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Case Report

Shoulder joint septic arthritis with scapular osteomyelitis a rare presentation in an infant with sickle cell disease: a challenging diagnosis; a case report from rural central India

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ABSTRACT

Sickle-cell disease (SCD) a hereditary autosomal recessive disorder is the most common haemoglobinopathy worldwide. In India, it is the second most common haemoglobinopathy next to Thalassemia, prevalent in the tribal population of Central and Southern parts of India. The pathophysiology of the disease is point mutation in the beta globin chain leading to sickling of RBCs which causes obstruction in microvasculature leading to acute events like Vaso-occlusive crisis. Patients with SCD are also at an increase of orthopaedic manifestation like osteomyelitis, septic arthritis or osteonecrosis. We report a case of a 10 months old male child who presented with anaemia and fever. Child was diagnosed with SCD, later developed swelling of right shoulder joint and restrictions of movements. Diagnosis of septic arthritis of shoulder with scapular osteomyelitis was made with the help of radiological and laboratory investigations. Child was managed with injectable antibiotics and symptomatic treatment. Patients with SCD as a result of occlusion of microvasculature along with immunocompromised state are at higher risk of bacterial infections. Osteomyelitis is one of the dreaded complications. Its clinical presentation is similar to that of VOC hence there occurs a dilemma in diagnosis. A multidisciplinary approach including high degree of clinical suspicion, laboratory investigation and radiological imagining can help in early diagnosis and management.

Keywords: Osteomyelitis, SCD, Septic arthritis, Vaso-occlusive crisis

INTRODUCTION

Sickle-cell disease (SCD) a hereditary autosomal recessive disorder is the most common haemoglobinopathy worldwide. High incidence seen in the sub-Saharan Africa, Saudi Arabia, India and Mediterranean Countries. In India, it is the second most common haemoglobinopathy next to Thalassemia. The disease is most prevalent in the tribal population of Central and Southern parts of India.[1]

Sickled haemoglobin occurs as a result of a point mutation at the 6th codon of the Beta globulin gene resulting in a single base pair change, thymine for adenine. The change encodes valine for glutamine in the 6th residue of Beta-globulin molecule. The structural change causes sickling of the RBCs, which gets precipitated in hypoxic conditions, acidosis, cold stress, decrease in 2,3-diphosphpglycerate concentration and increase in carbon monoxide concentration. The clinical presentation of SCD patients is extremely varied. The most common presentations being Vaso occlusive crisis (VOC), haemolytic anaemia and haematological infections. Patients with SCD are also at an increase of orthopaedic manifestation like osteomyelitis, septic arthritis or osteonecorisis.[2] Osteomyelitis is one of the most serious and potentially disabling orthopaedic

manifestations of SCD, but its differentiation from benign SCD conditions such as VOC can be difficult.[3]

In SCD, the accumulation of sickled red cells can cause occlusion of the microvasculature, causing acute clinical event. Such patients present with severe pain, swelling, fever and raised inflammatory markers. Repeated episodes can lead to bone infraction and necrosis creating a favourable environment for bacterial growth. Immunodeficiency caused by splenic dysfunction, tissue infarction, and excess iron content leads to an increased risk of osteomyelitis.[4]. It most commonly affects Tibia, Diaphysis of femur, humerus and vertebra. Radiographic findings such as osteopenia, sclerosis, and periosteal inflammation are seen in both stages of infection and infarction. Therefore, the radiographic features are nonspecific and primarily normal.[4] High decreased of suspicion with early diagnosis of osteomyelitis can help prevent severe bony destruction and life-threatening complications. We report a case of 10-month-old male child from Vidarbha region, Maharashtra with SCD having septic arthritis with scapular osteomyelitis.

CASE REPORT

A 10-month male child presented to paediatric unit with chief complaints of high-grade fever for 1 week. On examination child was afebrile. Pallor was present, no icterus or signs of bleeding. Vitals were stable with Heart rate 103/min, respiratory rate of 22/min BP 95/60 mmHg. Systemic examination was normal. On further enquiry, there was positive family history of sickle cell trait in the mother. Child was evaluated for acute febrile illness and for anaemia. Blood workup revealed, haemoglobin of 7.3 g/dl, RBC count 4.1 million/ µL, Total leucocyte count 15,580 cells/µl and platelet count of 1,17,000 cells/µL. Peripheral smear showed presence of microcytic hypochromic red cells along with some sickle RBCs. Anemia workup revealed serum iron 33 µg/dl, serum Vitamin B12 227 pg/dl. HPLC reports suggested HbA 3.8%, HbA2 2.6%, HbF 14.7% and HbS 78.9%. Inflammatory markers were raised. C-reactive protein (CRP) was 146 mg/l, serum ferritin 221.9 ng/ml. Blood culture and urine culture was sent. Considering septicaemia in a case of SCD, child was treated with injectable antibiotics and continued till culture reports were available which were sterile. Child responded well to treatment and was discharged after 10 days of hospital stay. He was started on iron, vit b12 and folic acid supplements. He was closely monitored in the outpatient department.

Patient followed up after 3 days, when mother notice child had pain, swelling with limitation of movement over right shoulder. Child was readmitted, considering vaso occlusive crisis. He was managed with IV fluid and analgesic for pain relieve. An X-ray of the right shoulder done which was suggestive of a heterogenous increased density diffusely involving right scapula. New bone formation, in the form of periosteal reaction was also present in the lateral margin of scapula. Homogeneous increased density within the soft tissue adjacent to the right shoulder joint was present. Features were suggestive of possibility of right scapular osteomyelitis with acute with right gleno-humeral septic arthritis. (Figure 1) ultrasonography of right shoulder joint was done which was suggestive of minimal right shoulder joint effusion. MRI of the shoulder joint was done to confirm the diagnosis. MRI findings showed presence of altered signal intensity peripherally enhancing of approximately 1.4×1.1×0.8 cm, volume 0.6 ml in deltoid muscle, diffusion restriction on DWI/ADC sequence. Hyperintensity was noted in the infra-scapular region of right scapula and post enhancement noted in all muscles of rotator cuff (Figure 2 and 3). No fracture or osteonecrosis changes were seen. Features were suggestive of septic arthritis with infective collection in the intramuscular plain.

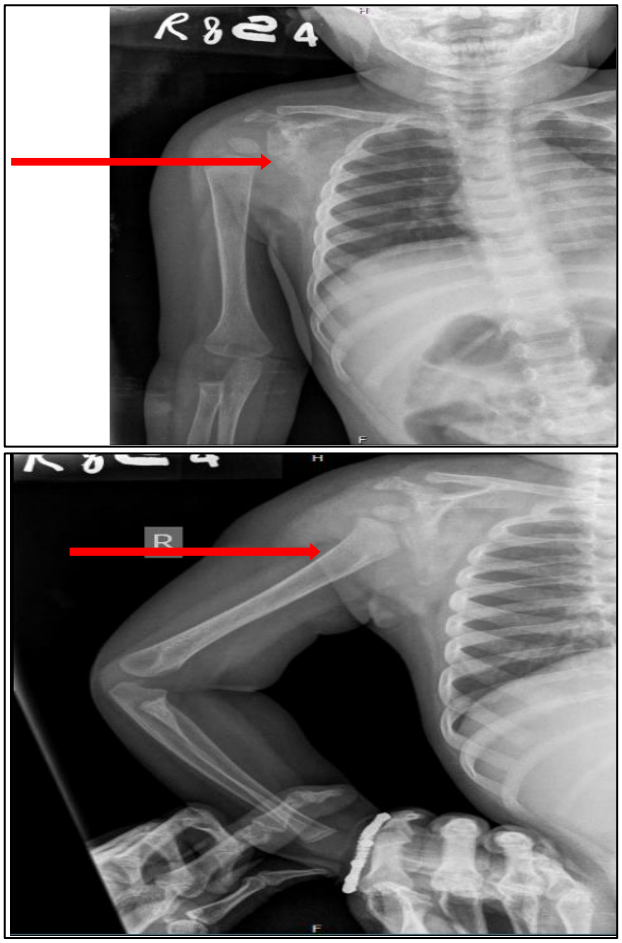

Figure 1: Heterogenous increased density seen diffusely involving the right scapula with indistinct lateral margin of right scapula, new bone formation seen in the form of periosteal reaction along the lateral margin of scapula, homogenously increased density within the soft tissues adjacent to the right shoulder joint and slightly homogenously increased density is seen involving the metaphysis and epiphysis of right humerus with irregular metaphyseal margins.

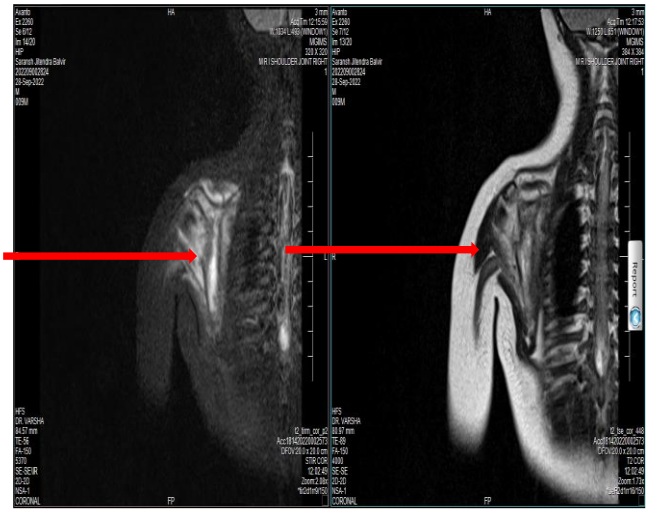

Figure 2: T2/STIR hyperintensity noted in infra-scapular region of right scapula.

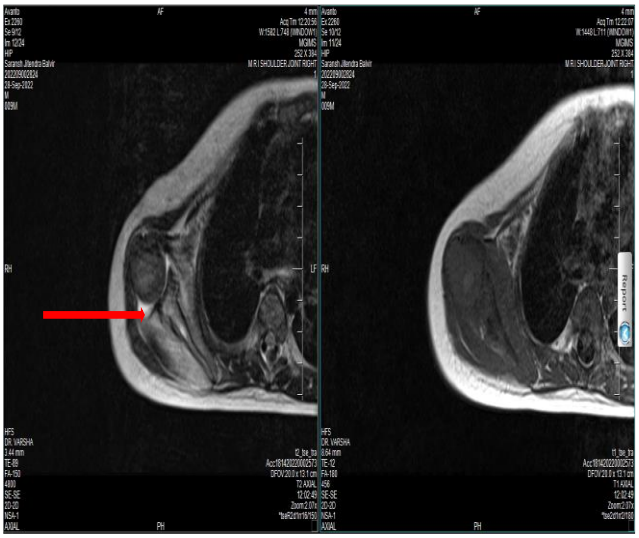

Figure 3: Fluid intensity collection noted in bicipital groove. Fluid intensity collection noted in subacromial-subdeltoid bursa and in right shoulder joint.

The case was discussed with a paediatric orthopaedic surgeon, considering osteomyelitis with septic arthritis patient was started on injectable broad-spectrum antibiotics, vancomycin (200 mg/kg/day) and ceftriaxone (100 mg/kg/day). Antibiotics were given for a total of 4 weeks. For pain, child was initially managed with NSAIDS but as the pain was severe, patient was given injectable tramadol (opioid analgesic) at 1 mg/kg three times a day. After 3 weeks of treatment, laboratory investigations were repeated which showed a decrease in CRP level (3.15 mg/L) and normal total leucocyte count (12,600 cells/µ). Child showed good response to treatment. Fever and pain subsided and child regained movement over his shoulder joint. Child was discharge and keep on the close follow up in the outpatient department.

DISCUSSION

We reported a case of a 10 months old male child who present with complaints of fever and anaemia. Child was diagnosed with SCD and given treatment for anaemia and septicaemia. Child later presented with complaints of pain and inability to mobilise right shoulder. A diagnosis of VOC was initially made but later was found to have right shoulder septic arthritis with scapular osteomyelitis.

SCD has a wide spectrum of clinical presentation, ranging from benign to life threatening complications. The immunocompromised state of SCD (due to hypo-splenism, defective compliment system) along with microtrauma following VOC, encourages the spread of infection during bacteraemia. Osteomyelitis often occurs in the long bones (in our case the Scapula was affected) in which due to looping of vessels in metaphysis there is slow flow of blood. In osteomyelitis, bone marrow is the primary site of infection. It may involve any part but metaphysis of long bone, preferentially adjacent to joints.[5] Bacterial osteomyelitis of the scapula is rare in paediatric patients, with frequency varying from 0.5% to 2.6%.[6]

Diagnosing osteomyelitis in children with SCD may be extremely difficult, as they mostly present with fever, pain, swollen, tender limb, with decreased range of motion. This presentation is similar to the VOC. There were no definitive features on history, physical examination, laboratory testing, that could reliably differentiate between osteomyelitis and VOC in our patient. In the early stages of osteomyelitis and VOC, plain radiographs are usually found to be either normal or only showing soft-tissue edema, periostitis, or osteopenia. The lytic changes suggestive of osteomyelitis lag at least 2 weeks behind the process of the infection on radiographs.[7] Thus this prompts the need of other imaging modalities. MRI is the imaging modality of choice for diagnosis of osteomyelitis, sensitivity has been reported up to 100%.[8] Radionuclide scans may be used in situations when the diagnosis of osteomyelitis remains unclear.[7] Given the high diagnostic accuracy of MRI, it is much less commonly used.

The optimal treatment of osteomyelitis includes a combination of antibiotics with adequate antimicrobial coverage and, if necessary, surgical management through debridement and wound reconstruction.[9] The typical duration of parenteral antibiotic treatment of osteomyelitis runs 4 to 6 weeks.[7] Some trials of extended courses of either parenteral or oral antibiotics have not suggested improved outcomes compared with 6 weeks of therapy.[10] S aureus, Salmonella, and other gram-negative bacilli are the most commonly isolated organisms from individuals with osteomyelitis in the SCD population. In an article published by Almeida and Roberts, recommended first-line treatment of confirmed or suspected osteomyelitis to be a third-generation cephalosporin to ensure coverage of the above-mentioned organisms.[11] Another reasonable combination for empiric

antibiotic treatment is a combination of vancomycin and ciprofloxacin.[12] Ideally, the results of a positive blood culture, biopsy, or aspiration should be used to direct the choice of antibiotics.

In our case, a diagnose was made through a combined approach of through clinical history, physical examination with laboratory investigations and imaging technology. Child was managed with IV hydration, analgesics for pain and broad-spectrum antibiotics for a duration of 4 weeks.

CONCLUSION

SCD patients are at a higher risk of bacterial infections. Sickle red cells causing occlusion of microvasculature along with their immunocompromised state make the ischemic bone marrow a hub for the bacterial to grow. Osteomyelitis is one of the dreaded complications of SCD. Its clinical presentation is similar to that of VOC hence there occurs a dilemma in diagnosis. A multidisciplinary approach including high degree of clinical suspicion, laboratory investigation and radiological imagining can help in early diagnosis and management. Early and intense treatment is thus required to prevent the crippling complications of osteomyelitis and septic arthritis.

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