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CASE REPORT

Odontogenic myxoma. Clinical case presentation

Mixoma odontogénico. Presentación de caso clínico

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ABSTRACT

Odontogenic myxoma are intraosseous lesions originating in embrionic connective tissue. They are benign lesions but locally aggressive and moderately recurrent. Objective: Surgical tumor resection with immediate primary reconstruction to return function and aesthetics. Case report: A 29 year old female patient attending ambulatory practice of the Maxillofacial Surgery Clinic for assessment and treatment. She presented volume increase and asymmetry in the right hemifacial region due to a tumor of approximately 5 year evolution, previously surgically treated to excise the lesion. Histological study reported piogenic granuloma. After surgery the patient experienced a slow and progressive recurrence of the tumor, disabling the patient in mastication, deglutition and breathing functions, and causing facial disfigurement. Treatment: Incisional biopsy was performed. It reported odontogenic myxoma. In the operating theatre, with the patient under general anaesthesia, it is decided to perform the tumor resection with immediate primary reconstruction of the orbital region with an autogenous graft of the iliac crest to later manufacture a palatine obturator.

Key words: Odontogenic myxoma, autogenous (autologous)graft.

RESUMEN

Es una lesión intraósea derivada del tejido conectivo embrionario asociada a la odontogénesis, de tipo benigno pero localmente agresiva y de moderada recurrencia. **Objetivo:** Resección quirúrgica tumoral con reconstrucción primaria inmediata con la finalidad de devolver funcionalidad y estética. **Presentación del caso:** Paciente femenino de 29 años de edad, que se presenta en la consulta externa del servicio de Cirugía Maxilofacial para valoración y tratamiento. Presenta aumento de volumen y asimetría en región hemifacial derecha por tumoración con evolución aproximada de 5 años, previamente tratada de manera quirúrgica para excisión de lesión. El estudio histopatológico reporta granuloma piógeno; posteriormente inicia con aumento de volumen lento y progresivo de masa tumoral recurrente, discapacitando a la paciente en su masticación, deglución y respiración ocasionándole desfiguramiento facial. **Tratamiento:** Se toma biopsia incisional la cual reporta mixoma odontogénico, se decide realizar en quirófano bajo anestesia general la resección tumoral con reconstrucción primaria inmediata de región orbitaria con injerto autólogo de cresta iliaca y posteriormente elaboración de obturador palatino.

Palabras clave: Mixoma odontogénico, injerto autólogo.

INTRODUCTION

There is great controversy about the origin of myxomatous tumors. In 1863 Virchow introduced the term myxoma to describe a group of tumors of similar histology to that of the umbilical cord. In 1948 Stout redefined the myxoma histological criterium as a true neoplasia, which does not produce metastasis and which excluded cellular components of other mesenchymal tissues such as chondroblasts, lipoblasts and rabdiomyoblasts. Myxoma is a tumour that can be found in the heart, skin, subcutaneous cellular tissue and bone. Nevertheless, head and neck myxoma is a rare tumor.[1-5]

Two kinds of myxoma have been identified:[1] myxoma derived from craniofacial bone tissue of facial bones which was previously subdivided into true osteogenic myxoma and odontogenic myxoma, and[2] the one derived from the soft tissue of the perioral region, parotid glands ear and larynx. Moshiri & al recently proposed an ultrastructural immunohistochemical study. In it, the odontogenic origin concept is reinforced, suggesting that fibroblasts that originate den-

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tal buds experience changes from which emerge the growth of the odontogenic myxoma.

This tumor also has an extremely rare, malignant version called odontogenic myxosarchoma.[6-9]

In contrast to this theory, Slootweg and Wittkampf showed that the maxillary myxoma matrix is totally different from the extracellular matrix of normal dental tissue. Therefore, myxoma can develop in the sinonasal tract and other facial bones originating from non odontogenic mesenchymatous tissue.[10]

The presence of odontogenic epithelium is not necessary to emit a bone myxoma diagnosis. In contrast with the aforementioned, McClure and Dahlin reviewed over 6,000 bone tumors at Mayo Clinic (Rochester, New York) and concluded that the presence of true myxomas could not be ascertained, excepting for those found in upper and lower jaws. Although no specific etiological factors have been demonstrated, upper and lower jaw myxoma is commonly accepted as an odontogenic tumor and represents from 3 to 6% of all odontogenic tumors.[11-13]

DEFINITION

Odontogenic myxoma are rare, benign intraosseous lesions, arising from embryonic connective tissue, associated to odontogenesis of dental bud mesenchymatous origin, either from dental papilla, periodontal ligament or follicle. They are locally aggressive. They generally appear in the maxillary and mandibular region, and affect patients in their first to fifth decade of life. Mean age is 30 years, without sex predilection. Two third of cases are in the mandible and the remaining third in the maxilla.

CLINICAL CHARACTERISTICS

Odontogenic myxoma can be found in any region of the upper jaw, and constantly involve the maxillary sinus, they cross the midline and affect the nasal region and the contralateral sinus. Lower jaw lesions are generally found in the molar and premolar area, and they extend up to the ascending ramus and the condylar region. These are painless, slow growing lesions which cause root dilacerations and even in some cases root resorption. They are generally associated to retained or missing teeth, with multilocular aspect, cause cortical expansion and eventual perforation, and result in tumefaction and facial deformity. Patients are generally aware of the lesions years before they seek treatment, therefore lesions reach substantial size due to lack of timely treatment.[14,15]

IMAGING FEATURES

Lesions of great size have a characteristic imaging aspect, constituted by a radiolucent multilocular zone with appearance of «soap bubbles» or «beehive». Nevertheless, sometimes they can have a radiopaque appearance, especially those associated to the maxillary sinus. In some area, thick or angular trabeculations can be found, and there is no clear delimitation with healthy bone. Displacement and non vital teeth caused by the tumor mass are relatively common findings. Root resorption is less frequent. Lesions are small and unilocular, and with a non specific radiolucent appearance. They can be confused with ameloblastoma, giant cell central granuloma and hemangioma.[16]

HISTOPATHOLOGY

This lesion is constituted by non differentiated fusiform mesenchymal cells widely dispersed in a non fibrillar mucoid fundamental substance *(Figure 1)*. It can present localized collagen areas as well as external hyalinization of blood vessels. When the lesion contains large amounts of collagen it is named myxofibroma. In peripheral areas, myxomatous tissue penetrates into trabecular spaces producing islets of residual bone; this characteristic explains the difficulty of conservatively removing the lesion. Moreover, two acid mucopolysaccharides i.e. hyaluronic acid and chondroitin sulphate have been observed.[17,18]

DIFFERENTIAL DIAGNOSIS

The calcifying epithelial odontogenic tumor (CEOT) must be considered in its translucent variety.

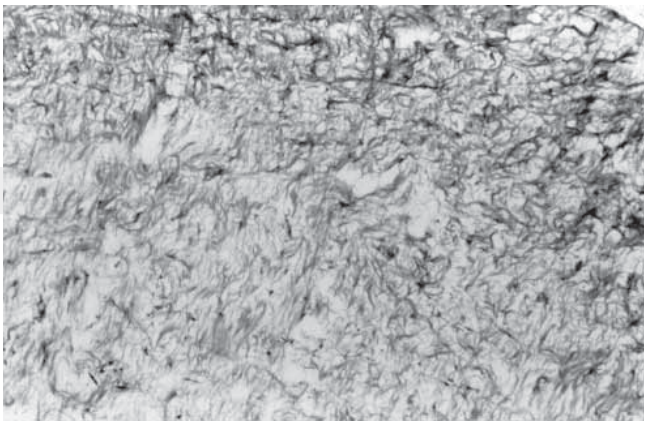


Figure 1. Microscopic view of odontogenic myxoma histological cut.

The dentigerous cyst, ameloblastoma, odontogenic keratocyst giant cell central granuloma and ossifying fibroma and a myxoid type of hyperplastic follicle must be considered.

TREATMENT AND PROGNOSIS

Since the lesions are mucoid or gelatinous in consistency, proper curetting is generally hindered. Treatment, therefore, will consist of broad and complete surgical removal at the moment when the tumor has invaded deep planes. If this is not carried out, there is a probability of recurrence. Recurrence has been reported in 25% of improperly treated cases. To avoid recurrence, it is recommended to perform a block resection with free bone margins of at least 2 mm. This lesion is not sensitive to radiotherapy. The combination of enucleation and cryotherapy based on liquid nitrogen for the treatment of large lesions has the advantage of preserving residual bone and is not opposed to the idea of a bone graft immediately after the cryotherapy procedure.[19]

CASE REPORT

29 year old female, born and residing in the State of Guanajuato, housewife, with six years of schooling. The patient denied blood transfusions and presence of any allergy. The patient attends the Outpatient Clinic at the Maxilofacial and Oral Surgery Service of the Juarez Hospital, Mexico City requesting evaluation and treatment of a tumoral mass in the right half of the face. The patient experienced facial deformity and difficulty to perform breathing, mastication and swallowing functions.

The patient informs that the lesion initiated 5 years previously, while experiencing a normal pregnancy of 36 weeks. She reports mobility of the partially erupted right upper molar and presence of pain and volume increase in that region. She also found in that region a solid mass of approximately 1 x 0.5 cm. The patient informed that, after the pregnancy came to term, the lesion was surgically removed, as well as the affected tooth, without any complications. The histopathological study revealed pyogenic granuloma. The patient was asymptomatic for four years, after which she experiences a new slow and progressive volume increase in the previously treated region. The patient received only medication, but no positive result was obtained. After this, the patient is informed that she can no longer be treated at that hospital, since there is a possibility that the lesion is malignant. The patient neglects herself. The lesion continued its growth process. The patients relatives, upon seeing the conspicuous facial asymmetry, move to Mexico City to secure a second medical opinion.

Physical examination revealed a facial deformity due to the tumor. Ocular globe with vision and movement preserved with severe right ocular globe proptosis, no nose deviation with deformity of the right nasal ala and right nasal vestibule obstructed by the tumor mass. Septum with posterior deviation to the left, with the presence of fetid hyaline rhinorrhea, non visible turbinate, diminished breathing flow through right nostril. An increase of volume is observed in the middle and lower thirds of the affected right hemifacial, due to the presence of a rounded solid mass. There were no further changes and no neurological signs *(Figure 2 A)*.

Intraorally, the permanent dentition is present with some extractions. A lesion is observed in the right maxilla. The lesion presents neoplastic appearance and fills a great proportion of the oral cavity. This lesion exceeds the midline and covers up to the posterior border of the palatine vault. There is a displacement of upper teeth in the lesion; this precludes occlusion, and there are impressions of the lower teeth surfaces on the tumor. The lesion is of an indurated consistency, with infiltrated base, irregular shape, lobed with scattered, reddish-purplish focal areas *(Figure 2 B)*. The patient experienced severe halitosis due to defective oral hygiene. The floor of the mouth, the tongue and the rest of the mucosa are normal.

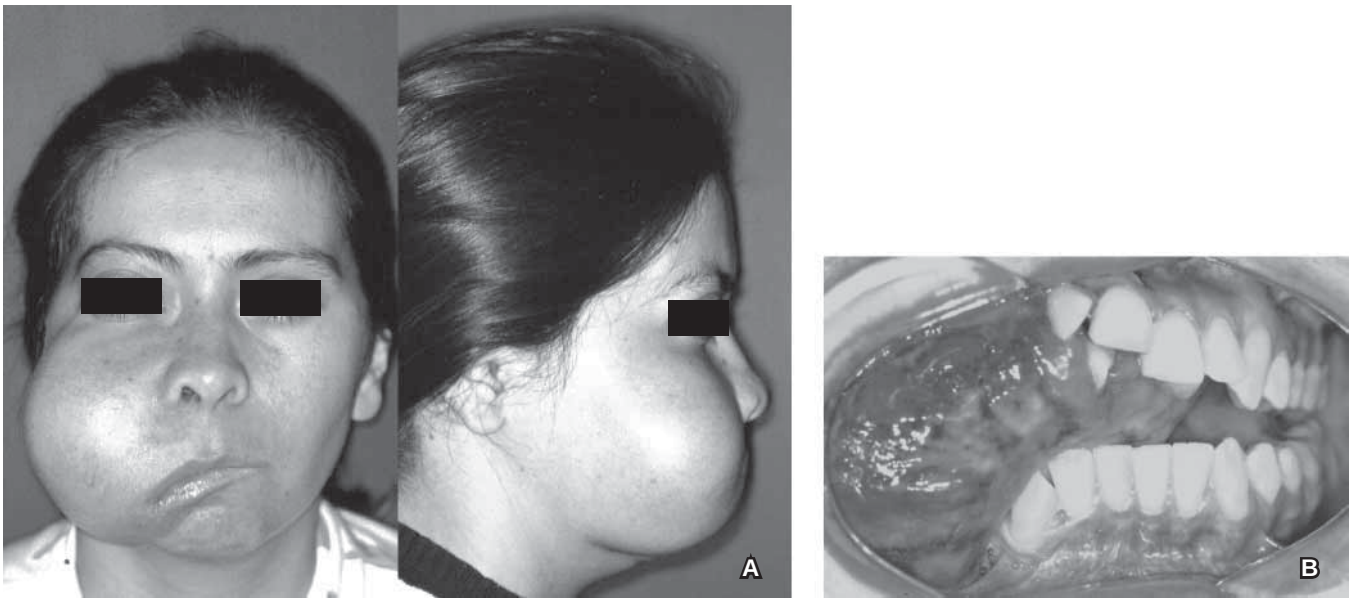
IMAGING FEATURES

The X ray examination: (Watters PA lateral and orthopantomography) destruction of the right hemimaxilar is observed. There is displacement of the upper teeth, destruction of the lateral nasal wall and right orbital floor, moderate deviation of the nasal septum to the left side. The computer axial tomography (CT scan) shows displacement of the right ocular globe. Same bone structures destroyed, occupancy of ethmoid cells and ipsilateral sphenoid sinus *(Figures 3B and C)*.

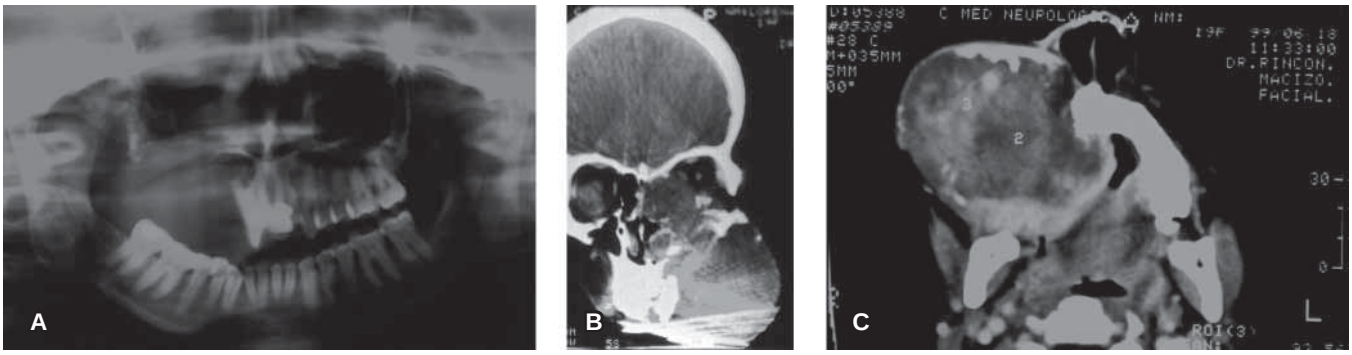
TREATMENT

Treatment was divided into three phases:

1. Incisional biopsy
2. Tumor resection with immediate orbital reconstruction with an iliac crest bone graft.
3. Prosthetic rehabilitation with an obturator.



Figures 2. A. Extraoral view, **B.** Tumor intraoral view.



Figures 3. A. Ortopantomography. **B** and **C**. CAT axial and coronary cuts.

PREOPERATIVE PHASE

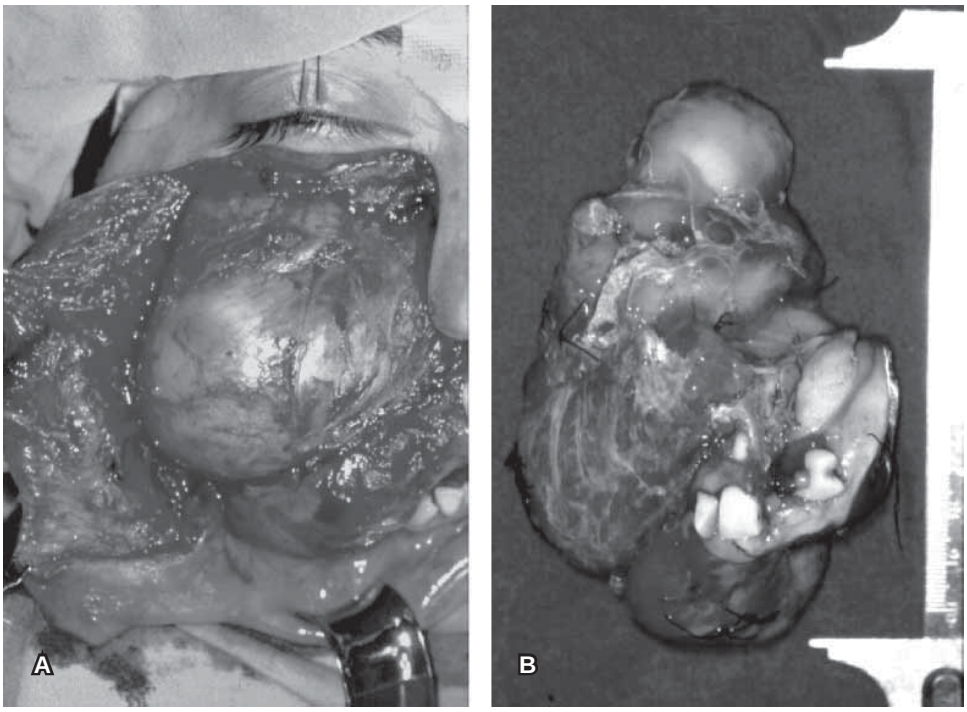
An incisional biopsy is taken which reports a pathological report of odontogenic myxoma. Presurgical protocol was performed with interconsultation with ophthalmologist and ENT specialist.

TRANSOPERATIVE PHASE

With the patient under general anaesthesia and nasotracheal intubation the tumor area as well as the hip area were prepared and infiltrated with local anaesthesia and epinephrine.

A Webber-Ferguson-Lynch type surgical incision is performed on the right facial region dissecting a full thickness flap to expose the tumor *(Figure 4A)*. The tumor is dissected. The lesion appears to be well encapsulated. After this, the lesion is dissected in cephalic direction and no severe compromise of the ocular globe is found. Nevertheless the lesion occupies an important section of the orbital cavity, which causes proptosis of the ocular globe and complete destruction of the floor and lower orbital ridge.

After this procedure, an osteotomy of the right malar region is performed, over the tumor, leaving free margins of 5 mm to ease tumor resection *(Figure 4 B)*. The bone graft is contoured to give a shape similar to that of the floor region and right orbital ridge. The slab is adjusted from the malar bone to the nasal lateral region. This procedure restores shape to the region



Figures 4 A. Weber-Ferguson-Lynch surgical approach. **B.** Resected tumor.

and achieves ocular globe support. The contoured graft is fixed with 2 mm titanium miniplates and 5 x 2 mm screws. Hemostasis is assessed, and the surgical wound is closed, having previously rotated the buccinator flap to protect the graft *(Figure 5)*.

The flap is then repositioned and the muscular plane is sutured with Polyglactine 910 3-0® with quadrangular mattress sutures. The dermal plane is sutured with subcuticular suture with Nylon 5-0. In the oral region, simple stitches are placed with Polyglactine 910 3-0. Gauze pads with iodoform are placed into the palatine cavity, to be changed on alternate days. A NG tube is placed for feeding purposes. The surgical event is completed entubating the patient, who shows no complications and is moved to the recovery room with stable vital signs and respiratory self-sufficiency.

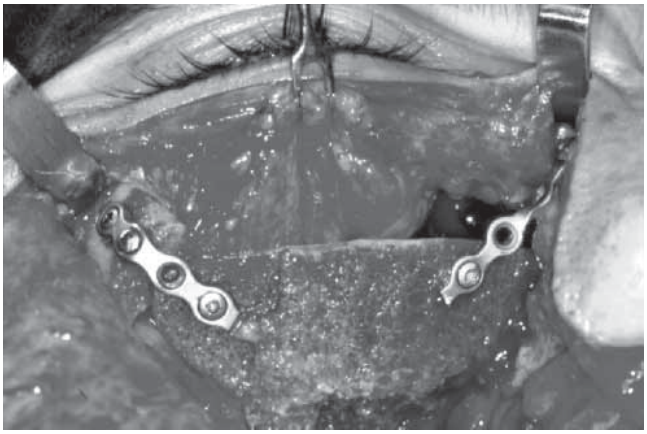


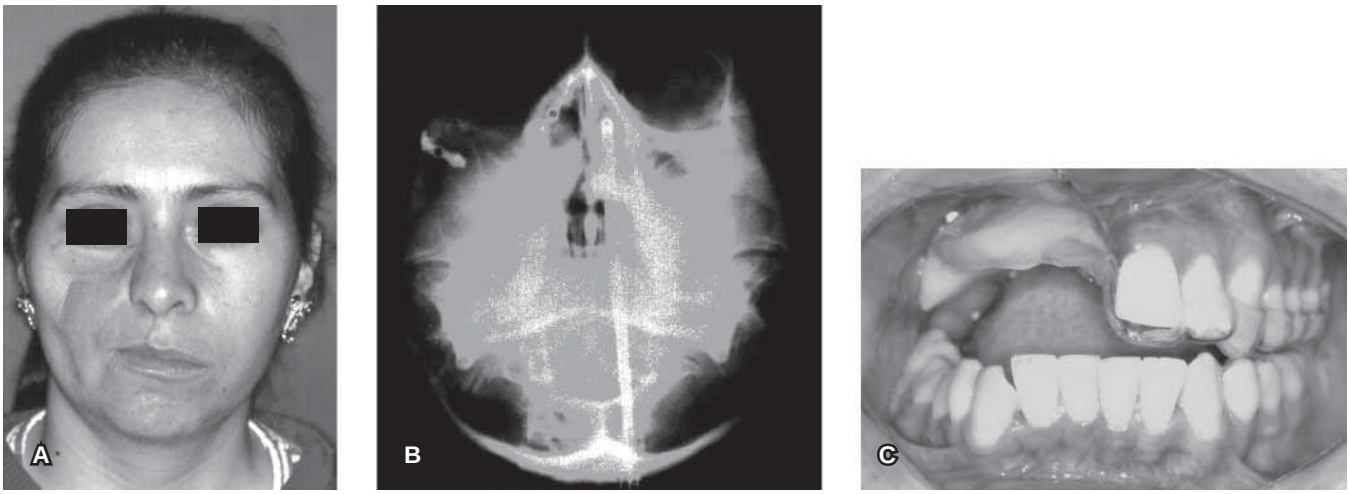
Figure 5. Contoured autologous graft fixed with miniplates and titanium screws.

POSTOPERATIVE PHASE

The NG tube is removed 8 days later, once initiated the oral feeding: the patient properly tolerated the procedure.

Seven days after the operation, the patient is assessed (evaluated). Moderate oedema of the facial region is observed. The facial region presents moderate asymmetry and right nasogenial depression. The surgical wound does not show clinical infection or dehiscence. The sutures are removed. The right ocular globe is assessed and shows preserved vision and movement, interpupillary axis showing no deviations, permeable nasal region which improves breathing air flow, and preserved facial mobility. The patient is sent to the maxillofacial prostheses department for the manufacturing of a palatine obturator, which is placed one week later.

Six weeks after the operation, the patient is examined. Appropriate movement of the facial mus-



Figures 6. A. Extraoral view, **B.** Postoperative radiograph, **C.** Intraoral view with palatine obturator.

cles is observed finding no evidence of paresis. The patient informs of dysesthesia in the right lateral nasal and maxilar malar region. A moderate maxillo malar depression is observed. Scarring of the surgical wound seems appropriate and alteration free *(Figure 6)*. Intraorally the palatine obturator can be observed in the right place and accomplishing its function. The velopharyngeal function is preserved.

DISCUSSION

Treatment of bone myxoma is subject of controversy. Some authors have informed of the use or radiotherapy, nevertheless, this lesion is not radiosensitive and studies reveal that presently there is no long term cure with this treatment. Surgical treatment of bone myxoma includes enucleation, curettage and chemical cauterization as well as partial or block resection and lesion removal with free bone margins. Davis et al informed of a 25% recurrence rate, which varied according to the aggressiveness of the surgical technique used to excise the tumor. It was also reported that the recurrence rate generally occurs two years after the excision.[20]

Deron et al, Abubaker and Benson recommend block radical primary resection of the tumor, with a 1 cm lesion free margin, preserving thus adjacent anatomical structures. Although myxoma is a non encapsulatead and infiltrating tumor, follow up of the case must continue indefinitely. Slootwegs in his report. mentions tumor recurrence in the lower jaw 15 years after its excision.[21-23]

CONCLUSIONS

Treatment of any tumor must, first of all, include a biopsy and histopathological study. These must be properly performed so as to arrive to an accurate diagnosis, and decide upon surgical treatment. To attain this goal, the oral pathologist, radiologist and maxillofacial surgeon must work as a team.

Presently, block resection is still the best treatment for odontogenic myxoma in large tumors. Small lesions are better treated considering first the use of curettage and enucleation before performing large and devastating operations which could cause disfigurement and loss of function as well as interfere in the growth process of pediatric patients.

In this case, it was indispensable to perform an immediate primary reconstruction so as to preserve function. Nevertheless a strict imaging survey is required, which must include simple X rays as well as CT scans to monitor probable recurrence of the tumors.

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