

Langerhans Cell Histiocytosis of Maxillary sinus

A B Azreen, MD, C Y Kwan, MBBS, N Prepagaren, FRCS (Edin)

Department of Otorinolaryngology, Faculty of Medicine, University Malaya Medical Centre, 50603, Kuala Lumpur, Malaysia

SUMMARY

Langerhan cell histiocytosis is a rare disease and usually occurs in paediatric age group. This disease may involve single or multiple organs system and has an unpredictable course of disease. The involvement of head and neck region are almost 90 % of cases, however maxillary sinus involvement is very rare. We report a case of 2 year old boy presented with multi organ LCH (orbit, skull, sinus and liver). The mainstay treatment for this high risk multi organ LCH group is chemotherapy. Unfortunately, although with the advancement of treatment, their mortality rate is still high.

KEY WORDS:

Langerhans cell histiocytosis, maxillary sinus

INTRODUCTION

Langerhans Cell Histiocytosis (LCH) is a rare disease. This disease was previously designated as histiocytosis X, eosinophilic granuloma, Letterer-Siwe disease and many more. Several theories have been suggested to explain the pathophysiology. These include uncontrolled immunologic stimulation of a normal Langerhan's cell resulting in the proliferation and accumulation of these cells, histiocytic reaction secondary to infection or excessive cytokines or lymphokines production and neoplastic proliferation of monocyte-macrophage cell's series¹. Currently it is recognized that LCH represents a spectrum of disease ranging from the benign unifocal lesion to a more aggressive multisystem disease.

CASE REPORT

LS, a 2 years old boy, presented with right periorbital swelling associated with facial puffiness for 4 days duration. The swelling was noted by his mother after he woke up from sleep. There was no pain, redness, blurring of vision, eye discharge, history of trauma, nasal symptoms such as nose block or purulent nasal discharge. He also had abdominal distension with dark-coloured urine but no jaundice, diarrhoea, vomiting, hematuria, leg swelling or fever noted. There was no similar illness among other family members.

On examination, patient was comfortable, pink and not jaundiced. His vital signs were stable. There were periorbital swellings involving both eyes, with right eye more prominent than left, but no redness or pus discharge. The range of movement of both eyes were full. His vision was normal. No facial tenderness noted. Otoscopy examination revealed normal findings. He had hepatosplenomegaly with mild ascities. There was no skin rash seen, no lymph node

palpable and no pedal edema. CVS, CNS and lungs examination were normal.

Blood investigation on admission showed abnormal of liver function with signs of sepsis, therefore he was started on IV antibiotic (Cefeprozone and Metronidazole). While in the ward, patient developed gradual right eye proptosis with a firm swelling over the right cheek. An urgent CT scan of the orbit and paranasal was performed which showed presence of enhancing mass in right maxillary sinus eroding and destroying the adjacent anterior, medial, lateral and superior walls. The tumour extended into the right nasal cavity and floor of orbit causing displacement of orbit (Figure 1). There was also presence of tumor deposited in the medial aspect of left orbit, left parietal bone and left occipital bone.

Nasoendoscopic assessment under general anaesthesia showed friable fleshy mass arising from the right maxillary sinus extending into the orbit and right middle meatus (Figure 2). Biopsy of the mass revealed tissue composed of abundant histiocytic cells admixed with eosinophils, the histiocytic cell stain positively for CD1a, CD68 and S100. Therefore, a diagnosis of Langerhans cell histiocytosis was made. Several other investigations done to stage the disease showed that he had multi organ involvement including right maxillary sinus, right orbit, left parietal and occipital bone and liver.

Chemotherapy was started. Assessment post induction chemotherapy showed good response, therefore he was continued with maintenance chemotherapy of IV Vinblastine and oral Prednisolone. Currently, after a year of completed treatment, child is well and is attending kindergarten school. On follow up noted, his liver enzyme is improving, no sign of recurrence of disease.

DISCUSSION

LCH diseases are usually considered to be diseases of childhood. Langerhans cell proliferation may involve one or many body systems. The head and neck are involved in almost 90 % of cases. However, involvement of paranasal sinus is very rare³.

Our patient presented with proptosis and cheek swelling. He had multi organ involvement including the orbit, skull, sinus and liver. Involvement of the maxillary sinus is extremely rare and to date only 3 cases have been reported³.

The diagnosis of LCH is based on histopathological examination of the biopsy specimens which shows

This article was accepted: 18 October 2012

Corresponding Author: Azreen Zaira A Bakar, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

Email: areein@yahoo.com

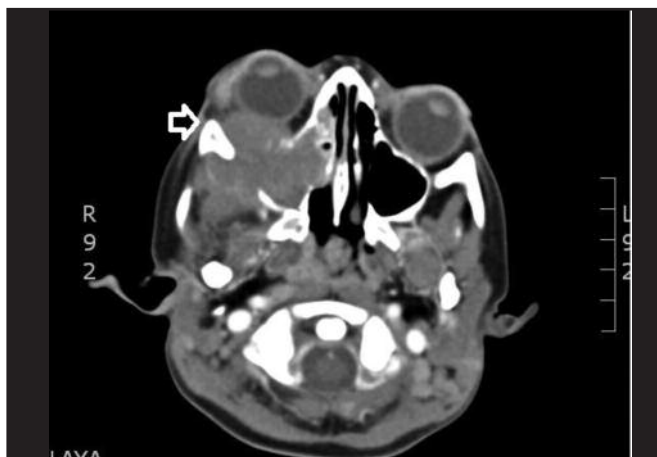


Fig. 1: White arrow shows CT scan findings of enhancing mass arising from right maxillary sinus extending to orbital cavity causing significant proptosis of right eye.

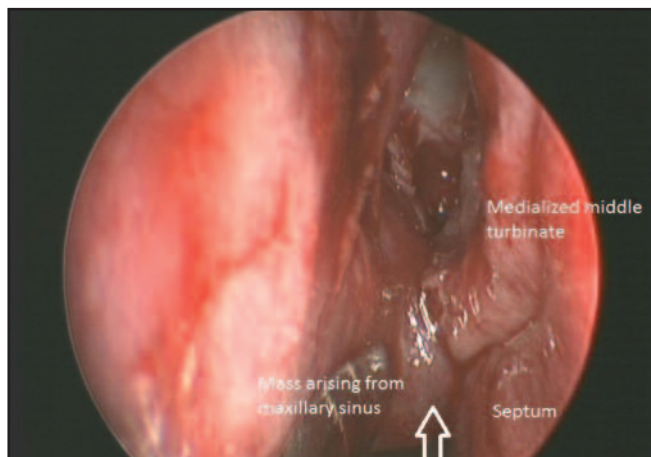


Fig. 2: White arrow shows naso-endoscopic finding of fleshy mass arising from right maxillary sinus.

multinucleated Langerhans’ cells, histiocytes, and eosinophils. In difficult cases, the presence of Birbeck granules on electron microscopic examination or the antigenic markers that react with CD1a glycoprotein and the cytoplasmic protein S100 detected by immunoperoxidase staining are considered diagnostic¹.

Treatment of multi-organ disease is controversial. Several large randomize studies have shown that multi-agent chemotherapy, given in longer duration will have better response rate and less recurrences compared to high dose prednisolone alone⁴. Currently, the LCH-III treatment protocol is probably the most common therapeutic guideline used for patients with multiorgan involvement. The regimen consists of one to two 6-week courses (continuous oral corticosteroids for 4 weeks, plus weekly IV vinblastine) of initial therapy, followed by a maintenance phase (3 weekly pulse of oral prednisolone for 5 days plus IV vinblastine). The addition of a third drug to the standard combination (etoposide or methotrexate) had shown no significant of improvement to the survival⁴. Other therapies currently being investigated include monoclonal antibodies that target CD1a or CD207, specific cytokine inhibitors, and a relatively new agent, 2-chlorodeoxyadenosine⁴.

Curettage of a circumscribed skull lesion with methylprednisolone injection may be sufficient if the lesion is single and does not involve the temporal bone, mastoid or orbital areas⁵. However, if the lesion involved the above bones, then chemotherapy is recommended. For mandible lesion, extensive surgery may destroy secondary tooth development therefore should be avoided. Usage of radiotherapy has been significantly reduced in paediatric

group. This mode of treatment is mainly used in single bony lesion that does not respond to chemotherapy or optic nerve involvement⁵.

The prognosis of this disease is closely linked to the age at onset, involvement of high-risk organs (liver, spleen, lung and bone marrow) and response to initial therapy (assessed after 6-12 weeks of treatment) . Children younger than 2 years old have higher mortality rate than older children. The presences of organ dysfunction and multi organ involvement are a poor prognostic sign. Thus, patients without involvement of risk organs (low-risk group) are not at risk for mortality but need systemic therapy in order to control the disease activity and avoid reactivations¹. The outcome for children with LCH involving low-risk organs has always been excellent, but the major challenge is to reduce the 20% - 30% incidence of relapse⁴. The overall survival rate for all was 79% at 1 year, 74% at 3 years, and 71% at 5 years; however, in patients with liver or spleen involvement 1-year survival was 33% and 5-year survival was just 25%⁴.

REFERENCES

1. Pizzo P, Poplack D. Principles and Practice of Pediatric Oncology (4th edition), Lippincott. 2002.
2. Eckardt A, Schultze A. Maxillofacial manifestations of Langerhans cell histiocytosis: a clinical and therapeutic analysis of 10 patients. Oral Oncol. 2003; 39: 687-94.
3. Bartnick A, Friedrich RE, Roser K, et al. Involvement of the maxillary sinus in Langerhans cell histiocytosis. Mund Kiefer Gesichtschir. 2003; 7: 36-41.
4. Minkov M. Multisystem Langerhans cell histiocytosis in children: current treatment and future directions. Paediatric Drugs. 2011; 13: 75-86.
5. Elizabeth KS, Whitney AH. Langerhans Cell Histiocytosis: A Review of the Current Recommendations of the Histiocyte Society. Pediatric Dermatology. 2008; 28: 291-5.