Hairy Cell Leukemia Masquerading as Infective Endocarditis

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Abstract Hairy cell leukemia is a chronic lymphoproliferative disorder affecting middle-aged adults, with the median age of 50–55 years. We report a case of hairy cell leukemia who presented with fever, splinter haemorrhages and moderate splenomegaly, simulating infective endocarditis. There was bicytopenia at presentation and hairy cells were seen in the peripheral blood.

Keywords Hairy cell leukemia · Infective endocarditis · Pyrexia unknown origin

Introduction

Hairy cell leukemia presents with varied manifestations like weakness and fatigue due to anemia, bleeding, early satiety due to massive splenomegaly, bacterial infection

secondary to neutropenia and with infiltrative skin lesions. Our patient also had unusual features like large skin blisters at presentation. Hairy cell leukemia masquerading as infective endocarditis has not been reported.

Case Report

A 50 year old male was admitted with high grade continuous fever for 1 month duration. He also had skin lesions which started as large blisters in limbs, chest and palms, which subsided spontaneously over 20 days. Patient also noticed yellowish discolouration of nails since the onset of fever. There was no cough, jaundice, bleeding manifestations or bone pains. On examination, he had multiple post inflammatory pigmentation with scaling in the limbs, chest and palms. There was nail pitting, onycholysis, discolouration of the distal half of the nails and multiple splinter haemorrhages. He had moderate pallor but no petechiae, lymphadenopathy, bone pain or jaundice. CVS, RS examination was normal. Abdomen examination revealed moderate splenomegaly, 7 cm below the left costal margin. As the patient had fever, splinter haemorrhages, anemia and splenomegaly, the diagnosis of subacute bacterial endocarditis was considered but blood cultures were sterile and transthoracic echocardiogram was normal (Figs. 1, 2, 3).

His hemogram showed bicytopenia with haemoglobin of 7.0 g/dl, total leukocyte count 3300/μl, polymorphs 21%, lymphocytes 79% and platelet count 2,67,000/μl. Peripheral smear showed hair like projections in some of the lymphocytes. Bone marrow biopsy showed hypercellular bone marrow with diffuse infiltration by lymphoid cells, increased marrow fibrosis, and fried egg appearance characteristic of hairy cell leukemia. The lymphoid cells were

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Fig. 1 (a) Skin lesions in the left forearm. (b) Palm. (c) Legs

positive for CD19, CD25, CD11c, CD45 and CD 103 confirming the diagnosis of hairy cell leukemia .

Discussion

Hairy cell leukemia (HCL) is a low-grade B-cell lymphoproliferative disorder with distinctive morphologic, cytochemical, and immunologic characteristics. The most common presenting symptoms/signs were weakness and fatigue (51% of cases), concurrent infection (17%), left upper quadrant pain (14%), and hemorrhagic phenomena, such as ecchymoses, purpura, and epistaxis (9%) [1, 2].

The most common category of skin lesion in HCL patients is infection (herpetic, cellulitis, abscess, pyoderma, dermatophytosis). In addition to bacterial, viral, and fungal infections, skin lesions may be due to atypical mycobacteria. Leukemia cutis, Sweet's syndrome, leukocytoclastic vasculitis are the other skin manifestations [3–7]. There has been a significant association between HCL and vasculitic syndromes. Vasculitis in HCL can be due to polyarteritis

Fig. 2 Nail changes: (a) pitting, onycholysis, (b) splinter haemorrhage

Fig. 3 (a) Peripheral smear showing hairy cell (b) Bone marrow imprint showing hairy cells $\times 200$

nodosa, leukocytoclastic vasculitis, or due to direct invasion of vessel walls by hairy cells [8, 9].

In our patients skin lesions had healed at the time of presentation and hence biopsy was not done. The presence of associated splinter haemorrhages suggests that the lesions may be due to vasculitis. Hairy cell leukemia presenting with nail changes like nail pitting splinter haemorrhages has not been reported in the literature.

Conclusion

Hairy cell leukemia can rarely mimic infective endocarditis in its clinical presentation. In a patient with pyrexia of unknown origin, splenomegaly and cytopenia the diagnosis of hairy cell leukemia should be considered.

References

- Hoffman MA (2006) Clinical presentations and complications of hairy cell leukemia. *Hematol Oncol Clin N Am* 20:1065–1073
- Bouroncle B (1979) Leukemic reticuloendotheliosis (hairy cell leukemia). *Blood* 53(3):412–433
- Finan MC, Su WP, Chin-Yang L (1984) Cutaneous findings in hairy cell leukemia. *J Am Acad Dermatol* 11:788–797
- Carsuzaa F, Pierre C, Viala JJ (1993) Cutaneous findings in hairy cell leukemia. Review of 84 cases. *Nouv Rev Fr Hematol* 35:541–543
- Lawrence D, Sun NCJ, Mena R et al (1983) Cutaneous lesions in hairy-cell leukemia. *Arch Dermatol* 119:322–325
- Frassoldati A, Lamparelli T, Federico M et al (1994) Hairy cell leukemia: a clinical review based on 725 cases of the Italian Cooperative Group (ICGHCL). *Leuk Lymphoma* 13:307–316
- Golomb HM, Hadad LJ (1984) Infectious complications in 127 patients with hairy cell leukemia. *Am J Hematol* 16:393–401
- Hasler P, Kistler H, Gerber H (1995) Vasculitis in hairy cell leukemia. *Semin Arthritis Rheum* 25(2):134–142
- Bouroncle B (1987) Unusual presentations and complications of hairy cell leukemia. *Leukemia* 4:288–293