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Research Paper

Recurrent oropharyngeal candidiasis with T-cell deficiency: a case report

Medical Science

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Anirban Chatterjee | Department of Pediatrics, Institute of Post Graduate medical education and research

ABSTRACT

We report a case of six-year-old boy, presenting with recurrent oropharyngeal candidiasis without Candidal sepsis. The most important immunological characteristics were T cell deficiency, anergy to common recall antigens but normal serum immunoglobulins, and phagocytosis. The patient responded to antifungal treatment. Considering clinical course of the disease and laboratory finding, patient was diagnosed as a case of chronic mucocutaneous candidiasis.

Introduction –

Chronic mucocutaneous candidiasis(CMC) is a rare and heterogeneous group of disease. CMC is manifested by superficial recurrent or persistent candida infection of mucous membranes and/or the skin, which is caused by a selective immune disorder of T-cells to Candida antigens However, CMC, is not associated with disseminated invasive Candidal sepsis[1], so mortality is uncommon. In addition, CMC is considered as a phenotypic expression of a range of immunologic, endocrinologic, and autoimmune problems.

Case report -

A 6yrs old male child was referred to our hospital for recurrent attack oral thrush. On admission, patient had fever (low grade, intermittent), non-productive cough and tachyponea. Parents and boy had no history of immunologic or endocrine diseases. The boy had no significant medical illness in past. Oral thrush first developed at two and half years of age. His growth and development history was within normal limit. On clinical examination, white adherent plaques of thrush were found over tongue .The rest of the cutaneous examination was normal. There was no localised or generalised lymphadenopathy. Nothing was significant in General survey and Systemic examination except fine crepitations in bilateral basal lung area.

Hemogram showed hemoglobin 10 gm%, total WBC count 14000, lymphocyte 57%, neutrophils 35% and eosinophils 8%. The chest X-ray and USG abdomen showed no abnormality.

KOH preparation of oral scrapping revealed fungus pseudohyphae, and the culture results showed Candida albicans growth. No growth was found in blood culture. To exclude CMC-associated endocrine dysfunction, random blood glucose, glycosylated hemoglobin test, thyroid function tests, liver functions tests, serum electrolyte evaluation - serum phosphorus and calcium were done and all are within normal limit. Screening for an HIV test was nonreactive.

IgA, IgM and IgG were within normal limits, but IgE was higher value of >2500 iu/ml. The phagocytic function assessed by the nitroblue tetrazolium dye reduction test was found normal. Skin tests has done using the killed *Candida albicans* cells and purified protein derivative (PPD) 10 IU were negative.

The phenotypes of lymphocytes showed B lymphocytes (CD19) 36.4% (N–14-23),T lymphocytes (CD3) 45% (62-80) and CD4 cells- 26.2%(N 35-51%),CD4/CD8 ratio was 0.39(N-0.84-3.05).

On the basis of clinical and laboratory investigations the boy was diagnosed as a case of Chronic Mucocuteneous Syndrome and was treated by topical clotrimazole and oral ketoconazole (3.3 mg/kg/day for 6 weeks). Respiratory symptom has improved with 14 days Trimethoprim-sulfamethoxazole (TMP-SMX) treatment**.** The patient became symptom free within 10 days of treatment.

At present, patient is advised that gurgling (2% Povidone Iodine mouth gurgle) whenever a sign of oral thrush is present to prevent severe recurrence.

Discussion-

CMC is a selective T cell-mediated immunity defect to Candida antigens but underlying basic pathobilogy is unknown.CMC is rare disorder. The reported cases from Asians are rare [2] A few cases CMC reported from India[4 5 6]. Of those, cases [4 5] has been diagnosed by mainly clinical parameters. Karnik AC et al[6] has mentioned T cell abnormalities in their reported case.

Even though CMC clinically presents at any age, typically appear before 3 years of age.[9]It may affect as single or multiple lesions and include the mouth, esophagus, nails, or skin. At the start, CMC are identical to familiar manifestations of mucocutaneous candidiasis[9], such as oropharyngeal candidiasis in the index case, over time CMC shows its chronicity. There are subtypes of CMC, as follows: oral, localized, diffuse, with endocrinopathy, with thymoma, interstitial keratitis, with the triad of keratitis, ichthyosis and deafness. The index case has been suffering from oral CMC and most distinguishing feature of this subtype is that it does not present with hyperkeratosis which is characteristics of other form of CMC[1].

We have investigated following Kirkpatrick guidelines which includes CBC with differential, Lymphocyte phenotyping, Endocrine function tests (Calcium, phosphate, TSH,), B lymphocyte counts, Serum IgG, IgA, IgM and IgE , HIV test. However we were not able to perform the test - Lymphokine production by antigen or mitogen-stimulated T cells- because of lack of laboratory facilities.

The findings of immunological investigations of the index case were slightly increased lymphocyte count, an increased proportion of B cells (CD19), a decreased proportion of T cells (CD3), a decreased CD4 cells count and altered ratio of CD4/ CD8 ratio. Delayed-type hypersensitivity skin test to tuberculin and candida were negative. In addition, Lisa J. Kobrynski et al[7] had demonstrated that CMC has a significant decrease in the percentage of CD4-positive lymphocytes. These facts point towards a defect in T cell-mediated immunity and anergy to Candida antigens. These findings are correlates with CMC in earlier studies. [2] Although underlying defects remains unknown, studies on animal models have indicated the role of type 1 cytokines, the T-helper 1 (Th1) and Th2 cells and important role of dendritic cells are present. [2]

Most of CMC patients associated with pneumonia. [2] It has seen that the respiratory symptoms of the index case probably because of Pneumocystis (carinii) jiroveci Pneumonia, which had resolved with TMP-SMX treatment. CMC with *Pneumocystis* jiroveci pneumonia has reported in several cases in the literature [3]

The azole antifungal agent is mainstay of treatment either con-

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tinuously or intermittently and CMC patient improve dramatically. Ketoconazole was the first and very successful agent [9]for CMC. However, like most patients have a relapse within a few weeks or months whenever treatment has stopped because the underlying defect could not be corrected[2]. The index patient has treated with ketoconazole but reappeared within few months after therapy-discontinuation. Next episodes were treated by fluconazole and put on 2% Povidone Iodine mouth gurgle whenever sign will appear that therapy advocated based on Kariuki E et al [8]study – a cost effective method to prevent severity and resistant candidiasis. . An alternative therapy for CMC, who fails long-term antifungal therapy, is cimetidine and zinc sulphate[2]

Message -

It is hard to make a well-timed diagnosis of CMC. Once the diagnosis has made, detailed studies should be needed to exclude endocrinopathy, thymoma, or autoimmune diseases. And follow-up is necessary because of associated disorders may come about after a long years.

REFERENCE

1. Kirkpatrick CH. Chronic mucocutaneous candidiasis. Pediatr Infect Dis J 2001;20:197-206 | 2. Shih-Jung Chiu Ching-Hsiung Tsao, Li-Chen Chen, Chun-Chieh Kao, Ko-Huang Lue, Jing-Long Huang Chronic mucocutaneous candidiasis in a 6-year-old boy J Microbiol Immunol Infect 2004;37:196-199 | 3. David T Robles http://emedicine.medscape.com/article/1091928-overview#showall | 4. Sathish K, Scott JX, George R. Chronic mucocutaneous candidiasis in a child. Indian J Dermatol Venereol Leprol 2005;71:432-3 | 5. Hassan Iffat, Mashkoor Ahmed, Hayat Bhat Mohammad et al, Chronic mucocutaneous candidiasis - A CASE REPORT , N Dermatol Online. 2010; 1(2): 19-21 | 6. Karnik AC, Vijayvargiya R. T-cell deficiency with recurrent mucocutaneous candidiasis. Indian J Dermatol Venereol Leprol 1990;56:43-4 | 7. LISA J. KOBRYNSKI, LISA TANIMUNE,LAURIE KILPATRICK,et al Production of T-Helper Cell Subsets and Cytokines by Lymphocytes from Patients with Chronic Mucocutaneous Candidiasis CLINICAL AND DIAGNOSTIC LABORATORY IMMUNOLOGY,1996;3:740–745 | 8. Kariuki E, Ngugi R, Muthotho J; International Conference on AIDS. Povidone iodine therapy for recurrent oral Candidiasis to prevent emerging Antifungal resistant Candida Strains. Int Conf AIDS. 2000;13:9-14 | 9. Kirkpatrick, C. H. 1989. Chronic mucucutaneous candidiasis. Eur. J. Clin. Microbiol. Infect. Dis. 8:448-456. |