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SCIENTIFIC LETTER

Perseverance in Persistent Pneumonia

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To the Editor: Persistent pneumonia is a clinical condition in which symptoms of pneumonia persist longer than usual or recur again unless the underlying cause is identified and treated [1]. We encountered an interesting case of persistent pneumonia, a 4-mo boy first born to non-consanguineous marriage, exclusively breast fed, immunized as per age who suffered viral prodrome which progressed to pneumonia which persisted longer than normal. History such as nasal regurgitation of feeds, excessive precordial activity, skin rashes, greasy stools and salty to kiss were absent except the kid failed to thrive. Investigations such as milk scan, ECHO, stool for fat globules were negative. X-ray and CECT chest showed bilateral diffuse ground glass opacities ruling out congenital lung malformation. Bronchioalveolar lavage for tuberculosis, fungal, *Pneumocystis jiroveci* and other atypical organisms were negative. HIV status of the mother was negative, immunoglobulin profile and TBNK levels were also normal. Finally, clinical exome sequencing was performed which was positive for X-linked Hyper IgM type 1 (HIGM). The child showed improvement after IvIg infusion and continued on monthly IvIg therapy and Co-trimoxazole prophylaxis without any recurrence of symptoms with adequate weight gain and normal development. The uniqueness of this case is that we have normal Ig profile in the face of hyper IgM syndrome. Why did this child respond to IvIg when the Ig profile is normal? Still, the function of the inherent immunoglobulins leaves us baffled as they might be dysfunctional in primary immunodeficiency syndromes. This case also emphasizes the importance of active search for effective management. X-linked HIGM is a class switching defect which affects the CD40 ligand on the T cells leading to failure of B cells to switch from one immunoglobulin production to another [2]. The definitive management is hematopoietic stem cell transplant, and supportive therapy such as IvIg and antibiotic prophylaxis will help in survival.

Declarations

Conflict of Interest None.

References

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