Case report

Neglected ameloblastomas: a case report

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Abstract – The aim of this article is to present a case of neglected ameloblastoma due to socio-economic status problem. Ameloblastoma is a benign odontogenic tumor though locally aggressive that usually occurs in the mandible. Its insidious development makes the diagnosis more or less late. Negligence plays an important part in the management of this pathology. It leads to a big tumor whose treatment will be more disabling without more expensive reconstructive surgery.

Mots clés :
améloblastome /
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Résumé – Améloblastomes négligés : présentation d'un cas. L'objectif de cet article est de rapporter un cas d'améloblastome négligé en raison d'un contexte socio-économique défavorable. L'améloblastome est une tumeur odontogène bénigne mais localement agressive, intéressant surtout la mandibule. Son évolution insidieuse rend souvent son diagnostic assez tardif. La négligence peut jouer un rôle important dans son évolution. Elle conduit à prendre en charge une tumeur très volumineuse dont le traitement est invalidant, même avec une chirurgie reconstructive coûteuse.

Ameloblastoma is a benign odontogenic neoplasm of the jaw whose development is locally aggressive. The most common location is on the mandible. Its development is generally slow with a progressive bone blowhole and cortical thinning. The clinic is very characteristic by the existence of a painless swelling associated with progressive multiple teeth mobility and movement with no sensory impairment [1]. In Madagascar, due to the insidious nature of the injury and socio-economic constraints of the country, problems ranging from diagnosis delays to management difficulties are very common.

We report a case of neglected mandibular ameloblastoma that care was performed in a hospital located far from the patient's home village.

Case report

In January 2009, a 36-year old man is sent for a huge tumor lasting for about ten years, involving the entire right mandible. The patient does not complain about pain, sensibility disorder, lockjaw or other functional disorder.

The skin of his whole face skin is normal, except for a scar, sequela of odontogenic cellulitis fistula on the left side of the mandible region (Figs. 1–3).

In intraoral, there are many dental malpositions with mobilities on the remaining right mandibular teeth, from 41 to 45. There is a fistula on the lateral mucosa of the 45 with bloody clear liquid issue. The existence of scale on almost all

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Figs. 1, 2 and 3. Ameloblastoma interesting the entire right mandible. Note the scar of old fistula of former odontogenic cellulitis on left side of the mandible region (red arrow).

Figs. 1, 2 et 3. Améloblastome intéressant toute l'hémimandibule droite. Noter la présence d'une cicatrice due à la fistule d'une ancienne cellulite d'origine dentaire dans la région génienne basse gauche (flèche rouge).

Figs. 4 and 5. Scan cuts showing the symphyseal injury (Fig. 4) and the loss of the right coronoid and condylar processes (Fig. 5). The tumor is well limited.

Figs. 4 et 5. Coupes tomodensitométriques montrant la limite symphysaire de la lésion (Fig. 4) et la disparition des processus coronoïde et condyliens droits (Fig. 5). La tumeur est bien limitée.

teeth and decay on nineteen teeth, among them, five are in root debris state is very remarkable.

Blood tests show no abnormality. The histological examination of the biopsy shows a plexiform ameloblastoma without malignancy sign. The surgery consists of a hemimandible resection.

Discussion

Ameloblastoma is a benign odontogenic tumor of unknown cause. Human papilloma virus (HPV) notion is mentioned,

especially in cystic forms [2]. Its incidence and behavior makes it the most important of odontogenic tumors [3-4]. Both sexes can be affected equally and it usually appears from the third decade, but patients under twenty may be encountered [1-5]. Negligence may cause significant morbidity and occasionally death [1].

In Madagascar, clinic and management of this pathology depend on two main parameters: its insidious development and socio-economic condition such as the cost of treatment and distance. This patient is a poor farmer living more than 700 km far from the hospital where care should be done. Negligence

Fig. 6. Plexiform ameloblastoma: odontogenic epithelium is arranged as a tangled network of anastomosing strands and cysts of various sizes.

Fig. 6. Améloblastome plexiforme : l'épithélium odontogène est disposé en réseau enchevêtré de brins anastomosés et de kystes de différentes tailles.

Fig. 7. Symphyseal bone resection of the tumor. Note the decay state of the anterior upper teeth.

Fig. 7. Résection osseuse symphysaire au large de la tumeur. Noter l'atteinte carieuse des dents antéro-supérieures.

is shown by the importance of scale, the bad dental status, the existence of former cellulitis fistula scar and the age of

Fig. 8. Surgical piece: tumoral right hemimandible.

Fig. 8. Pièce opératoire : hémimandibule droite tumorale.

the tumor. Giant ameloblastoma associated with poor oral hygiene is related in the literature [6].

The lesion begins most frequently in the molar region of the mandible by indurated painless swelling. This is the most common location and it gradually extends over the entire mandible. The development is slow and asymptomatic, explaining the negligence. The tumor gnaws all the bone and blows the cortical and swelling signs begin causing lesser or greater facial deformation depending on evolution stage of the lesion. The surrounding soft tissues are not infiltrated but they are distended and sensory disturbances of the inferior alveolar nerve can occur [7-9].

Macroscopically, its nature may be cystic, solid or mixed. Microscopically, the lesion is characterized by proliferation of epithelial cells arranged on a connective tissue stroma with a structure similar to the enamel organ at different stages of differentiation [10]. There are many types of ameloblastomas: follicular, plexiform, desmoplastic, granular... The histological appearance of our case is the plexiform type: islands of odontogenic epithelium are arranged as a tangled network of anastomosing strands, unlike the follicular type (Fig. 6) [13].

On the X-ray, ameloblastoma may be confused with odontogenic keratocyst, aneurysmal bone cyst, fibrosarcoma or giant cell tumor [10]. It shows uni- or multilocular radiolucency, more or less rounded with sharp delineation, giving characteristic soap-bubbles shape [7]. Dental root resorption is often associated, and it is more ameloblastomas than in other odontogenic cyst lesions [11, 12]. Tomodensitometry has particular interest in surgery (Figs. 4 and 5).

Fig. 9. Root resorption on 43, 44 and 45. Note the presence of decay on 41, 42 and 44.

Fig. 9. Rhizalyse intéressant 43, 44 et 45. Noter la présence de carie sur 41, 42 et 44.

The treatment of ameloblastomas is essentially surgical and recurrence is very common. The recurrence is higher in the solid or multicyst forms, especially in case of simple enucleation or curettage. Bone resection is the best treatment by taking a large margin [14, 15] (Figs. 7 and 8). Dental root resorptions are visible when the teeth are extracted from the surgical piece (Fig. 9). In developing countries, to do mutilating surgery such as segmental mandibulectomy is common due to negligence. Socio-economic constraints of the patient do not allow graft reconstruction. The efficiency of radiotherapy is uncertain, but it may be indicated in inoperable cases, primarily in the posterior maxilla or when recurrence is frequent [16, 17].

The high propensity for recurrence requires a long-term post-operative follow-up since the recurrence may appear very late [18]. In developing countries, this control is often not followed-up because of socio-economic problems.

Malignant transformation of odontogenic tumor is extremely rare. Officially, the WHO does not yet recognize the existence of odontogenic cancer [19]. However, a Nigerian study mentions twenty cases of ameloblastic carcinoma in 28 years [20]. Metastasis are very rare [21], and when they exist, they generally develop in the lungs [22].

In conclusion, ameloblastoma is a locally invasive tumor of odontogenic origin, located mainly on mandible, with propensity for local recurrence. Negligence in its management leads to a huge volume whose treatment is very mutilated and expensive.

Competing interests: none

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