ABSTRACT

Desmoid tumor is a fibroblastic proliferation of aponeurotic muscle tissue, fascia or periosteum of unknown etiology. It generally appears as a single tumor and can be related to Gardner’s Syndrome. It is considered a rare lesion, representing less than 0.03% of all tumors, with an annual incidence of 2-4 cases per 100,000 habitants. It usually appears as a mass (body) with symptomatology associated to location. It has the potential of achieving local invasion without progressing to metastasis. These tumors experience high rates of local recurrence after surgery, even when wide margins have been respected. A case of a 12 year old male patient is presented. He attended the Maxillofacial Surgery Service of the Hospital Regional Adolfo Lopez Mateos due to a volume increase of the left ascending mandibular ramus.

Key words: Desmoid tumor, extraabdominal, mandible.

INTRODUCTION

There is an ill-defined group of fibroblastic cells hyperplasia called fibromatosis. These can vary from a postinflammatory keloid scar up to non neoplastic fibrosis. Also included are lesions which can be considered as mid-way between fibromas and fibrosarcomas and are known as aggressive fibromatosis (desmoid tumors).1

Desmoid tumor is a fibroblastic proliferation of musculoaponeurotic tissue, fascia or periosteum. It consists of a painless, non encapsulated, ill-defined single tumor mass. It shows a firm consistency upon palpation, presents a grayish hue, is of slow and progressive growth, benign, locally invasive, and with a tendency to recurrence. It equally presents the ability to encapsulate adjacent neurovascular structures.2 It can present an unpredictable evolution, and when the lesion is located in the head or neck its proximity to vital and complex structures can complicate its evolution or treatment.

RESUMEN

El tumor desmoide es una proliferación fibroblástica de tejido musculoaponeurótico, fascia o periostio, de origen desconocido. Generalmente aparece como un tumor solitario y puede estar en relación con el síndrome de Gardner. Es una lesión rara, que representa menos del 0.03% de todos los tumores, con una incidencia anual de 2-4 casos/100,000 habitantes. Se suele manifestar como una masa cuya sintomatología dependerá de la localización. Tiene capacidad de invasión local sin ocasionar metástasis a distancia y altas tasas de recurrencia local tras la cirugía, incluso con márgenes amplios. Se presenta el caso de un paciente masculino de 12 años, quien acude al Servicio de Cirugía Maxilofacial, del Hospital Regional Lic. Adolfo López Mateos por presentar aumento de volumen en rama ascendente mandibular del lado izquierdo.

Key words: Tumor desmoide, extraabdominal, mandíbula.

INCIDENCE AND PREVALENCE

It is a rare lesion representing less than 0.03% of all tumors, with yearly incidence of 2-4 cases per 100,000 habitants.
Desmoid tumors associated with hereditary adenomatous polyposis are 1,000 times more frequent than those found in general population. There is a female: male predominance of 2:1. Prevalence of desmoid tumor in this condition is 7-12%. In these patients, abdominal location of the tumor is most frequent. 42% of these tumors are located within the abdomen and the remaining 40% are found in the abdominal wall.7

Figure 1. Patient front and chin photograph.

Figure 2. Orthopantomography.

Figure 3. Computer tomography, sagittal cut, size of lesion can be observed.

Figure 4. 3D reconstruction. Lysis of bone tissues can be observed in two thirds of the mandibular ramus.
CLINICAL OBSERVATIONS

These tumors appear as a mass whose symptomatology will depend on its location. Usually they present a slow growth pattern, and can reach considerable size. In 10% of cases, a rapid growth pattern can be found, spontaneous regression cases have also been described.8

DIAGNOSIS

Diagnosis is established based on findings obtained from clinical observation, as well as radiographic and histopathological studies.9

CLINICAL CASE

12 year old male patient attended the Maxillofacial Surgery Service of the Regional Hospital «Lic. Adolfo

Figure 5. 3D reconstruction of soft tissues.

Figure 6. Angiograph.

Figure 7. Histological cuts (slides).
Figure 8. Tomography with 3D reconstruction.

Figure 9. Stereolithograph where mandibular reconstruction plate can be observed.
López Mateos due to a one month evolution mass increase in the left mandibular ascending ramus. The lesion presented the following characteristics: limits the oral opening, is painful, indurated, of approximately 3 mm in diameter, non displaceable, non erythematous, non hyperemic or hyperthermic (Figure 1).

In orthopantomography evaluation, a radiolucid zone is observed in the left ramus and mandibular body, observing a third molar in proximity of the lesion region (Figure 2). A 3D reconstruction achieved through computerized tomography was requested. In it, a hypodense zone in the left mandibular ascending ramus zone was observed, as well as bone lysis and a tumor mass of approximately 6 to 8 cm diameter extending into the infratemporal fossa (Figure 3-5).

Figure 10. Surgical approach and hemimandibulectomy.

Figure 11. Stereolithograph where mandibular reconstruction plate is adapted.

Figure 12. Placement of reconstruction plate.
An angiography was performed to preclude vascular compromise (Figure 6).

Under localized anesthesia, incisional biopsy was performed and the sample was sent to the pathology department.

Anatomical and pathological study revealed a desmoid tumor located on the outside of the abdomen (Figure 7). Surgery was performed in the following fashion: temporal, preauricular retromandibular approach with submandibular extension to uncover left ramus and mandibular body; Tumor mass was removed from soft tissues, a hemimandibulectomy was performed with the placement of a mandibular reconstruction plate (Figures 8-13).

Based on the anatomical and pathological study the diagnosis of desmoid tumor was confirmed. The patient presently shows favorable evolution, facial paralysis in remission, no recurrence data, and is under strict surveillance (Figure 14).

**DISCUSSION**

Enzinger and Weiss\(^\text{10}\) differentiate between superficial and deep desmoid tumors. Included within the superficial tumor category are the following: palmar (palm), plantar (sole) and genital tumors.

Deep fibroid tumors include the following subtypes: abdominal, intraabdominal and extraabdominal (inside and outside of the abdomen). According to this classification, desmoid tumors of the head and neck belong to the deep fibroid group and extraabdominal type.

WHO describes it as a benign tumor, characterized by the presence of abundant collagen fibers formed by tumor cells. The tumor is cell-poor and nuclei are of ovoid or elongated shape. They present no cellularity.

In scientific literature there are very few reported cases of head and neck desmoid tumors. Incidence can be ascertained at 9.5 to 33%, out of which 85% are found in the neck. Other less frequent sites have also been reported like face, oral cavity, scalp, paranasal sinus and orbit.\(^{11-14}\) They can appear at any age rank, although their appearance is most frequent in patients between their 3rd and 4th decade of life.

There are no characteristic radiographic signs. In general terms, it is a translucent lesion which expands the cortical and thins it. The lesion is surrounded by a thin margin of reactive bone. When scintigraphically observed (gammagnostically), a radiotracer uptake increase can be observed in the inside of the lesion.

Radiation can be prescribed only in cases of inoperable tumors, or cases of residual disease after marginal surgery has been performed.\(^{15}\)

As final observation, it can be noted that the following have been described as effective: Progesterone, AINE, warfarine. Vitamins C and K, tamoxifen, testosterone, and some antineoplastic agents like adriamycin dacarbazine, vincristine etc.\(^{2}\) The use of antiestrogen and anti-inflammatory agents has also been recommended as treatment for tumors not suitable for resection.\(^{16,17}\)

**REFERENCES**


SUPPORTING REFERENCES


Correspondence address:
Dr. Doroteo Vargas López
Hospital Regional «Lic Adolfo López Mateos»
Av. Universidad 1321, Mexico City.
E-mail: dovarlo@yahoo.com.mx