

AGGRESSIVE INTRACRANIAL FIBROMATOSIS

Case report

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ABSTRACT - Fibromatosis is a locally aggressive, proliferative fibroblastic lesion affecting musculoaponeurotic structures, most often in the limbs and trunk. Intracranial fibromatosis is extremely rare and requires aggressive treatment to prevent recurrence. *Case Description* - We present the case of a 20 year old woman with aggressive skull base fibromatosis. The lesion extended through the sphenoid, ethmoid sinus and nasal cavity, destroying the right roof of the orbit and penetrating in anterior skull base. A combined anterior craniofacial approach was performed; complete resection with surgical margin was impossible due to the localization of the tumor and relation to important neurovascular structures. Complete resection with surgical margin is often impossible because of its widely infiltrative nature. Radiotherapy and chemotherapy are often required to improve local control of the lesion.

KEY WORDS: intracranial fibromatosis, skull base, craniofacial approach.

Fibromatose agressiva Intracraniana: relato de caso

RESUMO - Fibromatose é lesão fibroblástica proliferativa com agressividade localizada, afetando as estruturas músculo-aponeuróticas e acometendo, mais frequentemente, os membros e tronco. A fibromatose intracraniana é rara e requer tratamento agressivo para evitar recidiva. Descrevemos uma paciente de 20 anos portadora de fibromatose agressiva de base de crânio. A lesão se estendia através da cavidade nasal, seios esfenoidal e etmoidal, destruindo o teto da órbita direita e invadindo a porção anterior da base do crânio. A paciente foi submetida a uma via craniofacial anterior combinada, não sendo possível uma ressecção completa devido ao envolvimento de estruturas neurovasculares. A ressecção completa é, frequentemente, impossível devido sua natureza infiltrativa. Radioterapia e quimioterapia são indicadas para melhorar o controle da lesão.

PALAVRAS-CHAVE: fibromatose intracraniana, base do crânio, via craniofacial.

Aggressive fibromatosis are fibrous tissue proliferations that arise from the connective tissue of muscle and its overlying fascia. They tend to infiltrate surrounding tissues, making complete surgical resection difficult. The nomenclature that has been used in the literature to describe this lesion is confusing. The characteristics fall between fibromas and low grade fibrosarcomas. Unlike fibrosarcomas, fibromatoses are without metastatic potential but tend to behave locally in an aggressive and infiltrative manner, making complete surgical resection difficult. The terms "desmoid tumor" or "aggressive fibromatosis" are used as synonyms for fibromatosis by some

authors, as the lesions tend to recur after surgical resection.

The purpose of this report is to describe a case of intracranial fibromatosis involving the skull base^{1,2}.

CASE

A 20 years old woman presented with a two years history of right proptosis and nasal obstruction. Physical examination revealed a right proptosis with no neurological abnormality. A computerized tomograph (CT) scan revealed a lesion extended through the sphenoid and ethmoid sinus, into nasal cavity, destroying the right roof of the orbit and the anterior floor of the cranial base. The magnetic resonance image (MRI) findings confirmed a large

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Fig 1. Sagittal and axial pre-operative MRI showing a large cranial base lesion which enhanced homogeneously after contrast.

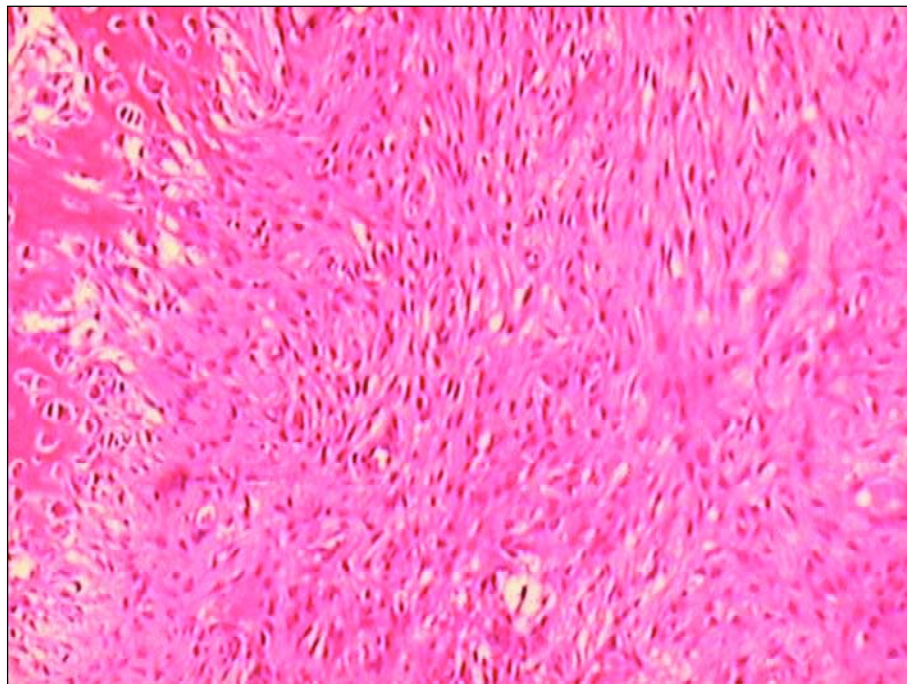


Fig 2. The tumor is composed of interlacing of fibroblasts and varying amounts of collagen. H.E., 200X.

cranial base lesion which enhanced homogeneously after contrast (Fig 1).

Surgical resection was performed through a combined frontal craniotomy and an anterior craniofacial approach (Weber Ferguson incision). A right frontal craniotomy permitted the dissection of the duramater that was not apparently infiltrated by the tumor. Inferiorly a midfacial, transmaxillar approach was performed and an extremely dense fibrous tissue infiltrating through bony structures was seen, and a sub total resection of the tumor, and surrounding invaded structures was achieved. Histopathological examination showed fibroblastic spindle cells with mild nuclear pleomorphism with a generous collagenous component (Fig 2).

The patient had an uneventful postoperative recovery with resolution of proptosis. Post-operative CT showed no

residual tumor. Follow-up MRI, eighth months post-operative showed recurrent tumoral tissue in the skull base centered upon the basi-sphenoid (Fig 3). The patient was submitted to a new surgical resection using an endoscopic nasal approach to the lesion, this time with partial removal.

DISCUSSION

Fibromatosis is a tumour-like fibroblastic proliferation characterized by a tendency to infiltrate surrounding tissues and to recur locally after surgical excision, but not associated with a capacity for metastasis^{3,4}. Fibromatosis can occur at any age, but is more common between the ages of 20 and 40 years, with a female predominance in most series. Head and neck

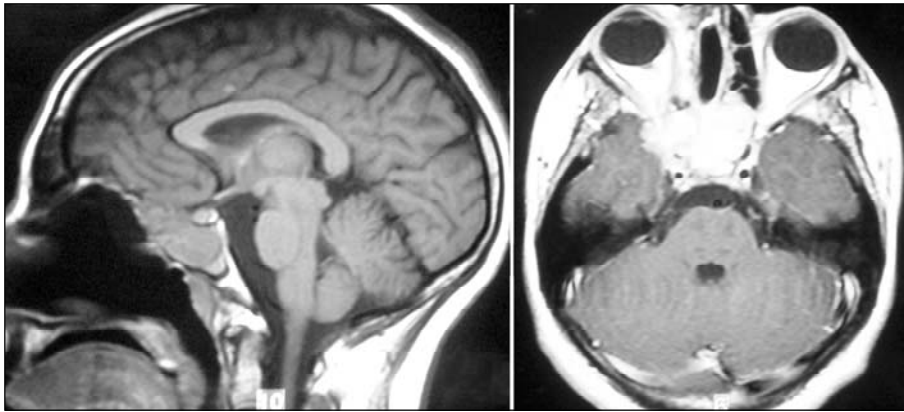


Fig 3. Follow-up MRI, post-operative showing recurrent tumoral tissue in the skull base centered upon the basi-sphenoid.

fibromatosis tend to display characteristics that differ somewhat from aggressive fibromatosis in other locations. They tend to occur in a younger age group, often in the first or second decade and have a high recurrence rate^{1,2,5}. Most cases of aggressive fibromatosis are sporadic, but fibromatosis can also occur in association with familial adenomatous polyposis as part of Gardner's syndrome^{6,7}.

The rate of local recurrence after surgical resection ranges from 27 to 77%. Most recurrences occur within two years. Death is uncommon but may occur as a result of compression of vital structures, such as the trachea. Approximately 9 to 27% of aggressive fibromatosis are located in the head and neck. Head and neck fibromatosis may behave somewhat differently from other aggressive fibromatosis².

The nomenclature that has been used in the literature to describe fibromatosis is confusing. Synonyms for aggressive fibromatosis include desmoid tumor, desmoid fibromatosis, deep fibromatosis, Grade 1 fibrosarcoma (desmoid type), desmoma, and desmoplastic fibroma of bone⁵.

Fibromatosis usually present as painless masses, however, pain is not an unusual symptom. Neuropathic "shooting" pain occurs rarely. In the head and neck area functional deficits, including trismus, speech impairment, dysphasia, nasal obstruction, and difficulty in closing the eye, have been reported^{7,8}. It is often difficult to ascertain the site of origin of an aggressive fibromatosis, but most authors tend to agree that intracranial fibromatosis arises from the dura^{3,4}. In our case due to the diffuse appearance of the tumor, it was impossible to precise the site of origin.

Histologically, desmoid fibromatosis arising in adulthood, irrespective of site, is composed of spin-

dle-shaped, uniform cells surrounded by abundant collagen and arranged in interlacing fascicles or a pseudolobulated pattern. The degree of cellularity is usually moderate but can vary from area to area within the tumor. Nuclei are never atypical or hyperchromatic, and mitoses can occur but are never abnormal. Mononuclear cells, usually mast cells, are scattered throughout the stroma. Infiltration into adjacent structures occurs frequently, even when the mass appears grossly well circumscribed. Differentiating tumor tissue from peripheral scar can be difficult. Bony erosions, as occurred in our case, may occur when the periosteum is involved. The differential diagnosis for pathologist includes fibrosarcoma, reactive fibrosis, and nodular fasciitis^{2,5,9}.

Fibromatosis have typical MRI features. They are isointense to slightly hyperintense in T1 weighted images, intermediate between muscle and fat in T2 weighted images and enhance after administration of contrast agent. These typical findings were evident in our patient. The lack of peritumoral soft tissue oedema and the presence of areas with very low signal intensity in T2 weighted images suggested a slowly growing and more benign proliferative soft tissue disorder¹⁰.

The treatment of choice for intracranial fibromatosis is surgery with adjuvant radiotherapy and chemotherapy being indicated for some authors, although the effects of these therapies is unclear^{3,4}. No clinical trials have been performed to clarify the best approach to treat fibromatosis. The aim of surgery in the treatment of any aggressive fibromatosis is complete excision with tumor-negative margins. Most series show that "more complete" excisions result in better local control¹¹⁻¹³. However, these lesions have

no capsule and so they tend to infiltrate surrounding tissues microscopically, becoming complete excision often impossible. This is especially true in the head and neck and particularly the cranial base, where the density of vascular and neural structures often prevents complete excision. In our case, a total excision with negative margins was impossible because of the extensive cranial base involvement with massive bone destruction and infiltration. The lesion was also very difficult to remove, being adherent to and infiltrating surrounding structures.

In the series from Perez-Cruet et al.², four of the five initial surgical procedures for skull base fibromatosis had gross macroscopic resections, but margins were involved in all cases. Two of the four patients experienced recurrences and the patient who underwent grossly incomplete initial resection had multiple recurrences. We recommend nonsurgical treatment for aggressive fibromatosis, particularly in situations in which a wide field resection would result in unacceptable morbidity¹⁴.

Several series have reported that radiotherapy can improve the local control of fibromatosis^{1,2}, and has been used in extracranial lesions to control marginal and incomplete resections and to treat recurrent disease. Radiotherapy was not used on gross tumour masses in the patients reported by Perez-Cruet et al.², so regression could not be assessed. Of the three patients treated with postoperative radiotherapy, two remained free of recurrent disease. Chemotherapy has also been used successfully to treat fibromatosis¹⁴, primarily in the pediatric population. Goepfert et al.¹⁵ reported six patients with desmoid fibromatosis or fibrosarcoma grade 1 desmoid type. The two chemotherapeutic regimens administered systemically were: 1. adriamycin and dacarbazine; 2. vincristine, dactinomycin and cyclophosphamide. Four patients responded partially, whereas two patients demonstrated complete regression of their tumors. Because tumor cell expression of estrogen receptors has been demonstrated in aggressive fibromatosis, hormonal therapy with tamoxifen has been attempted. Although the

data are limited, regression of tumors in response to tamoxifen has been reported¹⁶.

Transformation of aggressive fibromatosis to malignant sarcomas is exceedingly rare. Only a few cases of transformation have been reported in the literature².

In conclusion, fibromatosis is a histologically benign fibroblastic proliferative lesion that is locally aggressive but does not metastasize. An intracranial location is extremely rare. Treatment consists of an adequate three-dimensional imaging evaluation, complete resection is often impossible to achieve because of the infiltrative nature of the lesion. Postoperative radiotherapy, chemotherapy, and recently, hormonal adjuvant therapy have been used in an effort to control local recurrence.

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