

MASSIVE OSTEOLYSIS OF THE MAXILLO-FACIAL BONES : CASE REPORT AND REVIEW OF THE LITERATURE

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INTRODUCTION

Osteolysis of the facial bones is usually the result of a specific pathologic process, such as a periodontal and periapical infection, odontogenic cysts or neoplasia; less commonly, it is the result of an endocrine, metabolic or immunology disorder. In 1838, JACKSON (1) described massive osteolysis as an extreme rare disease entity of an unknown aetiology and uncertain pathogenesis. This disease is characterised ultimately by progressive resorption of contiguous structures (especially bones) and their replacement by fibrous vascular connective tissue. The predilection sites of involvement are the pelvis, the shoulder, the scapula and the clavicle. This process is usually monostotic but may occasionally be polystotic in character.

A review of the medical world publications has showed less than 200 cases of massive osteolysis involving virtually every bone in the body. Of these only 37 cases (2 - 26) have been found to affect bones of the maxillo-facial region.

This paper reports an unusual aggressive form of massive osteolysis, which originates from the left side of the mandible, extended to the maxilla, zygoma, orbit, frontal, temporal and occipital cranium, skull base and the upper cervical spine. The clinical evolution of this disease was about 4 years.

OUR CASE

Abdellah K. is a seventeen-year-old adolescent who was admitted to hospital in July 1997. He was immediately put under fastidious medical care in our department for he suffered from a massive temporo-maxillo-malar lysis. Bearing no prior pathological antecedent - personally or

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genealogically - this patient turns out to be a unique case. As a matter of fact, his June 1996 check-up indicated a gradual loss of the normal jawbone stiffness - a loss which was manifested first at the level of the zygomatic part, then it extended to the temporal bone and the mandible; eventually it reached the left maxilla. Such diagnosis did not seem to worry the patient enough to seek medical assistance, since there were no other related, physical symptoms; in other words, he did not feel any local pain, functional impairment and forewearing inflammatory signs.

After one full year of maturation, the patient started to feel a soft swelling tumefaction within the same area, generating hot spells foregrounding an imminent abscess. Such identification explained the patient's complaints about prolonged fever and a kinetic gingivitis, on one hand ; and an eventual teeth loss on both left maxilla, which entailed purulent otitis and rhinitis on the same side, as well as a compressed oral cavity, on the other. The neurological and ophthalmological tests, as well as the conjured somatic ones proved negative, though.

After consultation, we were inclined to a draining puncture of the tumefaction, which yielded 500 cc of purulent secretions. Hence, a bacteriological test recorded a viridens Streptococcus. Such draining helped reduce the tumefaction, but instead caused a depleting spot. Accordingly, the patient was prescribed an antibiotics treatment, together with a series of congruent tests : namely the standard radiography tests (Blondeau, Hirtz and contours of the skull base) and CT scan. The results revealed a lysis of the upper maxilla, the cheek bone (malar), the zygomatic arcade, the temporal bone, the larger wedge of the sphenoid, the occipital bone, as well as the ascending ramus of the left mandible. Closely related to this osteolysis, we could locate an expanding process of a hyper dense and intertwined tissue at the level of the infratemporal fossae where the temporal ear lobe was causing hernia.

Meanwhile, other biological and radiological check-ups were proctored as supplementary test towards a probing

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of an either hydatidical, metastatical or inflammatory etiology, but to no avail. Again, due to the negative results of the bacteriological and mycological tests, we had to ride out the osteitic hypothesis pertaining to specific germs, such as tuberculosis, actinomycosis or mucomycosis as potential causes of the impairment. Eventually, we vainly narrowed down our diagnosis to far-fetched causes such as dysembryoplastic or even local malignant tumor.

In due course, retro-auricular surgery in the surrounding areas of appending injuries was then recommended. The results revealed a total disappearance of the bone form within the temporal area. Instead, a fibrous tissue containing numerous, tiny vesicles, full of liquid substances, sloped upward the fading bone. Pushed out onto the edge, the bone looked delicately thin and brittle. We then removed a sample of the bony and fibrous tissue, which we subjected to a rigorous anatomopathological scrutiny. Thereafter, we found out that the blond-vascular system contained a number of fissures (cracks). Some of these vessels were blond-dry and partially covered with a sustained endothelium. The fissures were embedded in a fibrous stroma, hardly cellular, lacking any inflammatory infiltration. With such data at hand, we set up an initial lymphangiomatosis as a diagnosis, which was confirmed after a second reading of the slides.

Faced with an obvious discrepancy between the profuse bony lesions and the holistic diagnosis, we resolved to a further radiological check-up. This Magnetic Resonance Imagery (MAI) test revealed a diffuse progression with heterogeneous signets. It consisted mainly of a relatively thick, fleshy component posed in a level-headed sequence (T2), disseminating flabby soft parts in both the jugular areas and the temporo-mandible and causing hernia of the temporal lobe in the adjacent left infratemporal fosse and secreting a multikystic liquid substance that lay at the level of the retroauricular soft parts, which in their turn helped infiltrate the vasculonervous-retrostylian cluster, the intrapetrous carotid and pterygoidian muscles.

The cerebral parenchyma and the ocular globe were normal, though.

Still, an MTD/ technecium scintigraphy was required; it showed hypofixation of the radioactive plotter at the level of the maxilla-malair and the zygomato temporo-mandible areas. No further anomalies on the rest of the skull were indicated.

After a seven-month recess, the patient noted the reappearance of the temporal and facial tumefaction, paired

with tense painful muscular contraction of the back of the neck muscles and a diplopia of the left eye. Henceforth, a second draining puncture of the tumefaction was carried out. The apriori bacteriological test abstracted a purulent liquid. The microb isolated was Staphylococcus negative coagulase. The antibiotics treatment prescribed, helped improved the patients state clinically. That is why we carried out a 3D - tomodensitometric check-up, which specified an extension of the initial osteolytic lesions, more precisely at both the external orbital part of the occipital bone and the posterior arch of the atlas (C1) within the cervical spine

The latter's infection impelled us to maintain the patients cervical spine in an upright position using a surgical collar before resorting to a second surgery, this time using lesional biopsy, hoping to locate lesions whose anatomopathological outcome could explain this massive and idiopathic osteolysis. The expected result would reveal the existence of a hyaline-fibrous tissue, barely cellular, that contained a scarce number of regular fibroblasts. It was covered with empty and completely anastomosed sockets. In scattered spots, however, these sockets were excessively intertwined with regular endothelial cells. Within this fibrosis, we could also notice the remains of some osseous sequestration, together with a lack of any distinct infiltrating inflammatory cells.

These findings stood in contrast with the lymphangiomatosis, diagnosis initially reported. The GORHAM-STOUT diagnosis, accurately relevant to the clinico-radiological and histological data, was therefore maintained.

In May 1999, our patient was prescribed an external radiotherapy treatment with an optimal dosage of 35 Grays, tabulated over seventeen sessions. During the last check-ups, three and six months later, we could notice an improvement of the apparently infected temporo-facial array, which had strated to regress partially under the effect of antibiotics treatment. The extension of the osteolysis lesions within the occipital bone and cervical spine were more important and treatment with non-steroidiens anti-inflammatory were prescribed in order to reduce the daily pain.

DISCUSSION

Massive osteolysis or GORHAM's disease was first defined as a specific entity by GORHAM in 1955 (3). It develops in any part of the skeleton but the process affecting the facial bones was first presented by ROMER (2) in 1924. The mandible is one of the most common

bones concerned and its invasion undergoes partial or complete dissolution and spreads across joints to contiguous bones or involves the incumambient soft tissues (24, 25, 26).

Until the present time only 39 cases have been reported in the world publications, our case is an additional one. Approximately, the disease appears to originate initially from the mandible, in all cases, and spreads over. The mandible was affected alone by the osteolysis, partially or completely, in 23 cases whereas the maxilla was never involved alone. These cases reported to affect multiple contiguous bones of the head represent a more advanced stage of the disease and they have occurred in 14 cases. The age on onset has been reported from 1 month to 75 years, with a peak in the second and third decades. There is no particular sex or race and no associate endocrine, metabolic or immunologic disorder (26).

The authors described two phases in the massive osteolysis. The first is the active form of bone resorption and its progression. Clinically, it produces often the loss of the teeth, pathological fractures with mild pain ; less frequently tissue swelling and sepsis. The recurrent infections may reach the adjacent organs such as the middle ear, inner ear and meninges. The second phase represents the latent form. Moreover, bone defects are associated with both several and grave dysfunctions consisting of mastication, swallowing, speaking and respiratory functions and a notable facial deformity. The course and duration of the phases cannot be foreseen and the active form can last for months or years (22, 23, 25).

The radiological findings of the GORHAM's disease are not specific. JOHNSON and MC CLURE (27) noted in the early X-ray the evidence of one or multiple centromedullary and subcortical radiolucencies, usually with indistinct margins and no sclerotic borders. Later, these lesions can enlarge and coalesce together causing a disruption of the cortex than an intraosseous and extraosseous resorption. SAGE and ALLEN (28) have postulated that the osteolytic process was originated from the cortical bone and involved into the center.

Computerised tomography and magnetic resonance scans can more accurately detect the extent of the lesion to contiguous bones, particularly to adjacent soft tissues. According to DOMINGUEZ (29), the existence of patchy areas of inhomogeneously increased and decreased signal on T1 - T2 weighted sequences may represent hemorrhage at different stages. However, a nuclear bone revealed no increased activity in the-area (except at the-

advancing edge of the lesion) and disclosed no abnormalities in the remainder of the skeleton which usually shows a normal uptake. The values of haematological and biochemical tests often remain within normal parameters (24, 25, 30, 31).

The histopathological characteristic of the GORHAM's disease is the replacement of bone tissue by intrafibrous connective tissue containing many thin-walled vessels or anastomosing vascular spaces, including lymphatic capillaries; termed histologically hemangiomas as used by STOUT (3). Osteoclasts are generally but not always present and their number is, however, usually low (25).

Assessment of the diagnosis is difficult because of the rarity of the disease and there isn't any causative factors of bone's lesion identified. Therefore it is based mainly on histopathological examination of the lesional tissue correlated with clinical data.

Other conditions with bone facial lysis may be considered in the differential diagnosis, particularly the osteolysis in relation to infection, osteomyelitis or haemangioma of bone, metastatic malignancy, RECKLINGHAUSEN disease, fibrous dysplasia or primary osteolytic diseases of bone. Those occur rarely and involve hereditary multicentric osteolysis with dominant or recessive transmission ; no hereditary multicentric osteolysis with nephropathy or Winchester syndrome (TORG and al, PHERSON and al) (31).

HEFFEZ (32) and al suggested eight criteria for definitive diagnosis of massive osteolysis.

1. Positive biopsy in terms of angiomatous tissue presence,
2. Absence of cellular atypia,
3. Minimal or no osteoclastic response and absence of dystrophic calcifications,
4. Evidence of local bone progressive resorption,
5. Non expansive, non ulcerative lesion,
6. Absence of visceral involvement,
7. Osteolytic radiographic pattern,
8. Negative hereditary, metabolic, neoplastic, immunologic and infections aetiology.

The pathogenesis of GORHAM's disease remains mysterious. Important number of theories has been proposed postulating the role of the osteoclast activity in the osteolysis (HEYDEN and al, 1977; DOMINGUEZ and MUNDY, 1980). However, the histological findings have been discordant. Most studies reported that osteoclastosis

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was rarely associated with massive osteolysis (22, 33, 34). The GORHAM and STOUT's standpoint that angiomatosis consisted of congenital haemelymphangioma-tosis dysplasia with vascular component, hyperplasia, arterio-venous shunts and might be caused by active hyperhemia, mechanical force, change in pH locally and other unknown causes (34). Recently, elevated levels of interlock in n°6 (IL-6) were detected in the earlier course of the disease. That theory leads to suggest its role in the enhanced osteoclast activity (35). Another theory has advanced the possibility of agenesis of thyroid C cells which secrete calcitonin ; this agenesis could induce osteolysis (36).

The relatively good prognosis often shows spontaneous

arrest of the pathological process, but becomes much worse if there is involvement of the spine or chest (37). Bone regeneration is not usual but it seems possible fewer than 10 cases in all the publications (34).

Surgical intervention and radiotherapy are the suggested methods of treatment. Removal of the pathological angiomatous tissue was generally inefficient. Radiotherapy provided better results in selected cases with a total dose of 35 to 45 grays (21, 23, 25, 38). But the possibility of spontaneous arrest precludes any conclusion regarding to the efficiency of irradiation. Surgical reconstruction should always be carried out in the inactive phase because homologous grafts may also undergo resorption.

Figure 1 : Standard radiography showing complete disappearance of the upper maxilla, the malair and of the left body of the mandible

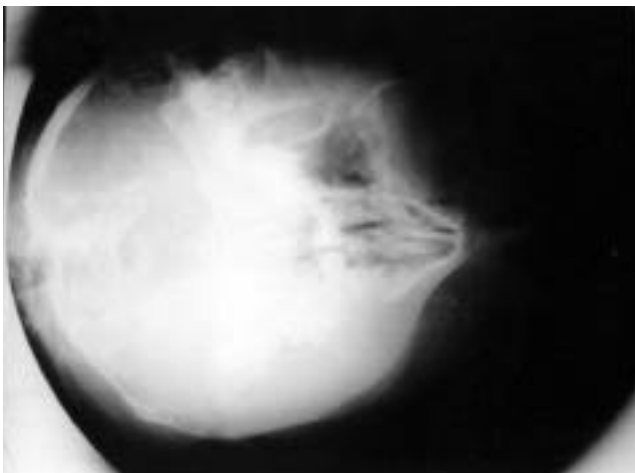


Figure 2 : Ct scan (3d-reconstruction) frontal view : Extensive resorption of the left mandible, maxilla, zygomatic bone, orbital and temporal cranium

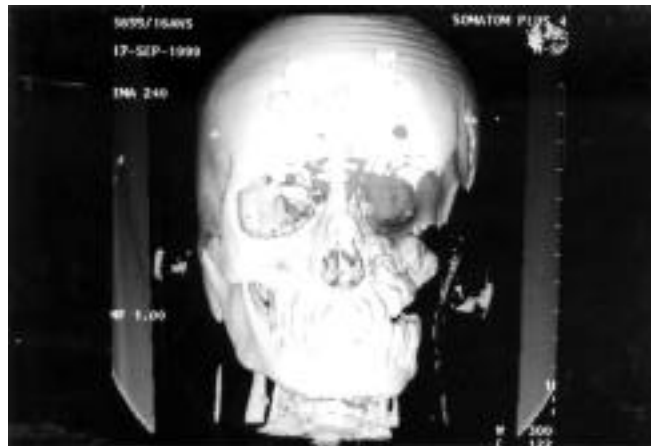
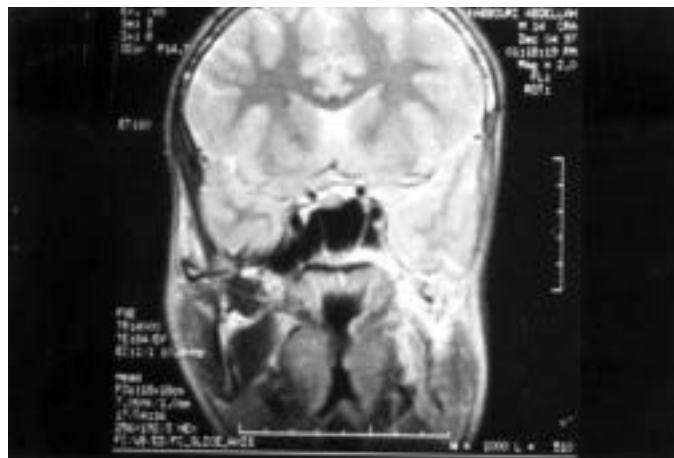


Figure 3 : Mri (t2) showing hernia of temporal lobe (middle fossa) through large bone defect



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ABSTRACT

An unusual case of massive osteolysis destroying the left side of the mandible, the maxilla, the orbit, the cranium bones and the upper cervical spine is reported. The evolution of this disease was observed over a period of 4 years. The literature was reviewed, only 39 cases have been found involving the maxillo-facial bones. In our case, attempts at surgery was without success. However, further radiotherapy with 35 Gy controlled the progression of this osteolysis.

Key words : *GORAHM - Osteolysis - maxillo-facial bones - cervical spine - angiomatosis*

RESUME

Il s'agit d'un cas très particulier d'une ostéolyse massive de toute l'hémi-mandibule gauche, du maxillaire homolatéral, de l'orbite, du temporal, du rocher et de la partie haute du rachis cervical. Ce cas survenant chez un adolescent de 17 ans, évolue depuis plus de 4 ans.

Avec l'aide d'une large revue de la littérature médicale, nous avons constaté l'extrême rareté de cette affection dont seulement 39 cas sont actuellement reportés dans sa localisation maxillo-faciale.

La chirurgie dans ce cas est totalement inefficace. Seule la radiothérapie à la dose de 35 Gy semble limiter l'évolution inexorable de cette maladie et de ses déformations.

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