



Mandibular desmoid tumor. Case report

Tumor desmoide mandibular.

Reporte de un caso

Doroteo Vargas López,* Ronald Rojas Villegas,[§] Víctor García Alcaraz,* Sara Guadalupe García Amador^{||}

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.

Palabras Clave: Tumor desmoide, extraabdominal, mandíbula.

RESUMEN

El tumor desmoide es una proliferación fibroblástica de tejido musculoponeurótico, 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.

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.

It generally appears as a single tumor and can be related to Gardner's Syndrome.^{2,3}

Its etiology is not well known. Some studies suggest the possibility of some genetic defect. Other correlations indicate the possible role of trauma⁶ and estrogen stimulation as causes of these lesions.^{1,4-6}

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

* Physician attached to the Maxillofacial Surgery Service of the «Adolfo López Mateos» Hospital ISSSTE. Mexico City, Mexico.

[§] Maxillofacial Surgeon, private practice.

^{||} Former resident attached to the Maxillofacial Surgery Service of the Regional Hospital «Adolfo López Mateos» ISSSTE. Mexico City, Mexico.



Figure 1. Patient front and chin photograph.

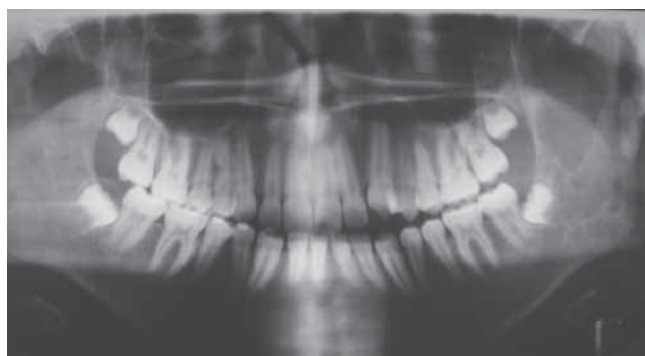


Figure 2. Orthopantomography.

people. They represent 0.03% of benign tumors and 0.06% of all bone tumors.

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

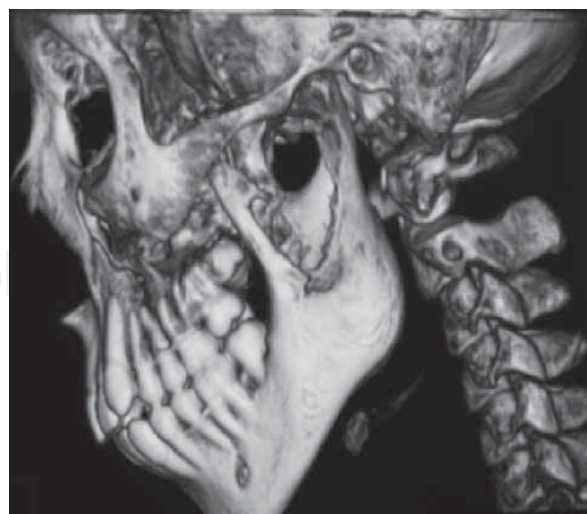


Figure 4. 3D reconstruction. Lysis of bone tissues can be observed in two thirds of the mandibular ramus.

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

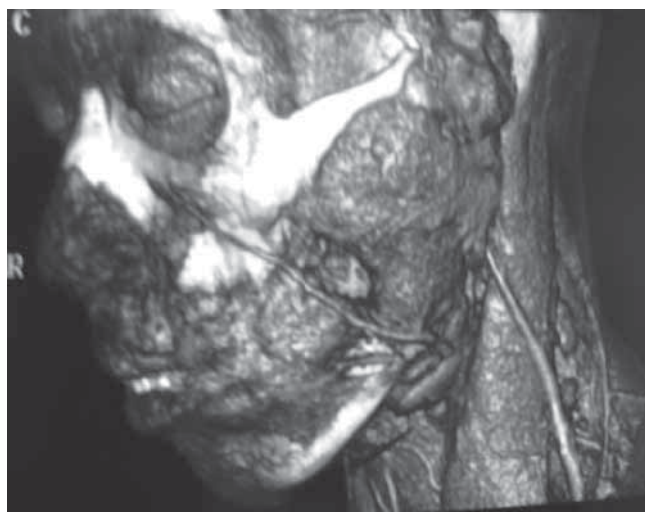


Figure 5. 3D reconstruction of soft tissues.

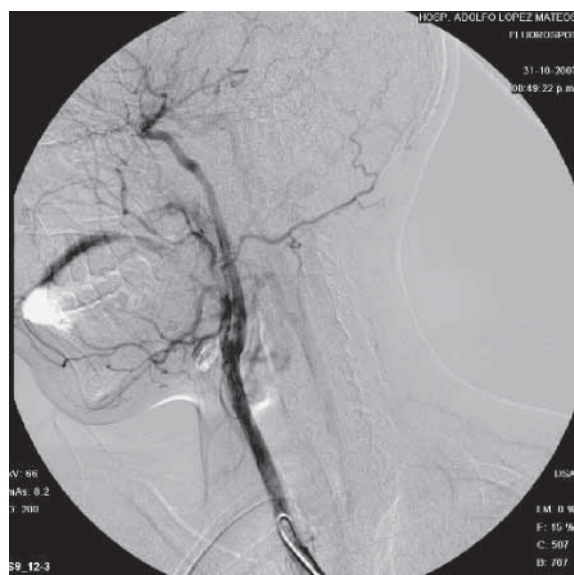


Figure 6. Angiograph.

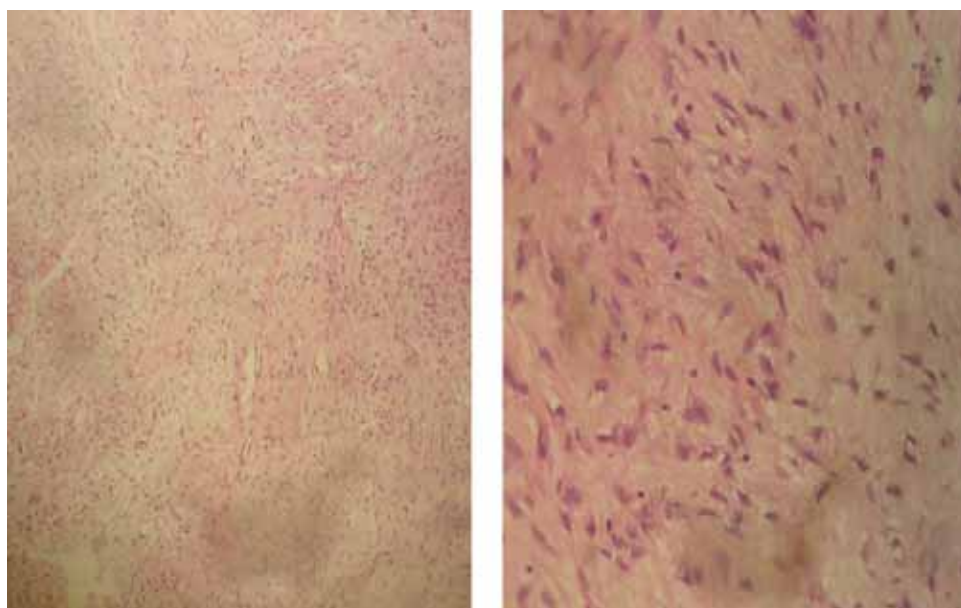


Figure 7. Histological cuts (slides).

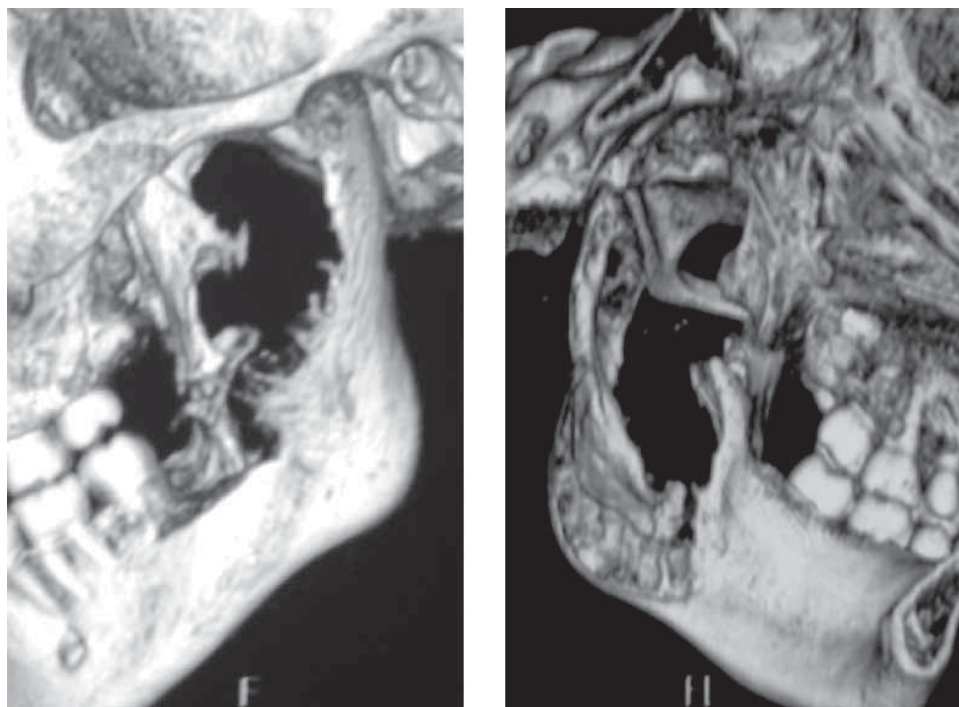


Figure 8. Tomography with 3D reconstruction.

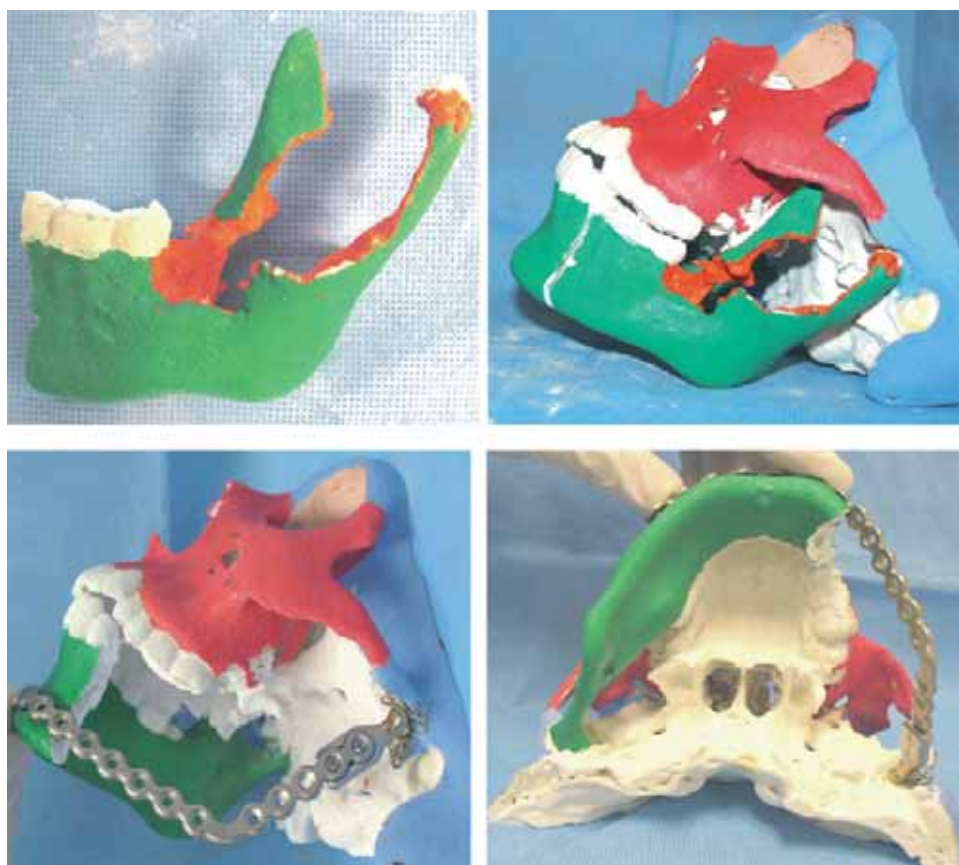


Figure 9. Stereolithograph where mandibular reconstruction plate can be observed.

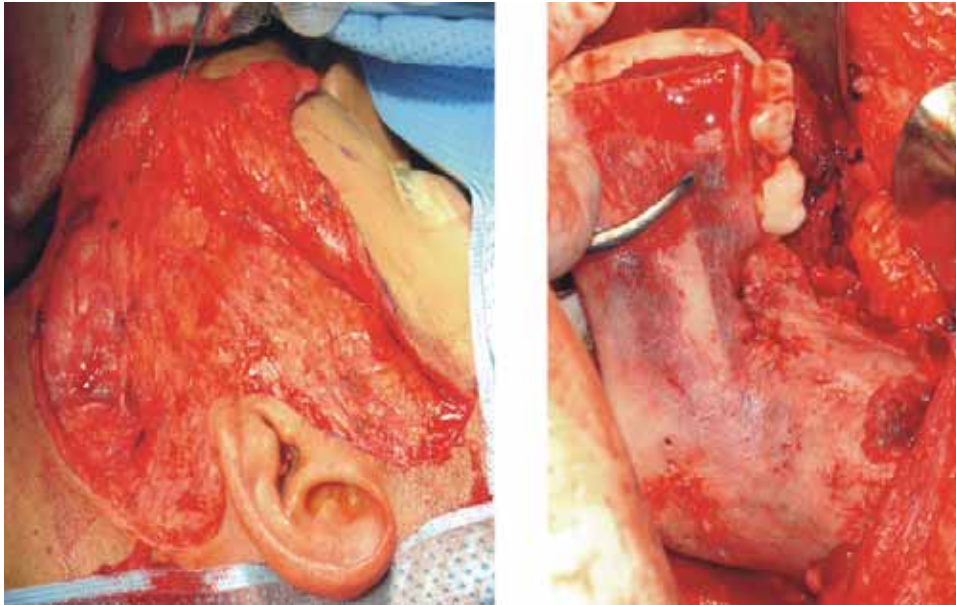


Figure 10. Surgical approach and hemimandibulectomy.

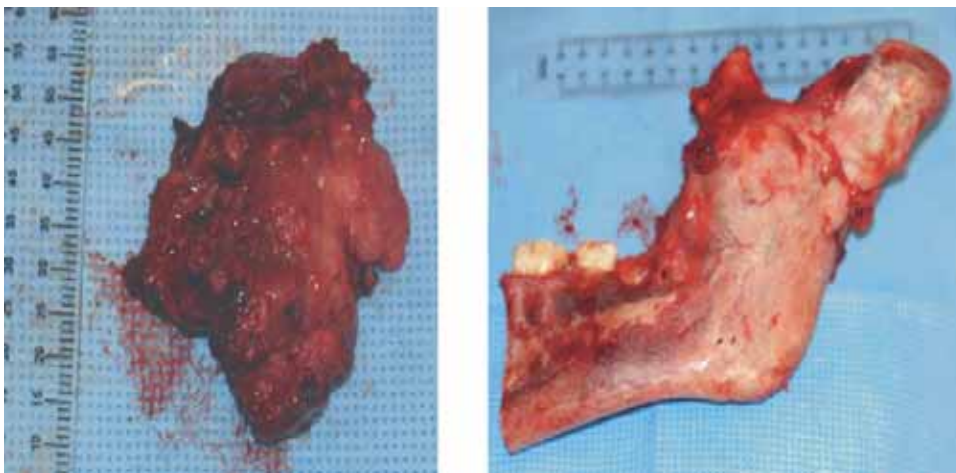


Figure 11. Stereolithograph where mandibular reconstruction plate is adapted.



Figure 12. Placement of reconstruction plate.

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 hyperhemic 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 13. suture by planes.

An angiography was performed to preclude vascular compromise (*Figure 6*).

Under localized anaesthesia, 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 Weiss¹⁰ 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



Figure 14. Control orthopantomography.

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 (gammagraphically), 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 anti-estrogen and anti-inflammatory agents has also been recommended as treatment for tumors not suitable for resection.^{16,17}

REFERENCES

1. Cotrán RS, Kumar V, Collins TR. *Patología estructural y funcional*. 6ª edición. Ed. McGraw Hill Interamericana: España, 2000: 1308-9.
2. Siegel NS, Bradford CR. Fibromatosis of the head and neck: a challenging lesion. *Otolaryngol Head Neck Surg* 2000; 123: 269-75.

3. Allen E. *Tratado de cirugía ortopédica*. Tomo I. Ed. Panamericana: México, 1996.
4. Tan YY, Low CK, Chong PY. A case report on aggressive fibromatosis with bone involvement. *Singapore Med J* 1999; 40 (2): 111-2.
5. Abrams GD. Disturbances of growth, cellular proliferation and differentiation. In: Price SA, Wilson LM. *Pathophysiology. Clinical concepts of disease processes*. 4th edition. International: Mosby, 1992: 94-5.
6. Tejpar S, Nollet F, Li C, Wonder JS, Michils G, dal Cin P. Prevalence of beta-catenin mutations and beta-catenin dysregulation in sporadic aggressive fibromatosis (desmoid tumour). *Oncogene* 1999; 18 (47):.
7. Bertario L, Russo A, Sala P, Eboli M. Genotype and phenotype factors as determinants of desmoid tumors in patients with familial adenomatous polyposis. *Int J Cancer* 2001; 95: (102) 6615-20.
8. Peterschulte G, Lickfeld T, Moslein G. The desmoid problem. *Chirurg* 2000; 71: 89.
9. Tostevin PMJ, Wyatt M, Hosni A. Six cases of fibromatosis of the head and neck in children. *Int J Pediatr Otorhinolaryngol* 2000; 53: 235-244.
10. Enzinger FM, Weiss SW. *Soft tissue tumors*. 3rd ed. St Louis: Mosby; 1995.
11. Masson JK, Soule EH. Desmoid tumors of the head and neck. *Am J Surg* 1966; 112: 615-22.
12. Conley J, Healy WV, Stout AP. Fibromatosis of the head and neck. *Am J Surg* 1996; 112: 609-14.
13. Gnepp DR, Henley J, Weiss S et al. Desmoid fibromatosis of the sinus tract and nasopharynx. A clinicopathologic study of 25 cases. *Cancer* 1996; 78: 2572-9.
14. Rao BN, Horowitz ME, Parham DM et al. Challenges in the treatment of childhood fibromatosis. *Arch Surg* 1987; 122: 1296-8.
15. Lackner H, Urban C, Kerbl R, Schwinger W, Beham A. Noncytotoxic drug therapy in children with unresectable desmoid tumors. *Cancer* 1997; 80 (2): 334-4.
16. Ballo MT, Zagars GK, Pollack RA. Desmoid tumor: Prognostic factors an outcome after surgery radiation therapy, or combined surgery and radiation therapy. *J Clin Oncol* 1999; (17): 158-167.
17. Leithner A, Schnack B, Katterschatka T, Wiltshcke C, Amann G, Windhager R et al. Treatment of extra-abdominal desmoid tumor with interferon-alpha with or without tretinoin. *J Surg Oncol* 2000; 73 (1): 21-5.

SUPPORTING REFERENCES

18. Batsakis JG, Raslan W. Pathology consultation extraabdominal desmoid fibromatosis. *Ann Otol Rhinol Laryngol* 1994; 103: 331-4.
19. El-Sayed Y. Fibromatosis of the head and neck. *J Laryngol Otol* 1992; 106: 459-62.
20. Eitamo JJ, Hayry P, Nykri E et al. The desmoid tumor I. Incidence, sex, age, and anatomical distribution in the finish population. *Am J Clin Pathol* 1983; 77: 665-73.
21. Welling RE, Hermann ME, Kasper GC. Experience with desmoid tumor in a community teaching hospital. *J of Surg Oncol* 1992; 49 (2): 113-5.
22. Enzinger FM, Shiraki M. Musculoaponeurotic fibromatosis of the shoulder girdle (extra-abdominal desmoid). Analysis of ten cases followed up for ten of more years. *Cancer* 1967; 20: 1131-40.
23. Das Gupta TK, Brasfield RD, O Hara J. Extra-abdominal desmoids: a clinicopathologic study. *Ann Surg* 1969; 170: 109-21.
24. Johns MM, Taylor RA, Bogdasarian RS. Quiz case 2. *Arch Otolaryngol Head Neck Surg* 2000; 126: 905.
25. Hayry P, Reitamo JJ, Totterman S et al. The desmoid tumor. II. Analysis of factors possibly contributing to the etiology and growth behaviour. *Am J Clin Pathol* 1982; 77: 681-5.

Correspondence address:

Dr. Doroteo Vargas López

Hospital Regional «Lic Adolfo López Mateos»

Av. Universidad 1321, Mexico City.

E-mail: dovarlo@yahoo.com.mx