

Langerhans Cell Histiocytosis: Rare Disease Treated With Chemotherapy and Radiotherapy

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Abstract: Langerhans Cell Histiocytosis represents a rare benign disorder previously designated as "Histiocytosis X Variety" Type II Histiocytosis or Langerhans cell Granulomatosis, it occurs when the body accumulates too many immature langerhans cell a subset of the larger family of cell known as histocytes. Langerhans cell are of white blood cell that normally help the body to fight against infection. Langerhans cell histiocytosis is most commonly characterized by single or multiple osteolytic bone lesion but also ulcerations of skin & soft tissue and also involvement of the CNS. Bones are the most common localization of disease (40-78%). Surgery is the treatment of choice in limited form of histiocytosis but in diffuse form only chemotherapy is used. The most of researches also recommends the radiotherapy in conditions, like Recurrence or progression of disease bone pain, exophthalmus, non surgical local changes. The aim of this study is to present four such rare patients having multi system diseases with multiple organs system treated with chemotherapy & radiotherapy at our institution.

Keywords: Benign, chemotherapy, eosinophilic granuloma, multifocal lesion.

I. Introduction

Langerhans cell histiocytosis (LCH) is a rare benign histiocytic disorder most commonly characterized by single or multiple osteolytic bone lesions but also ulcerations of skin and soft tissues and also involvement of the CNS are described. It is characterized by an uncontrolled clonal proliferation of langerhans cells which belong to the normal human mononuclear phagocytic system. Langerhans cell histiocytosis (LCH) formerly referred to as histiocytosis X, can be divided into three categories: eosinophilic granuloma, Hand Schuller Christian disease and Abt_Letterer-Siwe disease. LCH involves single (single-system disease) or multiple (multiple-System disease) organ systems. In 60% bony manifestations as single-system disease with uni or multifocal lesions are present but also in multi-system disease with activity in other organ systems are also described. In case of involvement of the CNS diabetes insipidus is a typical symptom. Patients with single system disease tend to do well. Patients with multiple system disease sometimes have an unpredictable outcome. Patients with only one bony lesion progression, disseminated disease and fatal outcome has been described. Nevertheless some self healing and self limiting courses has been reported.

II. Material Methods

We are presenting such rare disease of Langerhans histiocytosis who attended our OPD at Regional cancer Centre Raipur Chhattisgarh. Our study includes four patients with isolated and those with multifocal bone lesion treated with chemotherapy and Radiotherapy. The aim of our study is to find out disease free survival, recurrent disease death and therapy related side effects of chemotherapy & Radiotherapy. According to the LCH classification of Greenberger et al two child were having stage III disease & two adult having stage II disease.

Table 1: Clinical Presentation

S.No.	Age	Presentation
1	<1Y	Multiple bone lesion of long femur bone
2	2Y	Lesion in the oral mucosa, gums, palatal bone with skin involvement
3	21Y	Craniofacial bone involvement with lesion in the orbital soft tissue extension
4	16Y	Eye involvement with proptosis. Lesions in the orbit

Treatment Given

1. Chemotherapy Inj. Vinblastin, Tab. Prednisolone- Weekly for 8weeks.
2. Chemotherapy Inj. Vinblastin, Tab. Prednisolone weekly for 6wks.
3. Chemotherapy + RT- weekly 12wks Inj. Vinblastin + Radiotherapy with linac 2400cg/vinblastine 12#/tocranial bone+Maintaince therapy crossing of pulses of Inj. Vinblastin & Tab. Prednisolone.
4. Chemotherapy 1 cycle of Inj.Vinblastin + Tab. Prednisolone then lost to treatment.

III. Results And Discussion

In accordance with the nature of LCH all three patient were having NAD (Non Active disease) with regressive disease having regression of sign and symptoms with no new lesions. The patients are advised for 12month pulse therapy of maintenance and thereafter advised for the follow up investigation after and therapy. For clinical exam every 6weeks for 1 year then every 3months for one year and 6monthly for next 2years. Similarly height, weight and pubertal status every 6month for 5years. Patients were kept on follow up with Radiographs of some lesion only if new lesion or reactivations are suspected.

IV. Conclusions

Due to rarity of LCH and the variety of symptoms of the disease, oncologist encounters a number of diagnostic and therapeutic difficulties Chemotherapy and Radiotherapy is a safe and effective treatment option for patient suffering from LCH Chemotherapy for the multifocal lesion. Low RT dose to unifocal lesion shows sufficient local control. Side effect are low and no treatment related deaths were observed.

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