An overlooked case of mandibular histiocytosis: Case report and review of the literature

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ABSTRACT

Introduction: Langerhans histiocytosis is a rare disorder resulting from the proliferation and accumulation of a specific histiocytic cell called the Langerhans cell. Mandibular histiocytosis specifically can be asymptomatic or manifest as a jaw mass, Orthopanthogram as well as CT are required for the early diagnosis and prevention of total destruction of the osseous bone. Case Report: We report the case of a 7-years-old patient, with a history of a non-resolving right jaw swelling treated as a dental abscess with antibiotics and analgesics for 14 days. The patient has no history of trauma, and was referred to our department for a right jaw swelling evolving for three months. Oral examination found an ulceroproliferative process of the posterior aspect of the dental part of the right mandible. Dental contrast enhanced CT showed a destruction of the periodontium, with an extensive geographical osteolytic homogeneous mass destroying the mandible. The diagnosis of histiocytosis was suggested histologically after biopsy. After a 6 weeks regimen of vinblastine and prednisolone with a clear regression, a surgical curettage of the bone lesion was performed. Local control was achieved with no complications in the six months follow up examination. Conclusion: Our case suggests that rapid diagnosis followed by minimally invasive surgery can be determinant in the prevention of possible deformation, especially in the pediatric population. Further investigation of the role of microbiopsy for screening should be discussed. Computed tomography is the first choice imaging technique in the diagnosis of mandibular histiocytosis.

Keywords: Computed tomography, Histiocytosis, Lytic bone, Mandible

INTRODUCTION

Langerhans histiocytosis is a rare disorder resulting from the proliferation and accumulation of a specific histiocytic cell called the Langerhans cell. These cells, which are produced in the bone marrow, can be considered very important components of the cell-mediated immune response, acting as antigen presenting cells. It may occur at any age; and is mainly found in children and young adults however, it remains a childhood disease [1, 2].

The polymorphism of the disease is variable, LCH can present from a multi system disorder to an isolated bony lesion. In imaging, bone lesions are mostly found. Which makes radiology more essential to diagnostic process as well as an important tool for differentials of jaw bone lytic lesions [1, 2].

The jawbone localization can be suspected upon the discovery of an ulceration or gingival tumefaction and is characterized by teeth mobility and fall. X-rays shows destruction of the periodontium with progression to an osteolysis of the mandible [1].
CASE REPORT

A 7-years-old patient, with a history of a non-resolving right jaw swelling treated as a dental abscess with antibiotics and antalgics for 14 days. The patient had no history of trauma, and was referred to our department for a right jaw swelling evolving from three months.

The clinical examination found a painful non-inflammatory left mandibular mass measuring 15 mm, with a mastication difficulty. This mass was hard, immobile and attached to the deep plane.

Oral examination found an ulceroproliferative process developing in the posterior aspect of the dental part of the right mandible as well as a loss of an adjacent tooth.

Dental contrast enhanced CT showed a destruction of the periodontium, with an extensive geographical osteolytic homogeneous mass destroying the mandible, with a density of 40 HU enhancing to 110UH after contrast administration, and a floating teeth. It measured 17x18x19 mm with 21x22 mm of mandibular bone loss (Figure 1, Figure 2).

The diagnosis was confirmed histologically after biopsy samples.

A surgical curettage of the bone lesion was performed after a six weeks regimen of vinblastine and prednisolone with a clear regression.

DISCUSSION

Histiocytosis is a polymorphic condition related to the proliferation of dendritic cells in different tissues and organs. Among facial bone sites, the most common localization in patients under 20 years of age is the mandible [3]. It is a clonal pathology of unknown etiology, involving protein mutations in the MAP kinase pathway [4, 5]. The diagnosis is often delayed due to a multitude of differentials, and to this day, no screening test has been approved.

Histiocytosis are related to the proliferation of cells of the monocytic system and are divided into three groups [6]:

- Langerhans histiocytosis: characterized by a proliferation of Langerhans cells that can affect one or more organs, more frequently bone and skin.
- Non Langerhans histiocytosis: which is part of Rosai-Dorfman disease, are much more heterogeneous. Its typical presentation consists of cervical lymphadenopathy with a particular histiocytes infiltration.
- Malignant histiocytoses.

There are multiple clinical entities with a common histological substratum, localized eosinophilic granuloma (isolated benign lesion), Letterer-Siwe disease (rapidly progressive multi-organ involvement) and Hand Schuller’s Christian disease. (progressive condition with diabetes insipidus, intracranial bone deficiencies and exophthalmos) and transition forms which associates multiples lesions and locations [3].

The Histiocyte Society proposes a classification based on age, the number and topography of lesions criteria [7]:

- Type 1: unifocal involvement.
- Type 2: Multi-focal involvement without liver, splenic, pulmonary or spinal cord injury, and aged 2 years and over.
- Type 3: Multi-focal involvement with at least one impairment of the following four organs: liver, spleen, lung, marrow, or less than 2 years old.

We describe a case of a type 1 mandibular histiocytosis of the histiocyte society classification.

Langerhans histiocytosis preferentially affect children. The damage can be limited to an organ; more frequently skin and bones, as well as disseminated, the spread of lesions and the existence of organ dysfunction condition the prognosis. The distribution of lesions shows axial and cephalic predominance [8]. The lesion is unique in 70 to 75% of cases. Mandibular localizations are considered common. They represent 20% of non-odontogenic tumors. Mandibular involvement is much more common than that of the maxillary. The clinical manifestations depend on the number of lesions, their location in the mandibular bone and their extension to the soft tissues [3].

The diagnosis is based on the proliferation of CD1a-positive Langerhans cells or positive staining for lesional
cells with langerin (CD207) as it has demonstrated that these findings confirms the presence of Birbeck granules. The presence of Birbeck intracytoplasmic granules demonstrated by electronic microscopy is no longer necessary for the diagnosis [9].

The symptoms that point to the diagnosis are pain, the appearance of a swelling or loosenings of the teeth, mucosal ulceration, gingival bleeding, erythematous masses, periododontitis, candidiasis, precocious eruption of teeth and more rarely a pathological fracture [10]. A jaw swelling resistant to antiinfection with an ulceration proliferative lesion was the case for our patient.

The typical radiological image is a lysis beginning in the medulla of a long or flat bone. Usually histiocytosis affect long bones in children, isolated lysis of flat bones (mandibular) is more preponderant in the adult population. Sometimes the radiological aspect is worrisome, with a cortical rupture and a periosteal reaction suggestive of Ewing’s sarcoma. The lesion can in some instances reach the epiphyseal portion of a bone involving the cartilage [8].

The analysis of the transition zone, the margins and the periosteum is essential, it allows for the evaluation of the aggressiveness of the lesion and classify it according to Lodwick [9]:

- **Type I: Geographic.**
  - IA: Thin, sclerotic margin.
  - IB: Distinct, well-margined border, but not sclerotic.
  - IC: Indistinct border.
- **Type II: Moth-eaten.**
- **Type III: Permeative.**
  - IA and IB, which are characteristic for slow lesion growth, are the most encountered cases of Langerhans histiocytosis. In our case, the radiologic appearance of the lesion was suggestive of a Lodwick type IB.

Local complications may include pathological fracture, ocular compression and dental instability, functional outcomes are usually limited [8].

Nevertheless, since there is no proven link between the number of bone lesions, their inflammatory nature and the prognosis of the disease, a limited approach to bone assessment with a complete skeletal radiography is recommended [8]. Usually, a complete skeletal examination is performed in the initial evaluation, while during the reactiavtions of the disease, X-rays are centered on symptomatic anatomical regions [8, 11].

Computed tomography better analyze bone lesions

- It confirms osteolysis and the degree of bone destruction.
- Study the tumor matrix.
- Analyze the periosteal reaction.
- Extension to adjacent soft tissues after injection.

Typically, lytic intramedullary lesion homogeneous ranging from 40 to 50 HU in density, with an enhancement between 110 and 130 UH which is very characteristic. Severe lysis may produce a “floating teeth” aspect [1, 2]. MR depict a lesion with extra medullary extension decreased or intermediate T1 signal intensity and increased T2 signal intensity with a very intense enhancement after injection of gadolinium. MRI also makes it possible to better explore soft tissues involvement, which is often underestimated by CT [1, 2].

The treatment of LCH (Langerhans cell histiocytosis) is still depends on the extent and the severity of disease at diagnosis. The LCH Study Group adopted a stratification system, which separated patients into two major categories [12]:

- ‘Single-system’: subdivided further into single and multiple site.
- ‘Multisystem’: defined as involvement of two or more organs at diagnosis with or without organ dysfunction, this group was further divided into a ‘low-risk’ and a ‘risk’ group.
  - Low-risk patients (20% of patients), have an excellent prognosis and are characterized by the absence of involvement of ‘risk organs’ such as liver, lungs, spleen or hematopoetic system.
  - ‘Risk’ patients (80% of patients) have at least one or more risk organs involved and a high mortality rate.

A number of therapies were preconized among them:

- Local therapies such as intralesional instillation of steroids is an effective and safe treatment modality when therapy is required for a limited number of bony lesions.
- Systemic therapies include indomethacin, biphosphonates and chemotheraphy.
- Indomethacin, a PG-E2 inhibitor, was found to be a useful therapy for single or multiple LCH lesions. Whether it acts by slowing disease progression or simply as an analgesic allowing time for spontaneous healing is still unclear.
- Biphosphonates have also been found useful for pain control and possibly control of progression of bone LCH [13].

The Histiocyte Society considers an initial six-week course of therapy with vinblaztine and prednisone to be effective treatment with minimal toxicity and is therefore suggested for all patients with multi system LCH; regardless of risk organ involvement [13].

We prescribed for our patient a combination of prednisolone and vinblastine for 6 weeks followed by a surgical curettage of the residual tissue. There were no complications nor recurrence in the three and six months follow up.

**CONCLUSION**

In conclusion, our case suggests that rapid diagnosis followed by a six weeks regimen of vinblaztine and prednisolone and a surgical curettage can be determinant in the prevention of possible recurrence, especially in the pediatric population. Further investigation of the role of micro biopsy for screening should be discussed. Completed
tomography is the first choice imaging technique in the diagnosis of mandibular histiocytosis.

REFERENCES


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Author Contributions

Kaoutar Imrani – Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Othman Ayouch- Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Allali Nazik – Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission
The corresponding author is the guarantor of submission.

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Consent Statement
Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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