ORIGINAL RESEARCH PAPER

Volume-7 | Issue-3 | March-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

AGGRESSIVE FIBROMATOSIS OF THE NECK AND MANDIBLE; REPORT OF TWO CHALLENGING CASES

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ABSTRACT

We present our experience of two cases of rare connective tissue disease occurring within the midface. First case represented a very large painless slowly growing neck mass extending from mid neck to the clavicle displacing the trachea to the other side. Second case represented a painful mass in the right posterior mandibular region that had overgrows teeth. FNA and incisional biopsy did not confirm the diagnosis. A complete excision of the lesion with invasion of adjacent structure confirms the diagnosis of Aggressive Fibromatosis. Different modalities of the treatment will be discussed.

KEYWORDS

aggressive fibromatosis, desmoid tumor, neck mass, mandible mass

INTRODUCTION

Aggressive Fibromatosis, also called desmod tumor, is a rare benign that originate from musculoaponeurotic structures. Although, histopathologically benign with highly aggressive feature of adjacent structure

Fibromatosis usually present as painless masses, however, pain is not an unusual symptom. In the head and neck area, functional deficits, including trismus, speech impairment, dysphagia, nasal obstruction and difficulty in closing the eye, have been reported (Fowler, Hartman, Brannon, Am J Surgery 1999).

Reported most common cases between the ages 20-40 yrs with female predominance 2:1. The rate of recurrence after surgical resection ranges from 36 to 77% (Fowler, Hartman, Oral surg pathol 1994). Most authors recommend surgical excision as the initial approach, with radiotherapy and chemotherapy used adjunctively.

We report two cases challenging, young female and teenager girl with aggressive fibromatosis. Both cases underwent CT or MRI prior to complete excision of the lesion under general anesthesia to confirm the diagnosis. None of two cases were received radiotherapy or chemotherapy postoperatively

CASE REPORT – 1

A 24 years old female patient, presented with a history of painless left sided neck mass, which has been gradually increasing in size (Have been reported to another hospital 6 months prior to presentation). She also gave a history of mild odynophagia and difficulty in neck movement. There was no history of dysphagia, change of voice, or dyspnea. Her weight and vitals were normal.

On physical exam, the mass was large, firm, non-tender and non-pulsating. Filling the left aspect of the neck extending from the mid neck to the clavicle and from the posterior triangle to the midline displacing the trachea to the right side. The mass was fixed to the underlying structures but the covering skin was freely mobile.

There were no palpable cervical lymphadenophathy.

On radiological evaluation, computerized tomography scan of neck showed a large mass evolving the left prevertebral muscle from the mid neck to the thoracic inlet causing a displacement of the carotid and the jugular vein in anterior direction (Fig.1).

The patient was taken up for further investigation; FNA biopsy was obtained followed by incisional biopsy. The histopathologcal evaluation of biopsies showed Spindle cells, fibrohistocystic lesion, favouring benign without evidence of malignancy. There was no definite diagnosis yet.

On the basis of radiological and histopathologcal findings with no definitive diagnosis, a wide surgical excision of the lesion was preformed under general anaesthesia (Fig.2). The lesion aggressively invaded the internal jugular vein, which has been resected with the lesion. Final pathological evaluation revealed 10 x 6 x 5 cm mass with local infiltration to prevertebral muscle (Fig.3), the diagnosis of aggressive fibromatosis confirmed. No postoperative complications were experienced.

Unfortunately, recurrence occurred after 6 months follow up, complete surgical excision done. Local radiotherapy treatment was considered but the patient refused. The patient remains free of the disease for over 2 years with close follow up

CASE REPORT – 2

A 14 years old girl presented with mandibular pain since four weeks on the right site with impaired mastication in a dentist's office. On physical exam, a tumescence of about three centimeters was noted in the right posterior mandibular region that had overgrown teeth 47. Supposing an epulis biopsy was taken that showed an atypical proliferative fibromatosis. The young girl was subsequently addressed to our department for further assessment and treatment.

On radiological evaluation, CT scan and MRI showed an osteodestructive soft tissue process (Fig.5), evoking the suspicion for fibrosarcoma or osteosarcoma.

To assure the diagnosis a more extensive biopsy in general anesthesia was necessary, confirming an aggressive juvenile fibromatosis. Afterward the tumor was removed transoraly with partial mandibulectomy on the right site by the cervicomaxillary division. The postoperative follow-up was without complication.

Five years later mandibular atrophy occurred (Fig.6), that required an autologous osteosythesis. The now 21 years old girl is in good health without clinical signs of recurrence.

DISCUSSION

Aggressive fibromatosis is a proliferation of fibrous tissue that arise from fascia, periosteum and musculoaponeurotic structures, it is derived from mesenchymal cells. And characterized by a tendency to infiltrate surrounding tissues and to recur locally after surgical excision. [2, 3].

The nomenclature to describe fibromatosis in the literature is confusing.

Synonyms; Desmoid tumor, Desmoid fibromatosis, Grade I fibromatosis (desmoid type) and Desmoplastic fibroma.

Most cases are sporadic, but can occur in association with familial adenomatosis plyposis, osteomas of the skull bones as a part of Gardner Syndrome. [4,5,6]

There has been an observed incidence of trauma of about 19–49% preceding the development of cases of aggressive fibromatosis. The pathogenesis is thought to involve abnormal response of healing with

persistent immature fibroblast formation which later gives rise to the tumor. Steroid sex hormones are also believed to play a role in pathogenesis. [7]

Fibromatosis can occur at any age but is most common between the ages 20-40 yrs. (8) Most series report a female predominance 2:1. The rate of recurrence after surgical resection ranges from 36 to 77% [1]. Most recurrences occur within 2 years.

Approximately 9-27% of Aggressive Fibromatosis are located in the head and neck region.

Fibromatosis usually present as painless masses, however, pain is not an unusual symptom. In the head and neck area, functional deficits, including trismus, speech impairment, dysphagia, nasal obstruction and difficulty in closing the eye, have been reported [1]

A representative tissue biopsy is required for diagnosis. [9] Histologically spindle shaped uniform cells surrounded by abundant collagen, the degree of cellularity is moderate but can vary from area to area within the tumor. Nuclei are never at typical or hyperchromatic, and mitosis can occur but are never abnormal (Fig.4).

The differential diagnosis for pathologists includes: Fibrosarcoma, Reactive fibrosis, Nodular fasciitis , hypertrophic scars and keloids. [10]

MRI is the investigation of choice to assess the extension of the disease and for follow up, because of superior soft tissue representation in comparison with CT. [11, 12]

No clinical trials have been performed to clarify the best approach to treating fibromatosis. Most authors recommend surgical excision as the initial approach, with radiotherapy and chemotherapy used adjunctively.

The aim of surgery is complete excision with tumor-negative margins. Because fibromatosis tend to infiltrate surrounding tissues, microscopically complete excision is often not possible.

This is especially true in the head and neck region and particularly the skull base, where the density of vascular and neural structures often prevents complete excision.

Head and neck fibromatosis demonstrate a recurrence rate of 40 – 70 % after resection.The majority of which occur within 18 months of excision .[13]

The age of the patient and tumor site are the known factors associated with recurrence [14]. The role of the microscopic status of tumor margins is more complex. Some large retrospective studies showed high local recurrence rate with microscopically positive margins. [15, 16]; other studies failed to demonstrate an effect of microscopic margins on recurrence [17, 18]

Several series have reported that radiotherapy can improve the local control of the tumor. In the series of studied by **Plukker et al., 1998,** patients with incomplete resection or recurrent disease were treated with at least 50 Gy of wide field radiotherapy. [19]

Chemotherapy has also been used successfully to fibromatosis, primarily in the pediatric population. **Goeprfert et al. 1998** reported the treatment of six children with desmoid fibromatosis of the head and neck with one or two chemotherapeutic regimens. Because tumor cell expression of estrogen receptors has been demonstrated in Aggressive fibromatosis, hormonal therapy with Tamoxifen has been attempted. [20]

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

Death is uncommon but may occur as a result of compression of vital structures, such as the trachea and major blood vessels of the neck.

CONCLUSION

Aggressive Fibromatosis is a poorly defined, locally aggressive. histologically benign fibroplastic proliferative lesion that may occur in the head and neck. The lesion is highly cellular and locally infiltrative compromising vital structures within the head and neck.

Wide field resection is the aim of treatment. Radiotherapy, Chemotherapy and hormonal therapy are other modalities of treatment. Long-term follow up is necessary because aggressive fibromatosis may recur after surgical excision.

Fig.1

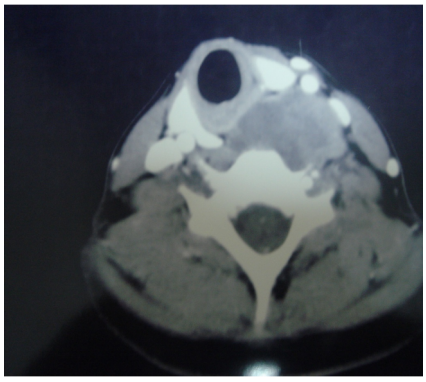


Fig.2

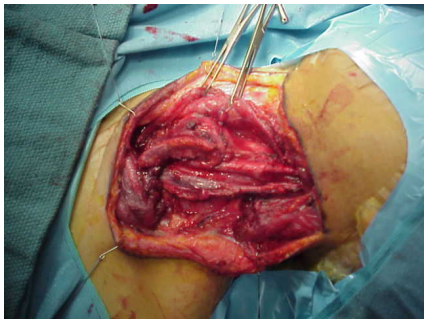


Fig.3

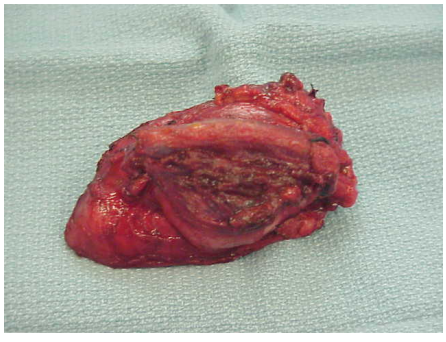


Fig.4

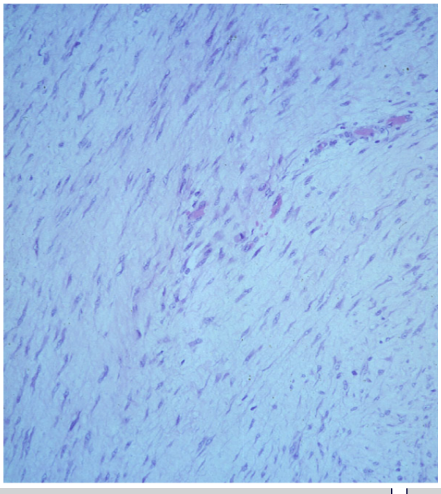


Fig.5

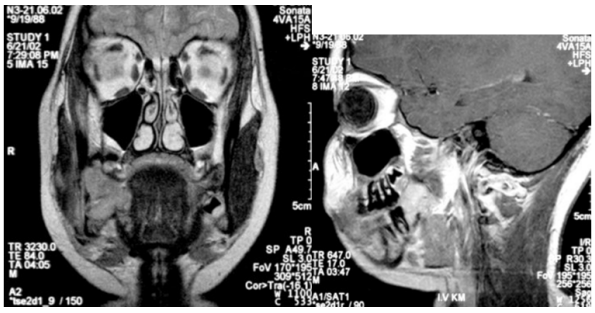
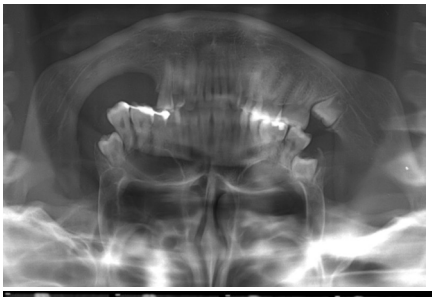
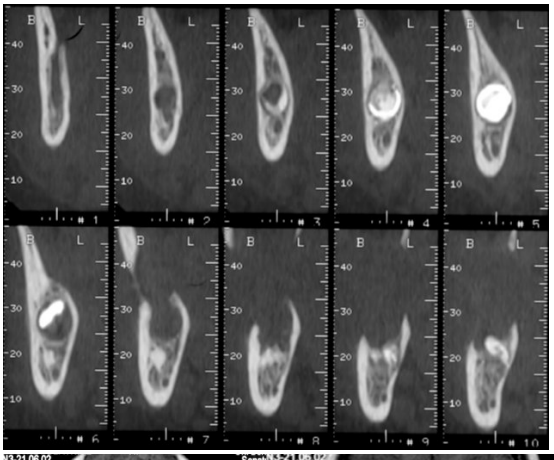
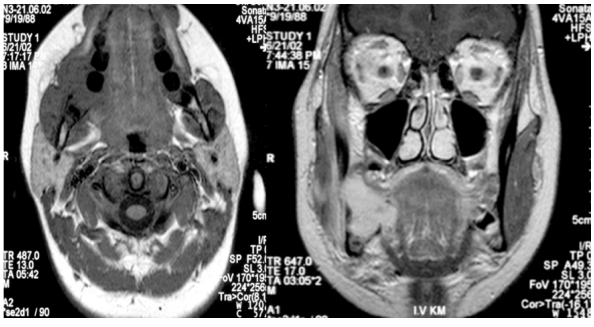


Fig.6







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