A case of retiform purpura secondary to sepsis, successfully managed with a combination of antibiotics and steroids


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Introduction
Retiform purpura (RP) is a skin manifestation secondary to blockage of blood vessels in the dermis and subcutaneous tissues. Vasculitis, sepsis and hypercoagulable conditions are common causes of RP. Therefore, it is essential to identify the exact aetiology of RP due to different treatment modalities. We describe a child with severe bacterial infection complicated with RP who responded well to antibiotics and steroids.

Case report
A ten-month-old baby boy presented with high-grade fever for eight days and loose stools for three days. He had no respiratory or urinary symptoms. Initially, the child was admitted to a peripheral hospital, and he was ill and toxic on admission. The child had no fever focus, and he was started on intravenous (IV) cefotaxime pending blood investigations. He had high C-reactive protein (CRP) of 108 mg/dL. The erythrocyte sedimentation rate (ESR) was 48 mm/hr and the procalcitonin (PC) level was 8.8 ng/mL (normal <0.05 ng/mL). The full blood count (FBC) showed anaemia (haemoglobin 8.9 g/dL), thrombocytopenia (platelet count 133,000/cu mm) and neutrophil leucocytosis (total 19,700/cu mm, neutrophils 70%). The blood picture showed severe bacterial infection without any evidence of disseminated intravascular coagulation (DIC), but the blood cultures remained sterile. Since the child remained ill, he was transferred to Teaching Hospital Karapitiya on the following day.

On day-9 of the illness, the child developed a non-blanching rash, suggestive of retiform purpura, over the lower limbs. It gradually spread over all four limbs, buttock and right pinna. Subsequently, he developed peripheral cyanosis with necrotic fingers and toes (Figure 1). However, the severity and the distribution were not compatible with the diagnosis of purpura fulminans. His blood pressure remained normal throughout the hospital stay.

He had persistently elevated CRP and ESR counts despite being on IV antibiotics. However, his subsequent procalcitonin levels came down to normal (0.025 ng/mL). His FBC showed persistent neutrophil leucocytosis and thrombocytopenia. He had normal coagulation profile (International normalised ratio 1.2, activated partial thromboplastin time 28.7 seconds). His fibrinogen level was 3.1 g/L (normal 2.0-4.0 g/L) and D-dimer level was 3.23 mg/L (normal <0.5 mg/L). His blood, urine and cerebrospinal fluid cultures remained sterile. He had negative antibodies and the rapid antigen test for Covid-19 was negative. He had a negative anti-nuclear antibody test. The rest of the autoimmune panel was not done due to a lack of resources. He was started on IV immunoglobulin (2g/kg) while continuing IV antibiotics, for which there was no response. A skin biopsy was performed, which showed fibrinoid necrosis of blood vessels in the upper and deep dermis and multiple occlusive fibrin thrombi (Figure 2). Moreover, mild to moderate neutrophil infiltration was noted in the vessel wall without features of leucocytoclasis. The biopsy favoured the diagnosis of sepsis-induced vasculitis over other medium vessel vasculitis such as polyarteritis nodosa (PAN).

After performing a bone marrow biopsy, he was started on IV methylprednisolone daily (30 mg/kg/day). He showed marked improvement for steroids. The steroids were tapered off gradually and stopped after two weeks. The child was discharged home after ten days of admission and reviewed in two weeks (Figure 3). He remained well, and all the inflammatory markers have come down.

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Figure 1: 10-month-old baby with retiform purpura complicated by gangrenous toes, fingers & plaques.

Figure 2: Skin biopsy showing focus of vasculitis with fibrin thrombi inside the lumen.

Figure 3: Skin appearance after 2 weeks of steroids.
Discussion
RP differs from other reticular eruptions due to vascular pathologies such as livedo reticularis and livedo racemosa because of its persistent nature. Blood vessel damage in RP could occur due to two main reasons; firstly, vessel wall damage due to vasculitis, secondary to sepsis or autoimmune conditions; secondly, it could be due to obstruction of the vessel lumen due to thromboembolic diseases. Hence, skin biopsy plays a crucial role in identifying the underlying pathophysiology of RP. However, certain disorders such as sepsis, cryoglobulinaemia and levamisole induced RP can present with both vessel wall abnormalities. In our case, the biopsy findings showed vessel wall abnormalities with minimal thrombus formation in the vessel lumen, highly suggestive of sepsis induced vasculitis. Therefore, it is unlikely to be due to hypercoagulable state or autoimmune vasculitis. Though this child had features of bacterial infections, the exact organism was not isolated. Covid-19 has been identified as a recognized cause of RP in children. However, in our case, Covid-19 antibodies were negative. Medium vessel vasculitis such as PAN and Kawasaki disease are important aetiologies of RP. In addition, PAN could be associated with streptococcal infections. However, in our child, features suggestive of vasculitis such as haematuria, thrombocytosis and eosinophilia were not found. Moreover, the histology did not support the diagnosis of vasculitis.

Mainstay of treatment for vasculitis induced RP is immunosuppressive drugs. Though this child presented with features of sepsis, skin changes and ongoing fever, despite being on broad spectrum antibiotics, could not be explained by the sepsis. Hence, he was started on IV methylprednisolone while continuing antibiotics. PC remains a marker of invasive bacterial infections in children. This child had a high PC level at the beginning but by the time he developed skin changes, his PC level came down despite elevated CRP and ESR.

Conclusions
Retiform purpura is a rare complication of severe bacterial infections in children. Though rare, PAN should be considered as a differential diagnosis since it is associated with streptococcal infections. Steroids remain the first line treatment for vasculitis; however, it is essential to decide the timing of starting steroids to prevent invasive bacterial infections. PC level is helpful to differentiate infections from inflammatory conditions. Skin biopsy is an important investigation to identify the cause of RP.

References