

Letters to the Editor

Hyper IgM Syndrome with Tuberculous Osteomyelitis and Scrofuloderma

Hyper IgM syndrome (HIGM) is a rare primary immunodeficiency characterized by low levels of serum IgA and IgG and a normal or increased levels of IgM due to a block in the B cell immunoglobulin switch from IgM to IgG(1). The estimated incidence is approximately 1 in 10,30,000 live births(2). Two types are described - X linked hyper IgM (XHIM) and autosomal recessive hyper IgM (ARHIM). XHIM is more common and affects males. Respiratory tract infections, pneumocystis carinii pneumonia (PCP), chronic diarrhea, cryptosporidium infection, sclerosing cholangitis and chronic neutropenia are the common clinical manifestations. ARHIM is much rarer and affects both sexes and generally has a more benign course.

The main treatment for HIGM is immunoglobulin replacement therapy with IVIG. Cotrimoxazole prophylaxis is required for PCP. Bone marrow transplantation is curative and is considered in patients with XHIM due to fatal infections(3). Patients with ARHIM can lead normal healthy lives provided they receive appropriate treatment for infections.

We report a 2-year-old girl with ARHIM who presented with tuberculous osteomyelitis and scrofuloderma. She had multiple discharging sinuses (in right forearm, lateral aspect of right orbit and right cervical lymphnode), intermittent fever since 7 months and multiple cold abscesses in cervical, submandibular and scalp regions since 15 days. Seven months back, she had right ulnar

osteomyelitis, which was now a chronically discharging sinus. She had multiple matted cervical and axillary lymphnodes. Her anthropometry was in the normal range. Systemic examination was normal except for a soft hepatomegaly. There was no history of tubercular contact. Her X-ray of right forearm revealed lytic lesion in the proximal ulna with periosteal reaction suggestive (*Fig. 1*). A bone scan showed increased tracer concentration in the right orbital bone and right proximal forearm. The abscesses were drained and histopathology showed caseating granulomas with epitheloid cells and acid fast bacilli suggestive of tuberculosis. Her mantoux test was positive (18 × 18 mm). Her X-ray chest was normal and hemogram showed neutrophilia and ESR of 70 mm. In view of diffuse cutaneous and skeletal tuberculosis, an underlying immuno-deficiency was considered. Her HIV ELISA test was negative. Nitroblue tetrazolium test showed 100% positive neutro-



Fig. 1. X-ray of ulna showing proximal lytic lesion.

phil function. Her serum immunoglobulins by nephelometry revealed low IgA (20 mg/dL), low IgG (<200 mg/dL) with elevated IgM (>240 mg/dL) suggestive of ARHIM. She was treated with 4 drug antituberculous therapy. She was started on trimethoprim-sulfamethoxazole prophylaxis and advised regarding immunoglobulin replacement therapy.

Osteomyelitis is a rare manifestation of HIGM and though infections with intracellular pathogens like cytomegalovirus, cryptococcus, candida, histoplasma, toxo-plasma, bartonella have been reported, tuberculosis is a relatively rare clinical manifestation of HIGM(2). Tuberculosis is common in Primary T cell immunodeficiency and IL-12 receptor defects(4). Cutaneous tuberculosis is seen predominantly in immunocompromised patients as was seen in our patient and when present one must suspect an underlying immunodeficiency(5).

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REFERENCES

1. Bejaoui M, Mellouli F, Chouanine K, Dellagi K, Barbouche MR. The Hyper IgM syndrome: 13 observations. *Presse Med* 2003; 32: 544-549.
2. Winkelstein JA, Marino MC, Ochs H, Fuleihan R, Scholl PR, Geha R, *et al.* The X-linked hyper IgM syndrome: Clinical and immuno-logic features of 79 patients. *Medicine* 2003; 82: 373-384.
3. Buckley RH. Primary Defects of Antibody Production. *In: Behrman RE, Kliegman RM, Jenson HB editors. Nelson's Textbook of Pediatrics.* 17th ed. Philadelphia, PA: WB Saunders 2004: 691-693.
4. Fieschi C, Dupuis S, Catherinot E, Feinberg J, Bustamante J, Breiman A, *et al.* Low penetrance, broad resistance, and favorable outcome of Interleukin 12 receptor β_1 deficiency: Medical and immunological implications. *J Exp Med* 2003; 197: 527- 535.
5. Hamada M, Urabe K, Moroi Y, Koga T, Takeishi M, Fujita M, *et al.* A case of multifocal lupus vulgaris that preceded pulmonary tuberculosis in an immune compromised patient. *J Dermatol* 2004;31:124- 128.

Pediatric HIV Infection/AIDS in Agra

Pediatric HIV infection is rapidly emerging as a problem among children in India. As per HIV/AIDS sentinel surveillance data, Uttar Pradesh comes in group-III(1), showing low prevalence of HIV infection. Several factors make Agra a specially vulnerable area from the point of view of HIV transmission. Agra, being major tourist destination, is well

connected by major highways and rail-routes to major cities. This also increases the likelihood of transmission to a significant degree.

This prospective study was conducted among hospitalized children between November 2001 and March 2003 at Department of Pediatrics, S.N. Medical College, Agra. For this study 120 children from 18 months to 12 years of age, suspected of having HIV/AIDS as per WHO criteria for