ABSTRACT

Post-Kala Azar Dermal Leishmaniasis (PKDL) as sequelae of Visceral Leishmaniasis is well described in literature. However in the Indian Subcontinent, these sequelae occur 6 months – 3 years after Visceral Leishmaniasis, in contrast to East African pattern, where it often occurs during the course of Visceral Leishmaniasis. This could possibly be an eye opener that there need not be significant differences in the time frame of presentation of Indian and East African PKDL.

Keywords: Post-Kala Azar Dermal Leishmaniasis; Visceral Leishmaniasis; East African PKDL

CASE REPORT

History

50 year old gentleman from Nilambur, Kerala, India presented with fever of 1 month duration. Fever was initially low grade for two weeks, but later he developed high grade fever with chills and rigors. He had significant loss of weight over past 1 month. There was no history of respiratory, abdominal or urinary symptoms. There was no history of recent travel outside or contact with tuberculosis. He had taken some self medications for his illness which included oral antibiotics. But in view of persistent fever, he was referred to our centre by a local practitioner. He hailed from a tribal community and was of poor socioeconomic status and was a chronic smoker and alcoholic.
EXAMINATION

On examination, he was conscious, oriented, moderately built and poorly nourished with a Body Mass Index of 17.2. There was diffuse hyperpigmentation of skin (Figure-1).

Figure-1: Hyperpigmentation of skin

His vital signs were stable and he had pallor and pedal edema. There was no icterus, cyanosis or lymphadenopathy. Examination of abdomen showed palpable liver 6cm below right costal margin in mid clavicular line and massive splenomegaly reaching up to umbilicus. Examination of other systems was within normal limits.

INVESTIGATIONS

Investigations showed Hemoglobin 7.8 g%, Total Count 3600 cells/mm³, Platelet count 90,000/mm³, MCV 74.3fl, MCH 23.6pg, MCHC 31.7g/dl, RDW-21% and random blood glucose was 123 mg/dl. Urine microscopy, liver function tests and renal function tests were within normal limits. ECG and Chest X-ray were normal and HIV screening was negative.

Peripheral smear showed Microcytic hypochromic with elliptocytosis, tear drop cells, target cells, few micro-ovalocytes and anisopoikilocytosis was seen. Peripheral smear was negative for malarial parasite. USG Abdomen showed hepatosplenomegaly with normal liver echoes. Mantoux test was negative and reticulocyte count was 2%. Serum ferritin and Serum LDH was within normal limits. Blood culture and Urine Culture was sterile and sputum AFB was negative. Direct Coombs Test and Indirect Coomb’s test were negative. Echocardiography did not show any vegetation.

CLINICAL POSSIBILITIES

At this point of time, with massive splenomegaly, hepatomegaly, pancytopenia and hyperpigmentation, possibilities entertained were that of myeloproliferative disorders, other haematological malignancies, lymphoma and disseminated tuberculosis. Considering the epidemiological background of Nilambur, visceral leishmaniasis was also considered. Chronic malaria was also kept in mind, though lower down in the list.

ROAD TO DIAGNOSIS

We proceeded with Bone marrow aspirate, which showed Erythroid-Normoblastic maturation, Myeloid-Maturation and Megakaryocytes seen with no atypical cells/infiltration.

We had started the patient on broad spectrum antibiotics in view of persisting fever. Anemia was corrected by blood transfusion. Meanwhile bone marrow trephine result was obtained which clinched the diagnosis. Trephine biopsy showed Erythroid-normoblastic maturation, Myeloid-maturation normal and Megakaryocytes seen. Also seen are histiocytes with LD bodies, suggestive of Leishmaniasis (Figure-2).

Meanwhile during the hospital stay, we noticed the appearance of a nodular swelling in the dorsum of right middle finger of the patient (Figure-3), which
was not present at hospital admission.

FNAC was taken from this lesion and the same yielded LD bodies (Figure-4).

A final diagnosis of Visceral Leishmaniasis with Post Kala Azar Dermal Leishmaniasis was made. However, something unusual about our case was that in Indian subcontinent, interval between Vischeral Leishmaniasis and Post Kala Azar Dermal Leishmaniasis (PKDL) is usually around 6 months to 3 years. In our case, the nodule on the dorsum of middle finger appeared during the course of illness. Such a pattern is usually seen in the East African region. The patient was started on Amphotericin B infusion given alternate days for 1 month. Fever subsided and patient improved clinically. Within 2 weeks of starting treatment, the spleen was not palpable. He was discharged after a total of 1.5 months of hospital stay and is now under follow up and is doing well.

DISCUSSION

This patient presented with massive splenomegaly, hepatomegaly, hyperpigmentation and pancytopenia. Pancytopenia in presence of massive splenomegaly may very well be due to hypersplenism, provided bone marrow is normal. But in our case, bone marrow revealed LD bodies diagnostic of Visceral Leishmaniasis. Pattern of Post Kala Azar Dermal Leishmaniasis between Indian Subcontinent and East Africa is different and is well described in literature. PKDL incidence is less among Indian subcontinent than among East African population. More striking is the observation that the interval between visceral leishmaniasis and PKDL is around 6 months to 3 years in the Indian subcontinent, whereas in East Africa, PKDL can occur either during the course of illness of VL or within 6 month of illness. However, in our case PKDL occurred during the clinical course of VL, which is unusual in the Indian scenario. This could possibly be an eye opener that there need not be significant differences in the time frame of presentation of Indian and East African PKDL.
LEARNING POINTS

1) What is Kala-azar?
1. Slowly progressing indigenous disease caused by a protozoan parasite of genus *Leishmania*
2. In India *Leishmania donovani* is the only parasite causing this disease.
3. The parasite primarily infects reticuloendothelial system and may be found in abundance in bone marrow, spleen and liver.
4. Post Kala-azar Dermal Leishmaniasis (PKDL) is a condition when *Leishmania donovani* invades skin cells, resides and develops there and manifests as dermal lesions.

2) What are the signs and symptoms of Kala-azar?
1. Recurrent fever intermittent or remittent with often double rise
2. Loss of appetite, pallor and weight loss with progressive emaciation
3. Splenomegaly – spleen enlarges rapidly to massive enlargement, usually soft and nontender
4. Hepatomegaly – enlargement not to the extent of spleen, soft, smooth surface, sharp edge
5. Lymphadenopathy – not very common in India
6. Skin – dry, thin and scaly and hair may be lost. Light coloured persons show grayish discoloration of the skin of hands, feet, abdomen and face which gives the Indian name Kala-azar meaning “Black fever”
7. Anaemia – develops rapidly

3) What are Signs & Symptoms of PKDL?
Types of morphological lesions:
1. Early hypopigmented macules similar to macular lesions of Lepromatous Leprosy but normally less than 1 cm. Usually occur on face but can affect any part of the body.
2. Later (after a variable period of months or years) diffuse nodular lesions on those macules
3. Erythematous butterfly rash which may be aggravated by exposure to Sunlight; an early sign of PKDL
4. Erythematous papules and nodules which usually occur on face, especially the chin.
5. Lesions progressive over many years, seldom heal spontaneously

REFERENCES