



Jornal Brasileiro de Patologia e Medicina Laboratorial

ISSN: 1676-2444

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Sociedade Brasileira de Patologia Clínica/Medicina Laboratorial

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Jornal Brasileiro de Patologia e Medicina Laboratorial, vol. 49, núm. 3, junio, 2013, pp. 208-211

Sociedade Brasileira de Patologia Clínica/Medicina Laboratorial
Rio de Janeiro, Brasil

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Pediatric fibromatosis involving mandible: case report and a five-year post-operative follow-up

Fibromatose pediátrica em mandíbula: relato de caso e avaliação pós-operatória de cinco anos

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ABSTRACT

Extra-abdominal fibromatosis is a benign fibrous neoplasm of locally aggressive behavior. Surgical excision with a wide margin is the treatment of choice. The aim of the present work is to report the case of fibromatosis in an 11-year-old melanodermic patient, who showed swelling in the area of the right mandible. Conservative surgery was performed based on the initial histopathological diagnosis of benign lesion suggestive of neural origin. The new anatomopathological examination of the surgical specimen was compatible with fibromatosis. The patient is still under periodic observation as part of the five-year surgical follow-up, showing no signs of recurrence.

Key words: desmoplastic fibroma; extra-abdominal fibromatosis; desmoid tumor.

INTRODUCTION

Extra-abdominal fibromatosis (desmoid tumor) is a benign fibrous neoplasm that arises from musculoaponeurotic structures with locally aggressive behavior^(12, 17). Fibromatoses are classified as superficial and deep. The deep type is aggressive, grows quickly and penetrates extensively, whether in a diffuse, multifocal way or locally, simulating malignancy^(2, 9, 10). Head and neck lesions are considered deep fibromatoses of the extra-abdominal type⁽¹⁴⁾.

Although extra-abdominal fibromatosis may affect any age group (newborn to elderly), it is an illness that predominantly affects children and young adults. It is frequently called aggressive juvenile fibromatosis^(7, 11). The most common affected places are the shoulder and trunk, with approximately 10% of cases occurring in the soft tissue of head and neck. Infantile extra-abdominal fibromatosis occurs more in females than in males at a ratio of 3:1⁽²⁾.

Its pathogenesis is unknown, but it is believed that hereditary factors and hormones play a role in the emergence of the lesion.

Some studies associate tumor growth with estrogen level, since there is a spontaneous regression found during menarche and menopause^(12, 16).

Clinically, extra-abdominal fibromatosis manifests as a painless, firm mass that grows rapidly and fixes to bone and soft tissue⁽²⁾. Invasion of bone or skin is not very common⁽¹¹⁾.

Histologically, the tumor consists of a proliferation of uniform-appearing, spindle-shaped fibroblasts, separated by dense collagen masses. The lesion has ill-defined proportions and tends to interlace with muscle fiber, hindering its surgical excision⁽¹²⁾. Although extra-abdominal fibromatosis does not generally metastasize or suffer sarcomatous degeneration, recurrence rate is high.

Treatment of head and neck fibromatosis is complete surgical excision, in which recurrence is high when negative surgical margins are not achieved^(2, 13).

Here is a case of extra-abdominal fibromatosis involving an 11-year-old patient.

First submission on 21/07/12; last submission on 22/04/13; accepted for publication on 09/05/13; published on 20/06/13

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CASE REPORT

An 11-year-old female, melanodermic patient came to the Oral and Maxillofacial Surgery and Trauma Unit of a municipal hospital with symptoms of pain and facial asymmetry. In the anamnesis, the patient's mother said that she did not know when the lesion had begun, but did say that swelling in the mandibular region of the right side was perceptible in the preceding two months, when the patient began to complain about sensitivity in the area. While performing the extraoral physical exam, a swelling of the lower third of the right side of the face was noticed. When performing the intraoral physical examination, it was discovered that the swelling extended from element 47 to the right mandibular ramus. Imaging examinations were requested. The panoramic radiograph showed a lytic lesion of infiltrative growth, with extensive destruction of the underlying mandibular bone area, causing upper displacement of tooth 48 (**Figure 1**). The computerized tomography showed the presence of a potentially expansive hypoattenuating lesion in the right mandibular ramus with a thinning of cortical bone. An internal and external expansion of the cortical bones could also be noticed, extending from the body to the middle third of the mandibular ramus (**Figure 2**). An incisional biopsy was performed and its diagnosis was suggestive of a benign lesion of neural origin. Based on the anatomo-pathological result, conservative surgery was performed. A new histological examination of the surgical specimen, in association with immunohistological evaluation, was compatible with fibromatosis. Five years after surgery, there was complete local bone repair, with no recurrence (**Figure 3**).

DISCUSSION

The term fibromatosis covers a range of rare benign soft tissue lesions that are locally invasive and may occur in various parts of the body. It is estimated that approximately 12%-15% of these lesions occur in the head and neck region and affect mostly

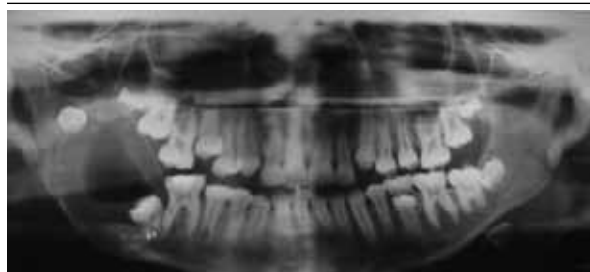


FIGURE 1 – Preoperative panoramic radiograph of the patient showed expansive lytic lesion located in the posterior mandible region

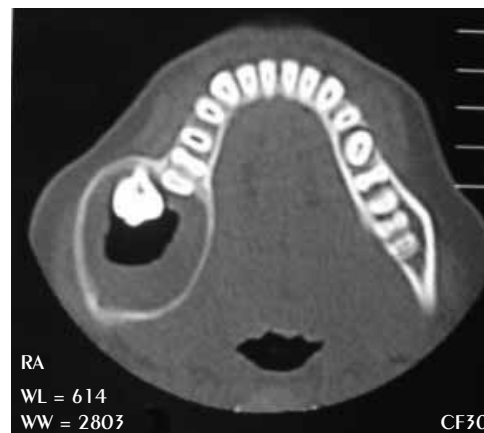


FIGURE 2 – Preoperative computed tomography showed the presence of a potentially expansive hypoattenuating lesion in the right mandibular ramus with a thinning of cortical bone

RA: right angle; WL: window level; WW: window width; CF: convolution filter.

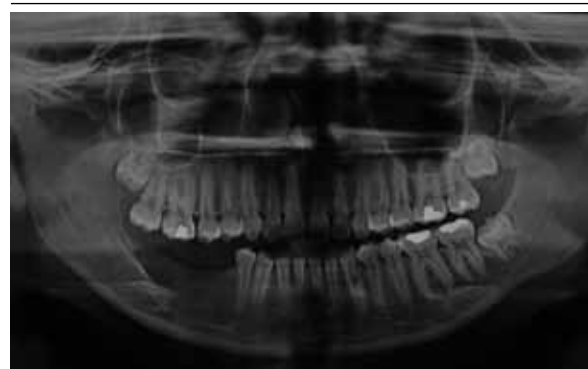


FIGURE 3 – Panoramic radiograph five years after surgery: observed complete repair of local bone with no recurrence

children and young adults. They are classified as extra-abdominal fibromatosis^(14, 15). However, there are other authors that claim that 34% of all cases of fibromatosis occur in the head and neck region⁽¹⁾.

As far as the growth of this disease is concerned, most studies^(2, 9, 10-12, 16) say that the lesion is of rapid growth and its differential diagnosis should include desmoplastic fibroma, nodular fasciitis, fibrosarcoma and neurofibroma⁽⁶⁾.

Due to histopathological likenesses, some authors believe that the desmoplastic fibroma is the osseous counterpart of fibromatosis. Furthermore, the desmoplastic fibroma may be distinguished from the nodular fasciitis by having a pattern of more infiltrative growth, larger production of collagen, less cellularity and less mitotic figures. In its turn, fibromatosis is distinct from fibrosarcoma due to the fact that malignant tumors present a herringbone pattern, pleomorphic and hyperchromatic nuclei as well as abundant and atypical mitosis^(7, 8).

Histologically, fibromatosis presents areas with many cells, alternating with areas with few, densely collagenized cells. Most cells correspond to mature fibroblasts with large uniform nuclei. In general, tumor cells are not atypical, being negative for S100 protein on the immunohistochemical test, which distinguishes them from neoplastic cells of neural origin^(2, 3, 11, 16).

According to Hauben *et al.*⁽⁴⁾, the presence of genetic mutations in the lesions may lead to an intranuclear accumulation of beta-catenin, resulting in an expressive nuclear immunoreactivity for this marker. However, in the study of Sharma *et al.*⁽¹²⁾, the authors affirm that beta-catenin is not a precise marker for the diagnosis of infantile fibromatosis.

In the present case, the histological examination of the surgical specimen revealed bundles of fibroblasts of large nuclei, ovoid or triangular, intertwined with large quantities of collagen (**Figure 4**). Highly cellular areas were intertwined with almost acellular, densely collagenized regions. There was no atypia or mitosis. The immunohistochemical examination showed a negative result for the S100 protein and positive for Vimentin (**Figure 5**), hence the appraisal report indicated fibromatosis.

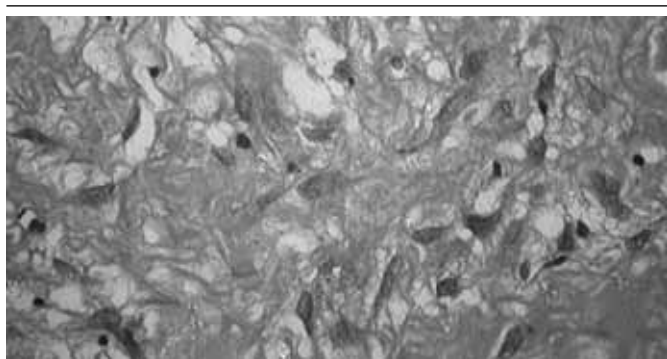


FIGURE 4 – The histological examination of the surgical piece revealed bundles of fibroblasts of large, oval or triangular nucleus, interspersed with large amounts of collagen (HE, 400x)

HE: hematoxylin and eosin.

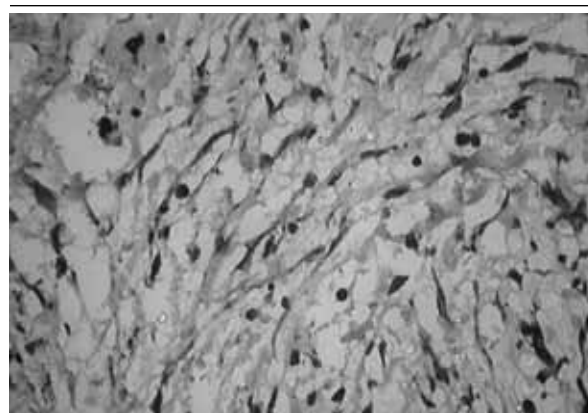


FIGURE 5 – The neoplastic fibroblasts were positive for vimentin (VIMENTIN, 400x)

Although there is still a lot of controversy and discussion in the literature, most authors agree that the treatment of choice should be the surgical excision of the lesion^(2, 5, 9). The extension of surgery varies considerably and depends on the size of the lesion, but excision with a wide surgical margin may be recommended, thus minimizing the risk of local recurrence^(2, 11, 16). However, this option is difficult in patients with tumors in the head and neck region due to the anatomical complexity and the proximity of vital structures⁽¹⁴⁾.

When complete resection of lesions does not occur, they have a local recurrence rate that varies between 50%-70% of cases^(3, 13). In children, surgical treatment of extra-abdominal fibromatosis is even more complicated because of the potential risk of functional incapacity⁽¹¹⁾. In the reported case, a less invasive treatment was chosen based on the initial diagnosis. As the final diagnosis of the surgical specimen was compatible with fibromatosis, careful follow-up has been undertaken. It is worth mentioning that in five years a complete repair of the affected bone area has been achieved, with no signs of recurrence to date.

RESUMO

Fibromatose extra-abdominal é uma neoplasia fibrosa benigna, caracterizada por comportamento agressivo local. Geralmente, o tratamento de escolha consiste na excisão cirúrgica com ampla margem de segurança. O objetivo deste trabalho é relatar um caso de fibromatose em uma paciente de 11 anos, melanoderma, que apresenta aumento de volume na mandíbula do lado direito. A partir do diagnóstico histopatológico inicial sugestivo de lesão benigna de origem neural, foi realizado tratamento cirúrgico conservador. Um novo exame anatomopatológico da peça cirúrgica foi compatível com fibromatose. A paciente encontra-se sob controle periódico de cinco anos da cirurgia, não havendo recorrência clínica da lesão.

Unitermos: fibroma desmoplásico; fibromatose extra-abdominal; tumor desmoide.

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