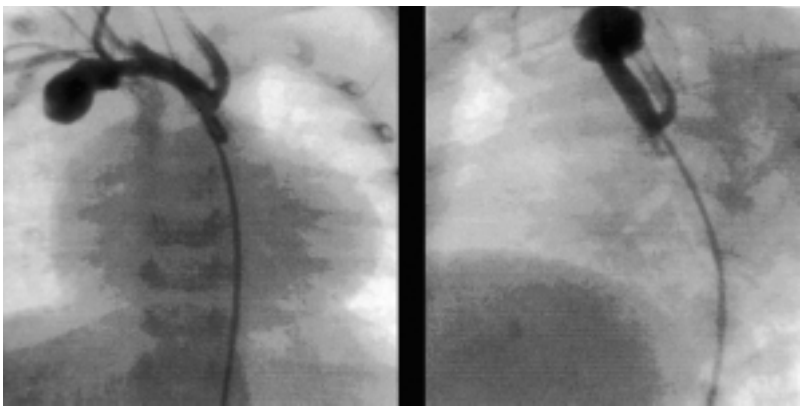

Figure 2. *Selective angiography in the postero-anterior* (left panel) *and lateral* (right panel) *views of the right innominate artery that shows a bulky pear-like mass completely occluding both the distal subclavian artery and the Blalock-Taussig shunt.*

gical damage of the feeding artery or to kinking of the prosthetic conduit[1-6]. Rarely, hypercoagulable states[7], shunt infection[8,9] or extrinsic compression by a seroma[10,11] or pseudoaneurysm[9,12] have been reported as a cause of shunt malfunction and repeat surgery. These complications tend to become clinically evident over a few days or weeks after surgery and manifest as progressive cyanosis and signs of shunt occlusion. In our patient, the shunt malfunction became clinically significant unusually late after surgery, without any apparent triggering factor. It might be hypothesized that the false aneurysm of the subclavian artery slowly and progressively developed after surgery, manifesting once it had reached a critical size. Then, it caused a sudden sharp bending of the modified systemic-to-pulmonary shunt. On the basis of the spatial relationship between the false aneurysm and the prosthetic conduit, it might be suggested that a too proximal extension of the surgical incision at the site of shunt anastomosis damaged and weakened the wall of the subclavian artery. Again, the use of a larger than usual PTFE conduit during the neonatal period might have contributed to weaken the site of shunt implant. Finally, this resulted in progressive periarterial bleeding proximal to the PTFE conduit that became clinically evident when a critical size was reached several weeks after discharge. Despite the timely instrumental confirmation of shunt occlusion and the immediate "bail-out" shunt procedure, the patient died of multiorgan failure, presumably due to the long-standing systemic acidosis.

In conclusion, this rare surgical complication should always be suspected and ruled out in patients with a rapidly progressive shunt malfunction occurring late after surgery. It might be correctly diagnosed at echocardiography showing an abnormal dilation of the shunt-feeding artery responsible for a decrease in the blood flow through the shunt. However, in our opinion, cardiac catheterization is mandatory to confirm the diagnosis and to attempt the restoration of blood flow through the shunt, in order to avoid a new surgical procedure whenever possible. Thus, regardless of the time elapsing from the shunt procedure, and even in the presence of a recent documentation of an adequately functioning shunt, a sudden increase in cyanosis should always raise the suspicion of shunt occlusion.

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