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Langerhans Cell Histiocytosis: An Unusual Presentation

ABSTRACT

Objective: To describe an unusual presentation of Langerhans cell histiocytosis in the craniofacial skeleton in a patient previously diagnosed with Pott's disease.

Methods:

Design: Case report.

Setting: Tertiary care center.

Patient: One (1)

Result: A 30-year-old male who previously underwent cervical spine surgery for Pott's disease, presenting with watery ear discharge and mandibular resorption was initially diagnosed with tuberculosis. On MRI, a mildly contrast-enhancing soft tissue mass involving the left infratemporal and middle cranial fossae consistent with residual or recurrent tumor as well as an inflammatory process was seen. He underwent transtemporal excision of the mass with external auditory canal blind sac closure and obliteration. Final histopathology revealed Langerhans cell histiocytosis. A review of slides of the specimen from the previous spine surgery was done and signed out as Langerhans cell histiocytosis.

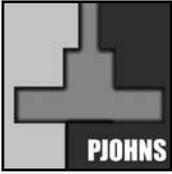
Conclusion: Langerhans cell histiocytosis occurs rarely, but has frequent head and neck manifestations. It may also be confused with other diseases in the head and neck, such as tuberculosis, in this case. Otorhinolaryngologists and head and neck surgeons should be well aware that isolated lesions in the bony framework of the head and neck should include Langerhans cell histiocytosis in the differential diagnosis. Early detection is the key to preventing disease progression and instituting timely definitive management.

Key words: *Langerhans cell histiocytosis, watery ear discharge, mandibular resorption*

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No funding support was received for this study. The authors signed a disclosure that they have no proprietary or financial interest with any organization that may have a direct interest in the subject matter of this manuscript, or in any product used or cited in this report

Presented at the Interesting Case Contest (1st Place)
Philippine Society of Otolaryngology Head and Neck Surgery
Midyear Convention, Legazpi City, Bicol, April 28, 2006



considered, for which cytologic correlation was recommended.

The patient was referred to a neurosurgeon who requested an MRI of the skull base. This showed a mildly contrast-enhancing soft tissue mass in the left infratemporal and middle cranial fossae (Figure 2). A biopsy was the next logical step but there was a dilemma on how to approach the mass.

Another otolaryngologist saw the patient and noted watery, clear, pulsatile non foul-smelling discharge from the left ear. Retrognathia was noted, for which a panoramic radiograph was done, showing total absence of the mandible (Figure 3). An assessment of CSF otorrhea secondary to TB lytic destruction with mandibular resorption was made.

In order to address both biopsy of the primary lesion and treatment of the CSF otorrhea, transtemporal excision of the mass with external auditory canal blind sac closure and obliteration under general anesthesia was done at the Philippine General Hospital. Intra-operative findings include the absence of the posterior bony external auditory canal wall, and an "eaten-out" temporal bone. A fleshy mass was seen in the area of the mastoid cavity. Part of the mass was sent for frozen section which was read as chronic inflammation with gliosis. Near total gross excision of the mass was done. Blind sac closure of the external auditory canal was followed by temporalis muscle flap rotation and obliteration of the temporal bone defect to address the CSF leak.

Final histopathologic diagnosis was Langerhans cell histiocytosis. A review of slides of the specimen from the previous spine surgery was requested, which likewise revealed Langerhans cell histiocytosis. To strengthen the diagnosis, immunostaining with S-100 protein was done. Both specimens were positive.

The post-operative course was uneventful except for episodes of dizziness and vomiting. The patient was discharged after 12 days, but was again lost to follow-up. Despite repeated calls, the patient refused the recommendation to undergo chemotherapy.

DISCUSSION

Langerhans cell histiocytosis (LCH), formerly known as histiocytosis X, denotes a group of diseases characterized by infiltration or proliferation of histiocytes in various body tissues. The clinical spectrum includes the acute fulminant, disseminated Letterer-Siwe disease, solitary or few, indolent and chronic, lesions of bone or other organs called eosinophilic granulomas, (under which the patient may fall) and the intermediate form

called Hand-Schüller-Christian disease.²

LCH is seen in different ages, but a clear predominance exists for the pediatric age group.³ It is a rare condition in adults.⁴ Thus, a 30-year-old male presenting with otorrhea, mandibular resorption and cervical spine erosion does not typify a patient with Langerhans cell histiocytosis.

Destruction of the temporal and mastoid bones manifest as mastoid swelling, middle ear polyps, and in this case, CSF otorrhea, and erosion of the posterior bony external auditory canal. Infiltration of the mandible will present with loose teeth and eventual resorption as seen in this patient. Vertebral collapse with spinal cord compression has occasionally been described, which may have happened in the patient when he was initially diagnosed with cervical Pott's disease four years prior to the onset of watery ear discharge.

There may be confusion with Langerhans cell histiocytosis and other diseases in the head and neck, particularly granulomatous inflammations, such as tuberculosis (TB). Many of the clinical features of Langerhans cell histiocytosis mimic this highly endemic disease in the Philippines.⁵ As seen in this case, radiographs were not able to differentiate between TB and LCH. Even the final histopathology of the cervical lesion was initially read as tuberculosis.

Both Langerhans cell histiocytosis and tuberculosis may present with multisystem involvement. Symptoms in this patient presented in a sequential manner with cervical erosion, followed by mandibular resorption, and finally otorrhea, which is more in accordance with Langerhans cell histiocytosis.⁶ In the case of tuberculosis, symptoms usually appear simultaneously although progressive multi-organ involvement sequentially occurs in many cases.

Histologically, a typical tuberculous granuloma would normally be described as chronic granulomatous inflammation with caseation necrosis and Langhans' type giant cells mixed with chronic inflammatory infiltrates consisting of lymphocytes. (Figure 4a) In the patient's cervical spine specimen, cells were arranged in a granuloma-like pattern, and as such may have been mistaken for a tuberculous granuloma. (Figure 4b) Both diseases show aggregates of histiocytes. Histiocytes of tuberculosis are epithelioid and spindle-shaped. (Figure 5a) On the other hand, histiocytes seen in Langerhans cell histiocytosis are atypical, in that they are large, ovoid, mononuclear cells. (Figure 5b) Aside from this, LCH also presents with a large component of eosinophils in contrast to lymphocytes seen in TB.⁷

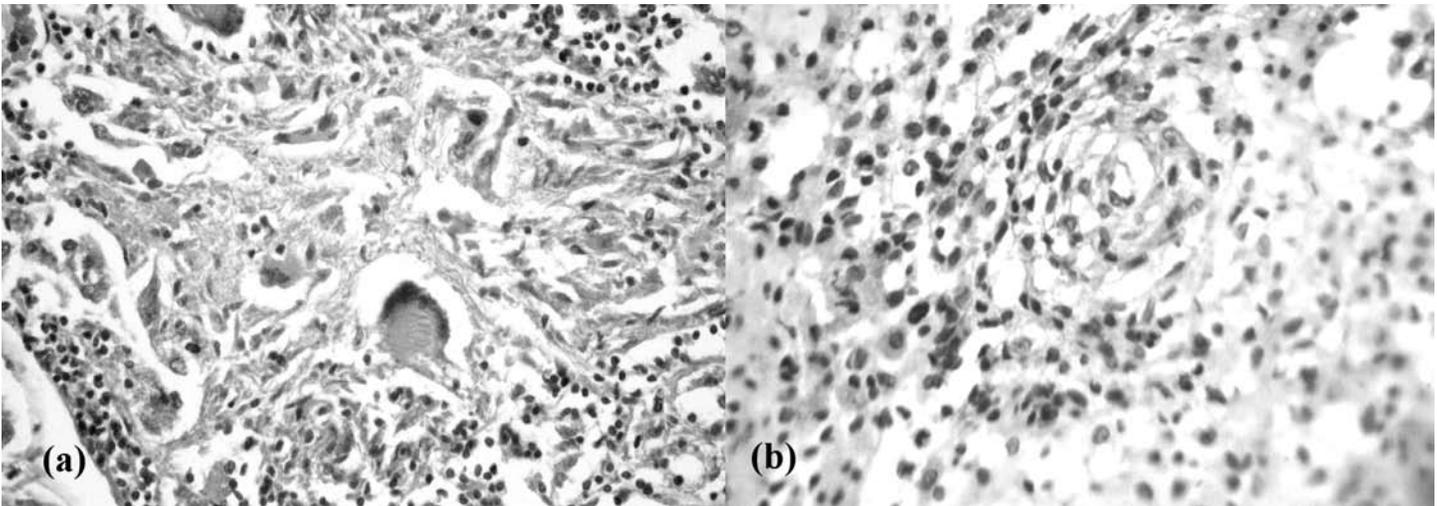


Figure 4. H&E photomicrographs. (a) Typical tuberculous granuloma versus (b) granuloma seen in the cervical spine specimen of the patient.

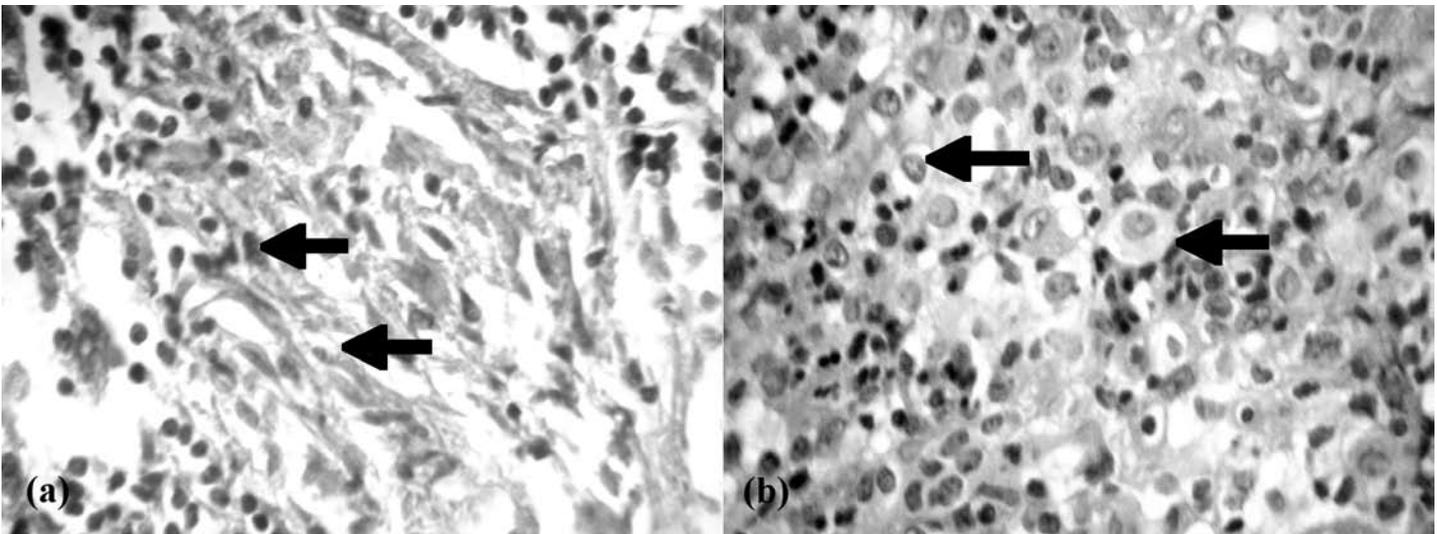


Figure 5. H&E photomicrographs of tuberculosis specimen and LCH specimen of the patient. Histiocytes (arrows) of tuberculosis (a) versus histiocytes (arrows) of Langerhans cell histiocytosis in the specimen of the patient (b).

S-100 staining strengthened the diagnosis of LCH in this case. Both normal and diseased Langerhans cells will stain positive for S-100. Tuberculosis cells do not.

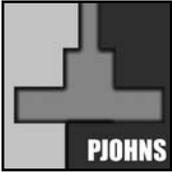
The similarities and differences between Langerhans cell histiocytosis and tuberculosis noted in this case are summarized in Table 1.

The initial misleading histopathologic diagnosis of tuberculosis of the cervical spine specimen allowed progression of the disease to involve the mandible and then the temporal bone. The patient's poor follow-up record compounded this progression, adding to delays in diagnosis and treatment.

Transtemporal excision of the mass with blind sac closure of the external auditory canal was considered for the definitive

Table 1. Similarities and differences between Langerhans cell histiocytosis and tuberculosis noted in the case

	Langerhans cell histiocytosis	Tuberculosis
Multisystem involvement	Present	Present
Appearance of symptoms	Sequential	Often simultaneous, may be sequential over time
Histologic arrangement of cells	Granuloma-like pattern	Chronic granulomatous inflammation with caseation necrosis and Langhans' type giant cells
Appearance of histiocytes	Atypical; large, ovoid mononuclear cells	Epithelioid, spindle-shaped
Inflammatory infiltrates	Eosinophils	Lymphocytes
S-100 reactivity	Positive	Negative



management of CSF otorrhea in this case while, at the same time, obtaining a good specimen for biopsy.

In retrospect, had a correct diagnosis been made at the time of cervical spine surgery, the resultant CSF otorrhea could have been prevented. Though initially managed by topical drops, the diagnosis of CSF otorrhea and definite management with external ear canal blind sac closure was very important. The latter is most relevant in preventing the possible life-threatening condition of meningitis. However, the contralateral temporal bone is likewise at risk. Definitive treatment, in the form of chemotherapy consisting of low to moderate doses of methotrexate, prednisone, and vinblastine hopefully can save this patient from further lytic destruction of other bony sites.⁸ This option has been presented to the patient, as well as the possible side effects of chemotherapy, including hair loss and vomiting. The patient was not open to these particular side effects, thus refusing this therapeutic option.

Otorhinolaryngologists and head and neck surgeons should be well aware that isolated lesions in the bony framework of the head and neck should include Langerhans cell histiocytosis in the differential diagnosis because, as in the case of the different diseases that have plagued mankind throughout the centuries, early detection is still the key to preventing disease progression with timely definitive management.

ACKNOWLEDGEMENT

The authors wish to thank the following individuals from the University of the Philippines College of Medicine - Philippine General Hospital without whose help this paper would not have been possible: Dr. Jose Maria Avila, Chairman, Department of Pathology who was our resource person for pathology; Drs. Joselito Jamir and Romeo Villarta Jr. former chairman and consultant, respectively of the Department of Otorhinolaryngology, who were our scientific advisors; Dr. Gerardo Legaspi, consultant, Department of Neurosciences Section of Neurosurgery who was the co-managing neurosurgeon; and Dr. Sherjan Kalim, resident of the Department of Pathology for assisting with the photomicrographs.

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