Case Report

A rare case report - case of idiopathic CD4+ lymphocytopenia with palatal perforation

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Abstract

Introduction: Idiopathic CD4+ lymphocytopenia (ICL) is very rare medical syndrome in which CD4+ T lymphocyte count is decreased. It is sometimes characterized as HIV negative AIDS. People with ICL have weakened immune system and susceptible to opportunistic infections. We are reporting this case because we have found palatal fistula in patient with ICL. This case report illustrates the difficulty in diagnosing a rare condition with non-specific clinical manifestations.
Keywords: Idiopathic CD4+ lymphocytopenia (ICL), Palatal perforation.

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INTRODUCTION

Idiopathic CD4+ lymphocytopenia (ICL) is a presumed heterogenous syndrome with key element low CD4+ T-cell counts (below 300/mm^3) without evidence of HIV infection or other known immunodeficiency. The etiology, pathogenesis, and management of ICL remain poorly understood and inadequately defined. The clinical presentation can range from serious opportunistic infections to incidentally diagnosed asymptomatic individuals. Cryptococcal and tuberculous and non tuberculous mycobacterial infections and progressive multifocal leukoencephalopathy are the most significant presenting infections, although the spectrum of opportunistic diseases can be similar to that in patients with lymphopenia and HIV infection. Idiopathic CD4 + lymphocytopenia (ICL) was described in 1992 as an immunodeficiency syndrome characterized by opportunistic infections and low CD4 + T-cell counts in the absence of HIV infection. Despite the 22 years that have elapsed, the clinical spectrum, pathogenesis, and possible treatment for ICL remain obscure. Palatal perforation may be developmental, autoimmune, neoplastic, drug related, iatrogenic or infectious. Immunocompromised patients are succeptible to opportunistic infections leading to palatal perforation. In leprosy, tertiary syphilis, tuberculosis, rhinoscleroderma, naso-oral blastomycosis, leishmaniasis, actinomycosis, histoplasmosis, coccidiomycosis and diphtheria the palatal roof may be perforated.

CASE REPORT

55 years male patient was presented to outpatient department of otorhinolaryngology of Gokuldas Tejpal hospital mumbai with chief complaint of regurgitation of food from left nostril since one and half month. detailed history of patient was taken. There was no history of facial trauma, any oral instrumentation, tuberculosis, diabetis mellitus or any immunosuppressive drug intake or drug addiction. Over the previous period, the patient was prescribed antibiotics for sore throat for duration of three weeks six month back. Patient's built was average. On examination of the lesion, it was found that there was 4 x 2 cm non tender ulcer with fibrotic margins

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communicating with left nasal cavity. slough was present over the edges of the fistula. Adjacent of oral mucosa was normal. Patient was investigated thouroully. HRCT of paranasal sinuses was done which was showing left hard palate defect with pansinusitis. Laboratory analysis of patient was done. Hemoglobin level, blood sugar level ,serum urea and creatinine level, serum calcium level were within normal limits .Urine routine microscopy and urine protein were negative .White blood cell count was 6900/mm with decrease of the percent of lymphocyte.CD4+ T lymphocyte count was 260 cells/cumm. Platelet count was 2,45000 cells/mm. blood coagulation profile and ESR were within normal limits.

- Chest X ray, ECG, USG abdomen and pelvis were within normal limits.
- VDRL test, test for hepatitis B antigen, HIV test, tests for acid fast bacilli were negative.
- RA factor, c ANCA, p ANCA test were also done to rule out collagen vascular disease.
- Peripheral blood smear tests for Malaria and Lesmanias is were negative.
- Biopsy from edge of fistula was suggestive of infected granulation tissue with negative Gomori methenamine silver (GMS) and Periodic acid Schiff (PAS) stains for fungus along with no evidence of malignancy.

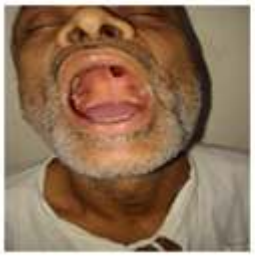

Figure 1: Case of ICL with palatal perforation

Figure 2: CT scan of paranasal sinuses suggestive of pansinusitis with hard palatal defect

DISCUSSION

The immunodeficiency syndrome of Idiopathic CD4+ T – lymphocytopenia was defined for Adults by the Centre for disease control and prevention (CDC) as

1. Depressed numbers of circulating CD4+T cell lymphocytes (<300cell/cumm or < 20% of total T cells) on more than one occasion.
2. No laboratory evidence of infection with Human Immunodefiency virus Type I (HIV I) or Type II (HIV -2).
3. Absence of any defined immunodefiencies or therapy associated with depressed levels of CD4

The differential diagnosis of ICL is large and this diagnosis remains one of exclusion and requires an extensive immunologic, hematologic, rheumatologic, and infectious disease workup as well as follow-up testing to confirm the persistence of lymphocytopenia. As stated in the syndrome definition, other forms of immunodeficiency have to be excluded and the provisional ICL diagnosis has to be laboratory-confirmed at least twice during a period of 1 to 3 months. A comprehensive immunologic work-up should be applied in order to exclude HIV infection, lymphoma, autoimmune diseases, other forms of immunodeficiency (such as common variable immunodeficiency), and sarcoidosis. As mentioned above with tuberculosis, special attention should also be paid to the possibility that the presenting infection or treatment may be the cause of lymphocytopenia. The diagnosis of ICL is typically suspected when an opportunistic infection is identified in an otherwise healthy individual. The literature is dominated by such cases and is constantly expanding. The majority of cases report opportunistic infections - most notably cryptococcal or mycobacterial disease and progressive multifocal leukoencephalopathy (PML) - that are normally seen in HIV-infected patients Opportunistic infections at or before the time of presentation were the most common clinical illness. 40% of the patients had AIDS defining illnesses and 53% had other illnesses that were non AIDS defining and 6% were asymptomatic. In immunocompromised patients mucormycoses, aspergillous, tuberculosis, histoplasmosis, blastomycosis are some common apportunistic infections causing palatal perforation. Our present case satisfied the case definition criteria of Idiopathic CD4+ T–lymphocytopenia by having no evidence of HIV infections on repeated testing (spot and ELISA) and having a CD4 count of 260cell/cumm. He neither had high-risk behaviour or blood transfusions in the past hence reducing the possibility of HIV infection. He was also screened for other causes of immunosuppression like leukemia, diabetes, and immunosuppressive therapy.

CONCLUSION

At this point we can reasonably conclude that idiopathic CD 4 + T cell Lymphocytopenia is a rare syndrome, it is

not new as cases have been reported as early as 1983, it is not caused by HIV I, HIV -2, HTLV 1,HTLV 2, is epidemiologically, clinically and immunologically different from HIV and does not appear to be caused by an infectious agent. It is a possible cause of opportunistic infections in the so-called immunocompetent host unless they are screened with CD4 Counts after an HIV negative report. It is yet unclear whether in some patient's idiopathic CD4+T cell lymphocytopenia develops as a consequence to an infection or the infection is the cause of low CD4 +T cell count. Further studies of these cases could give us an insight into the complexities of the immune system and its role in host defence mechanism.

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