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Unusual Presentation of Arthritis in an Immunodefecient Child

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Abstract: Arthritis may be the first clinical manifestation of primary hypogammaglobulinaemia. We report a case of primary hypogammaglobulinemia who presented with migratory polyarthritis and sore throat. The diagnosis of primary hypogammaglobulinemia (Brutons disease) was delayed as it was previously thought it was migratory polyarthritis, due to Rhematic fever, Which is very common in Asian countries. These patients are likely to develop recurrent infections complicated by arthritis. This delay resulted in considerable joint destruction. The measurement of serum immunoglobulin concentrations readily differentiates immunodeficiency from conditions such as Stili's disease and dictates subsequent management. The importance of presenting this case was that one should be in look out for something more in cases where child is not responding well to antibiotics and getting various bacterial infections on and off.

Keywords: Child, Septic Arthritis, Psedomonas.

INTRODUCTION

Primary hypogammaglobulinaemia was defined as a serum IgG concentration of less than 2-0 g/l with impaired immunoglobulin synthesis owing to a defect of unknown aetiology in the B lymphocytes or plasma cells [1]. Patients who have hypogammaglobulinemia have a typical presentation with recurrent bacterial infections which usually involve respiratory tract [2]. XLA (brutons agammaglobulinemia) is caused by a mutation on the X chromosome identified in 1993 which produces an enzyme known as Brutons tyrosine kinase (Bruton's disease) or common variable immunodeficiency; they may also have arthritis as a presenting or complicating factor, these patients do not generate mature B cells which manifests as a complete lack of antibodies in their blood stream. Patients with untreated XLA (brutons agammaglobulinemia) are prone to develop serious and fatal infections. It is first known immune deficiency and is classified with other inherited (genetic) defects of immune system. Btk is particularly responsible for mediating B cell development and maturation through a signaling effect on the B cell receptor BCR. Patients typically present in early childhood with recurrent infections, in particular with extracellular, encapsulated bacteria [3]. It occurs in a frequency of about 1 in 100,000, male newborns, and has no ethnic predisposition. X linked hypogammaglobulinaemia (Bruton's disease) was diagnosed according to the following criteria of the World Health Organisation, male patient; onset of disease in infancy or early childhood; serum IgG concentration less than 2-0 g/l, with appreciably decreased IgA and IgM concentrations for age; absence of functional antibodies; normal cell mediated immunity; and pre-B cells present in bone marrow but no mature B cells in peripheral blood [4]. This child suffered from recurrent bouts of respiratory tract infections, ottitis media and urinary tract infections from the age of one and half years. He presented with typical symptoms of migratory polyarthritis, hence initial diagnosis of Rheumatic fever was made and he was treated for the same.

CASE REPORT

A four year old boy was brought with history of on and off complaints of fever, history of inability to walk and history of sore throat. Our patient was the eldest male child offspring born out of a consanguineous marriage. He was full term normal delivery. At the time of birth, baby weighed about 2.7 kg. Patient was normal for his age, height and weight. His weight was 15 kg and height was 99 cms. There was no history of delayed milestones. There were no major complaints till the age of 1 year. On examination there was swelling and tenderness at wrist joint, knee joint. He had skin lesions all over the body (old pyoderma lesions). The vitals were within normal

limits, so were the other systems. There was a past history of empyema, two years back, patient also gave the history of admission to the hospital for urinary tract infection an year back and history of ottis media six months back. There was no relevant family history. There was no history of any abnormal rashes on body, nor was there any history of pain involving any small joints in the body. His blood investigations revealed, a hemoglobin (10 g/dl), total leukocyte count of 41600/mm^3 with differential count of P 92%, L 5%, E 03 % and peripheral smear showed a picture of microcytic hypochromic anaemia with neutrophilic leucocytosis, hysegmented neutrophils seen. Reticulocyte count was 1 %. Platelets were adequate. Negative for any parasites. His ESR was 49 mm at end of hour and Serum chemistry ([blood urea nitrogen: 16 mg/dl, serum creatinine: 0.7 mg/dl, serum glutamate oxaloacetate transaminase (SGOT): 20 IU/l, serum glutamate pyruvate transaminase (SGPT): 25 IU/l. Total Proteins – 6.4 gm/dl Albumin 4.3 gm/dl all blood chemistry parameters were within normal range for age. Urine showed presence of 15- 20 RBC, His CRP was 1.2 mg/dl. RA test was negative. Montoux test was negative. The sickling test was negative, Hb electrophoresis was done to rule out thalassemia, which did not show any abnormality. He was also tested for HIV which was non reactive. However his blood culture tested positive for Psedomonas and during his entire stay in hospital he developed recurrent spikes of fever. X ray chest was WNL, X ray wrist, bilateral mastoid, Knee and Forearm showed signs of septic arthritis. CT thorax revealed consolidation in anteroposterior basal segment of left lobe with enlarged lymph nodes. Sonography of Ischiopubic rami showed ischaemic changes and osteomyelitic chages. X ray of right femur showed fluid in right iliac fossa with right femoral Lymph nodes. Bone scan(Fig1) showed infective etiology of right acetabulum, ischemium and pubic bones. MRI pelvis showed moderate right hip joint effusion with myositis, fasciitis with mild right knee joint effusion.MRI hip showed increased hip joint effusion, myositis, osteomyelitis (right femur) Iliac fossa(right) , free fluid and presence of femoral lymph nodes. MRI wrist showed signs of septic arthritis. Right hip arthrotomy was done which revealed septic arthritis. Blood Immunoglobulins study was done which revealed all were below the normal ranges. IgG 31.2 g/l, IgA 5.78 g/l, Ig M 4.23 g/l, Ig E 5.3 g/l Ultra Sonography , ECG, ECHO and blood gases showed no abnormality. The child was treated with intravenous antibiotics Vancomycin, Maropinem and NSAID. On this regimen his arthritis decreased and he had no further episodes of arthritis. However the infection had severely damaged his hip.

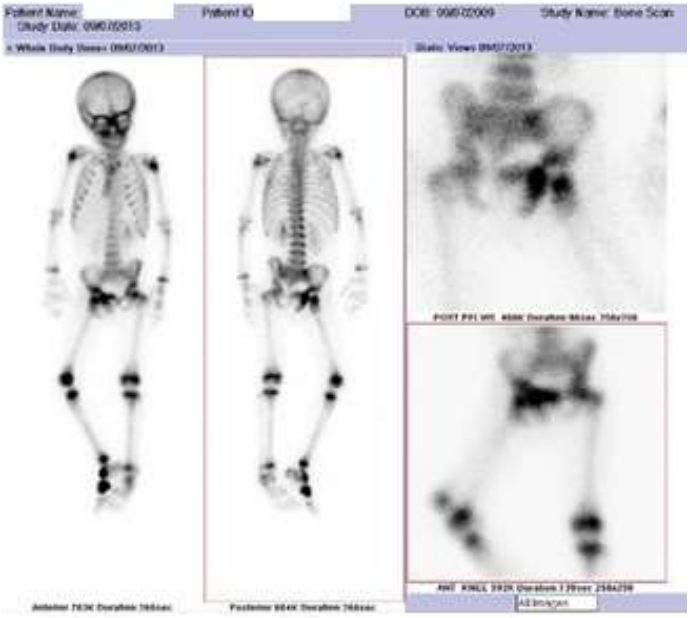

Fig. 1: Bone scan of child

DISCUSSION

Children who have hypogammaglobulinaemia may present with a spectrum of arthritic disease, such as immunecomplex or vasculitic polyarthritis, echovirus syndrome, which mimics joint disease, knee hydroarthritis, and polyarthritis, some of which may be confused with juvenile chronic arthritis, leading to delay in diagnosis and inappropriate treatment. A distinction among these disorders is important in view of their different management gram negative bacilli are an unusual cause of septic arthritis Pseudomonas aeruginosa has been associated with variety of infections as was found in this case. In general population less than 1 % of cases are due to this organism. Most cases of gram negative septic arthritis follow an episode of bactremia and a source, such as

urinary tract,can often be identified [5]. Webster *et al*. have reported that Respiratory infections with Psedomonas infection may be associated with arthalgia and arthritis. Late onset primary hypogammaglobulinaemia is associated with an increased incidence of chronic inflammatory polyarthritis which usually remits on gammaglobulin replacement [6].

Majority of children with hypogammaglobulinemia present with recurrent and severe lower respiratory tract infections and the diagnosis in such cases is usually clinically obvious. In some children, however chest infection may not be very prominent at presentation and instead joint involvement may be the major clinical finding. Such patients have been misdiagnosed as having juvenile rheumatoid arthritis [7]. Recurrent sepsis, sinusitis, otitis media and pneumonia are the usual presenting complaints in patients with hypogammaglobulrnemia [8]. Some of the children, however, can present with arthritis as the major clinical finding Though immunodeficiency as a cause of arthritis is well described m standard text books [9]. The diagnosis is often missed by treating physicians as it clinically resembles oligoarticular JRA, but may progress to polyarticular disease [7].

Juvenile chronic arthritis of systemic onset (Still's disease) presents classically in children under the age of 5 as a trio of fever, characteristic rash, and arthritis. As Bruton's disease occurs in boys of a similar age there may be a diagnostic difficulty in the early stages, but measurement of serum immunoglobulin concentrations and antibody activity readily distinguishes hypogammaglobulinaemia from these conditions. They are also susceptible to joint infections with mycoplasma and ureaplasma that require specialised culture facilities to isolate them from joint aspirates. Although non-steroidal anti-inflammatory agents are a useful adjunct in treating the active stages of arthritis in patients who have hypogammaglobulinaemia, a long term solution is likely to be achieved only after the correct diagnosis has been made and treatment with replacement immunoglobulin started.

Like all children with hypogammaglobulinemia, those who present with arthritis also require prolonged IVIG therapy however this patient was not affording hence we could not give him IVIG. Appropriate antimicrobials should be given in the initial phase depending on the organism isolated Majority of such children show significant improvement with these measures.

CONCLUSION

It is important to recognize hypo-gammagobulrnemia as a cause of arthritis in children; in this case the diagnosis of Bruton's disease was delayed. This delay resulted in considerable joint destruction. It is necessary to make correct diagnosis and to prevent many unnecessary invasive investigations and toxic medications to the child and avoids unnecessary mental and economic burden to the families. We thank the clinicians, paediatricians, and haematologists who cared for these patients and provided many details on which this report is based.

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