Atypical Kawasaki disease: Diagnosis underneath diapers

We report a 5-month-old baby, who presented with fever, irritability with rash for a total duration of 10 days, cough, and loose motions for 2 days. He was being treated as sepsis with broad-spectrum antibiotics before being referred to us. There were no history of erythema of tongue, conjunctival congestion, breathing difficulty, altered sensorium, or convulsion. Past history and perinatal history were unremarkable. On admission, the patient was febrile, irritable, and vitals were stable. Anthropometric measures including weight (=6 kg) and length (=63 cm) of the baby were within normal range as per WHO standards. General physical examination revealed presence of urticarial rash, unilateral cervical lymphadenopathy, and edema of hands and feet but there were no oral or eye changes nor any periungual desquamation. On further examination and unfolding the diapers, we noted scrotal edema with perineal maculopapular rash which led us to consider Kawasaki disease (KD) as a possibility [Figure 1]. Laboratory evaluation revealed raised erythrocyte sedimentation rate (ESR) and C-reactive protein (50 mm/1st h and 34 mg/dL, respectively), anemia (Hb 5.8 g/dL), neutrophilic leukocytosis (20,200/mm^3), and thrombocytosis (platelet count: 6,00,000/mm^3).

Ultrasoundography of scrotum was suggestive of bilateral hydrocele. Echocardiography did not show any abnormalities consistent with KD. As our case did not met all the criteria for KD, a label of incomplete (or atypical) KD was considered as per American Heart Association (AHA) guidelines and patient was started with intravenous immunoglobulin (IVIG) (2 g/kg over 12 h) and aspirin (75 mg/kg/day) on day 1 of hospitalization. After 24 h of IVIG therapy, the patient became afebrile, irritability reduced, and scrotal edema resolved in 48 h [Figure 2]. Repeat inflammatory markers (ESR, CRP, leucocyte count) were normal after 72 h although thrombocytosis (500,000/mm^3) continued. Blood culture was sterile. The patient was discharged on antithrombotic doses of aspirin. At 2 months follow-up, the child is doing fine and afebrile with normal echocardiography and inflammatory markers were normal.

KD is a self-limited vasculitis of unknown etiology often preceded by symptoms of upper respiratory or gastrointestinal illness like in our case. Our patient had both upper respiratory infection and gastroenteritis at presentation. In the absence of classical features, these children often get misdiagnosed as sepsis and receive unwanted antibiotics without any benefit. The persistence of fever, presence of skin rash, edema of hands and feet and the scrotal edema pointed toward underlying vasculitis in our case.

Presence of scrotal swelling and pain due to testicular inflammation is characteristic of vasculitis including polyarteritis nodosa (PAN) (6%), Henoch-Schönlein Purpura (2%–38%) and KD. Presence of rash especially over perineal area is seen in KD in initial few days of illness. The appearance of scrotal edema in KD ranges from 4 to 18 days. It is important for the physicians to know about this finding in KD to avoid unnecessary surgical exploration.

Diagnosis of KD remains a challenge, mainly when a child presents with incomplete or atypical features. A meticulous clinical examination can reveal clues such as scrotal edema and perineal rash to enable early diagnosis and timely initiation of therapy to prevent long-term complications.

Figure 1: Presence of scrotal edema and perineal rash

Figure 2: Scrotal edema has resolved in 48 h after IVIG therapy
Declaration of patient consent
The authors certify that appropriate patient consent was obtained.

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Conflicts of interest
There are no conflicts of interest.

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