Systemic arterial aneurysm complicated by thrombosis in an infant with resistant Kawasaki disease

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ABSTRACT

Kawasaki disease (KD) is a systemic vasculitis commonly associated with coronary artery aneurysms. Small-sized and medium-sized systemic arterial aneurysms have also been described, particularly in infants and patients with resistant KD. This case illustrates the presentation of a systemic arterial aneurysm complicated by arterial thrombosis and successful interventional management in a young infant.

Keywords: Arterial thrombosis, infant, Kawasaki disease, systemic aneurysm

INTRODUCTION

Kawasaki disease (KD) is a febrile pediatric systemic vasculitis with a predilection for medium-sized arteries, generally the coronary arteries. However, other systemic arteries may also be involved. We present the case of an infant with resistant KD who developed brachial artery aneurysm and thromobosis.

CASE REPORT

A 2-month-old (6 kg) boy was initially diagnosed with KD after 6 days of symptoms and treated with intravenous immunoglobulin (IVIG) (2 g/kg) and high-dose aspirin. Over the 1st month after his diagnosis, he had several readmissions for recurrent symptoms. Although his initial fever subsided with IVIG, he represented a week later with fever, rash, and conjunctivitis. He received a second dose of IVIG (2 g/kg) and remained on high-dose aspirin. Several days later, he developed a new fever, rash, and erythematous lips. He was readmitted and treated with methylprednisolone (30 mg/kg/day) for 3 days. He defervesced with this, but 4 days later, he again developed fever, conjunctivitis, and edema of his feet. He was ultimately treated with infliximab (5 mg/kg) intravenous for one dose and steroids (1 mg/kg/day tapered over 2 weeks). High-dose aspirin was continued from the time of his initial diagnosis through the 1st month. On echocardiogram, his coronary arteries had small aneurysms of the right and left main coronary artery which remained stable.

He remained well for the next 2 months without recurrent fever or symptoms. He then presented at 5 months (7.4 kg) with a 2-day history of intermittent left upper extremity pallor and coolness. He was afebrile with stable vital signs. The left lower arm was cool to touch and appeared pale when the infant was supine but pink when he sat up. There was fullness noted in the left axillary region on palpation. The left brachial pulse was unable to be palpated, while the pulses of the remaining extremities were easily appreciated and this was confirmed by Doppler. There were no signs of conjunctival injection, rash, edema, mucosal changes, or lymphadenopathy.

Computed tomography (CT) angiography of the left arm showed multiple aneurysms in the left axillary artery and a thrombosed aneurysm in the proximal left brachial artery. Angiogram of the left arm confirmed the CT findings [Figure 1] with thrombosis of the brachial artery aneurysm. Collaterals along the upper arm were

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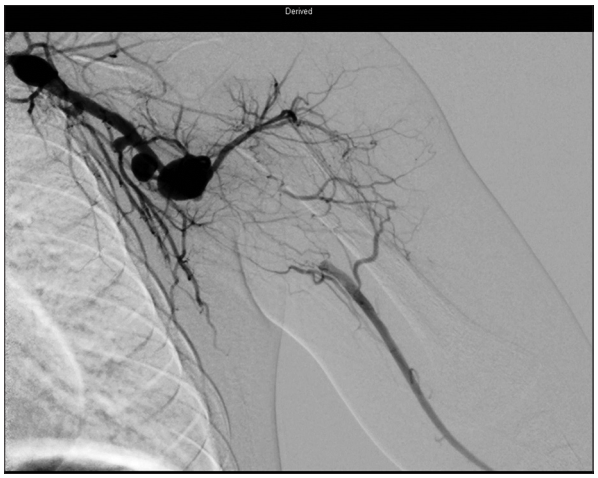

Figure 1: Angiogram of the left upper extremity reveals multiple aneurysms of the left axillary artery and a thrombosed aneurysm at the left axillary–brachial junction. The distal brachial artery remains patent receiving flow from small collaterals

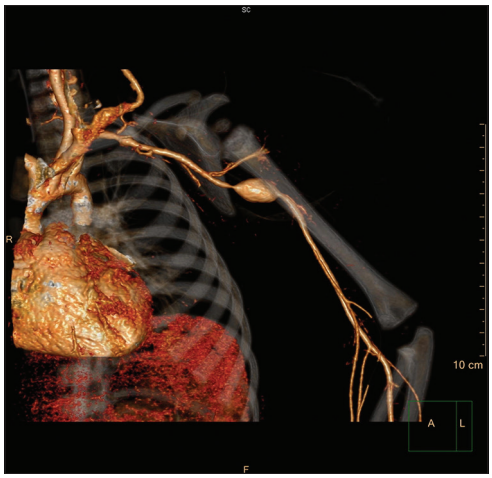

Figure 2: Computed tomography angiogram of the left upper extremity shows resolution of the axillary aneurysms and resolution of the thrombus in the brachial artery though there remains a persistent large aneurysm

present bypassing the thrombosed region but were prone to compression when the infant was supine, thus leading to worsening symptoms when supine. Additional full-body imaging also revealed an aneurysm in the right brachial artery and small aneurysms in the axillary arteries bilaterally. No aneurysms were noted in the abdomen, pelvis, lower extremities, or the head and neck. Rheumatologic and hematologic evaluations were negative for any other etiology for peripheral aneurysms. Given concerns for limb ischemia, catheter-directed thrombolysis with tissue plasminogen activator (tPA) at the site of thrombosis was pursued in addition to systemic heparinization. The patient was started on tPA initially at 0.02 mg/kg/h and then increased to 0.04 mg/kg/h.

After 48 h of treatment with catheter-directed tPA, the patient had a brachial pulse auscultated with Doppler, and there were no further ischemic changes noted to the distal left arm. Intravenous heparin was continued and repeat angiogram still showed thrombus, but with some improvement in flow. The patient was transitioned to enoxaparin. After 6 months, repeat CT angiography of the left arm showed persistent large brachial artery aneurysm, but resolution of the thrombus [Figure 2]. No progressive or giant coronary aneurysms were seen during the disease course.

DISCUSSION

Systemic arterial aneurysms in childhood are uncommon but can be seen in association with infection, trauma, connective tissue diseases, congenital vascular malformations, or vasculitides.[1] KD is one such acute systemic vasculitis with predilection for medium-sized arteries, especially those of the coronary circulation. However, late complications associated with coronary artery aneurysms are more common, and systemic aneurysm formation can also be a significant cause for morbidity as revealed by this patient. Systemic artery aneurysms have been reported to occur in 2% of untreated patients with KD[2,3] and are often detected in infants. The most common areas of distribution include brachial and internal iliac arteries.[3]

Aneurysm-related complications may include progression to rupture, thrombosis, or embolic complications.[4-6] Systemic aneurysms can develop acute thrombosis similar to coronary aneurysms and isolated case reports of infantile KD describe thrombosed aneurysms of the upper extremities leading to bilateral hand loss or finger necrosis.[5,6] Prompt catheter-directed anticoagulation was able to reverse the thrombus and avoid possible ischemic complications.

Predisposing risk factors to systemic complications in this patient include young age at diagnosis and resistant KD. The acute phase of KD is a self-limited inflammatory process of unknown pathophysiology. Intervention during the acute phase of the illness is directed toward limiting long-term complications of vasculitis, most notably coronary aneurysms. Early recognition and intervention with IVIG and aspirin have been shown to decrease the incidence of coronary aneurysm formation from 25% to 3-5%.[7,8] Most patients with systemic arterial aneurysms generally have giant coronary aneurysms >8 mm in diameter;[3] however, this was not the case in the patient.

Infants diagnosed under 6 months are more likely to develop coronary aneurysms as compared to that of the older patients.[9,10] This observation has been largely attributed to delays in diagnosis and prompt treatment due to incomplete clinical presentation.[10] Reports of those diagnosed with systemic arterial aneurysms due

to KD were detected almost exclusively during the first 8 months of life, as in this patient.[3] Although this patient was initially treated in a timely fashion, he had resistant disease that appeared to place him at risk for systemic arterial aneurysms.

This case illustrates the importance of having a high index of suspicion for systemic arterial aneurysms in the high-risk patient with KD. Infants diagnosed with KD and/or patients with resistant KD should be screened on physical examination for pulsatile masses by palpation of the axillary, brachial, abdominal, and inguinal region. CT or magnetic resonance imaging of the systemic arteries, such as the brachial and iliac arteries, as a screening tool should also be considered. Patients with a history of KD presenting with signs of limb ischemia should undergo further evaluation of possible systemic arterial aneurysms and thrombosis. Initiation of catheter-directed thrombolysis for clinically significant systemic aneurysms may be indicated to avoid associated complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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