Pneumocystis carinii, cytomegalovirus, and severe transient immunodeficiency

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Infection with Pneumocystis carinii is rare in infants, and raises strong concerns of immune deficiency. The report describes the unusual case of a male infant with concurrent chest infections caused by P carinii and cytomegalovirus. Investigation was complicated by the strong suspicion of non-accidental injury, including subdural haematomas. The case illustrates how to investigate for possible immunodeficiency. Low immune function tests at presentation slowly improved and have remained normal on longterm follow up. Possible explanations for the transient severe clinical immunodeficiency in this case are discussed.
other forms of T cell deficiency. T cell function tests consistently gave acceptable results, as judged by phytohaemagglutinin responses in vitro. Stimulated T cells expressed CD40 ligand normally (important in the context of “hyper-IgM” (HIGM) syndrome). Neutrophil numbers were normal. Adenosine deaminase and nucleotide phosphorylase concentrations, human leucocyte antigen (HLA) class I and II expression (on peripheral blood lymphocytes by flow cytometry), bone marrow aspirate, chromosome karyotype, DNA testing for ataxia telangiectasia, and response to tetanus immunisation were all normal/negative.

The initial lymphocytopenia and borderline low IgG resolved rapidly, and have remained normal on three year follow up (fig 1). Immunophenotyping of blood lymphocytes improved within months, to show normal numbers and percentages of T (CD3 positive) cells, B (CD19 positive) cells, and natural killer (CD56 positive) cells. IgA and IgM have been normal since 2 years of age. Thus, possible diagnoses such as HIV, SCID, and HIGM syndrome were all excluded.

The examination of maternal serum between early pregnancy and this acute illness showed CMV seroconversion. Therefore, the patient probably acquired the infection in the late antenatal or early postnatal period.

On follow up over a period of three years, the patient shows severe developmental delay, but has been strikingly free of infection on cotrimoxazole prophylaxis. He has received all routine immunisations, omitting live vaccines. Prophylactic septrin was stopped without adverse consequence at this stage.

DISCUSSION

This patient had three major problems: P carinii infection, CMV infection, and evidence of non-accidental injury.

Pneumocystis carinii is a common infectious agent. Seroconversion can be demonstrated in most infants and small children, but clinical disease occurs almost exclusively in the setting of immune compromise. Therefore, this infection should always trigger a search for possible HIV infection or other immunodeficiency. In this case, the search was surprisingly unrewarding. Had we missed something?

“Clinical disease occurs almost exclusively in the setting of immune compromise”

Cytomegalovirus is a relatively common congenital infection with a reported incidence of between 0.2% and 2.2% of live births. Transmission may occur during birth through cervical secretions, or later via breast milk. The virus has been shown to suppress immunity through a variety of mechanisms.

We postulate that CMV transiently impaired this child’s immune function and predisposed him to pneumocystis infection. Indeed, a similar case of congenital CMV complicated by pneumocystis has been described.

Neither fractures, subdural haematomas, nor non-accidental injury have previously been reported in association with CMV or pneumocystis.

Take home messages

- Pneumocystis carinii infection is rare in infants, and raises strong concerns of immune deficiency
- We describe the unusual case of a male infant with concurrent chest infections caused by P carinii and cytomegalovirus (CMV) who did not appear to be immunocompromised
- We postulate that CMV transiently impaired this child’s immune function and predisposed him to pneumocystis infection

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References