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AGGRESSIVE FIBROMATOSIS: A CASE REPORT

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Abstract

Aggressive fibromatosis is an uncommon locally invasive non-metastasizing neoplastic lesion. The aetiology of this tumor is unknown and its treatment is controversion. A case of an aggressive fibromatosis in a 32 years old woman which recurred 6 times in 2 years was presented and relevant literature was reviewed on this rare manifestation.

Key words: *fibromatosis, aggressive fibromatosis, desmoid tumor*

INTRODUCTION

Aggressive fibromatoses are fibrous tissue proliferations that arise from connective tissue. They tend to infiltrate surrounding tissues which make it difficult for complete resection. Although they do not metastasize, fibromatoses tend to recur after surgical resection. This report describes a patient with an aggressive fibromatosis which recurred 6 times in 2 years and reviews the recent literature on this rare manifestation.

CASE REPORT

A -32- year old woman was originally referred 2.5 years ago to a dental hospital withmandibular pain and swelling. She had had oral aphtous lesions since her childhood, but she suffered mandibular pain for the past 12 months. She had the tumor excised and the pathology report was malignant mesenchymal tumor (leiomyosarcoma with intermediate differantiation). She underwent subsequent rebiopsies in following months because of unremitting pain and swelling at this same location. These biopsies were diagnosed in different pathological reports as malignant mesanch.ymal tumour, gingival fibromatosis , chronic fibromatosis , gingival fibromatosis or odontogenic fibroma. She had not received any further therapies apart from surgery during that period. Her family history was noteworthy that her older sister died of a bone tumour of unknown type at age 18. Finally in September 1999 she was admitted to Marmara University Hospital with poorly controlled pain. In examination, her mandible was found to be painful on palpation, tongue and chin had restricted movements; an infiltrative tumour had been observed in the mandibular corpus mainly to the right side of the midline, deeply penetrating into the dermis. The MR study revealed a mass which was placed in the base of mouth, caused a destruction in the anterior and right anterolateral part of the mandible (no progression or regression when compared with the previous MR examinations) and lympadenopathies in the submental (a few mm in diameter), suboccipital and bilateral servical regions (15x9 mm on the right and 16x7 mm on the left).Hemimandibulectomy was performed.

Macroscopically 7x4.5x4 cm mandibulectomy specimen included 3x2.5cm skin on its surface and a tooth. The specimen had bone at the edge of the long axis. The cut surface was firm and had white fibrillary appearance. Morphologically the tumour was poorly

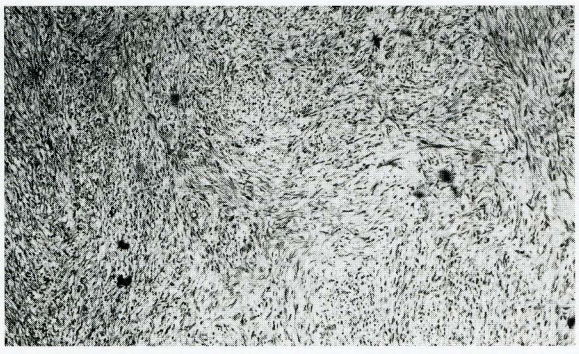

Figure 1. The spindle cells in a fasicular pattern (H&E X200)

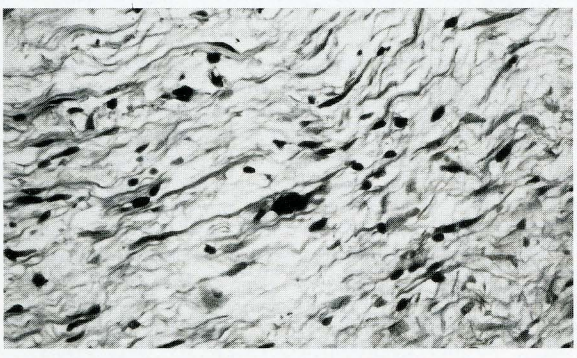

Figure 2. The spindle cells wiith prominent pleomorphism (H&E X400)

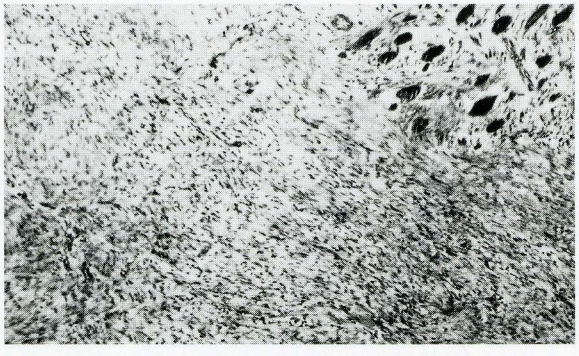

Figure 3. Entrapped skeletal muscle fibers at the advancing margin of the tumor(H&E X200)

circumscribed and composed of spindle shaped uniform cells surrounded by abundant collagen and arranged in interlacing fasicular pattern (Figure 1). Reticulin and Masson trichrome stain clearly brought out the abundance of collagen between the individual tumour cells. The differential diagnosis included fibrosarcoma, reactive fibrosis and nodular fasciitis (1). The degree of cellularity was generally moderate but showed variance from area to area within the tumour. Nuclei were uniform and normochromatic in the majority of the cells, but there were hyperchromatic, bizarre nuclei in some areas (Figure 2). Mitoses were rare (less than 3/HPF).There were mononuclear cells which were mainly mast cells scattered throughout the stroma. İnvasion through the striated muscle and bone was observed (Figure 3). Finally with all these findings the tumour diagnosed as aggressive fibromatosis.

Treatment: This patient had to receive postoperative radiotherapy as the tumor had an aggressive behaviour, recurred several times locally in spite of surgery and never received adjuvant radiotherapy, it had infiltrated into bone and muscle with positive surgical margins. She had received 60Gy external radiotherapy in 30 fractions 6 MV photon beams, parallel opposed fields, with shields placed for maxillary protection. Following the radiotherapy the patient was screened for disease beyond the original region and none was found. An experimental adjuvant combination chemotherapy with an anthracycline and Ifosfamide was suggested but she refused any treatment that may cause hair loss. Therefore she was treated with 1250mg/m2 Ifosfamide for 5 days for 2 cycles at 28 days intervals.

DISCUSSION

Aggressive fibromatosis is a proliferation of fibrous tissue characterized by a tendency to infiltrate surrounding tissues and recur locally after surgical excision. In the literature the nomenclature that has been used to describe aggressive fibromatosis is confusing and includes desmoid tumour, desmoid fibromatosis, deep fibromatosis, Grade 1 fibrosarcoma (desmoid type) as synonyms (2,4). However It differs from malignant tumors such as fibrosarcoma as it has no metastatic potential (5).

Molecular studies examining the patterns of X-chromosome inactivation have confirmed that these lesions are the result of a clonal process, demonstrating that desmoids are neoplasms and not the product of an intense inflammatory response (6).

Fibromatosis can occur at any age but is most common between the ages 20-40 years (4). Approximately 9-

27% of aggressive fibromatosis are located in the head and neck. The rate of occurrence after surgical resection ranges from 36 to 77% and mostly it recurs within 2 years. Head and neck fibromatoses demonstrate a recurrence rate of 46 to 70% after resection which may be slightly higher than the recurrence rates for fibromatoses in other areas. Death is uncommon but may occur as a result of compression of vital organs such as the trachea (3,4,8,9).

Most cases of aggressive fibromatosis are sporadic , but fibromatosis can also occur in association with familial adenomatous polyposis as part of Gardner's Syndrome (1,10).

Histologicaly fibromatoses are composed of spindle shaped uniform cells surrounded by abundant collagen and arranged in intertwining fascicles or a pseudolobulated pattern.The degree of cellularity is usually moderate but can vary from area to area within the tumour .Nuclei are uniform , normochromatic and mitotic figures are rare. There may some chronic inflammatory cells which are scattered throughout the stroma.

Many clinical trials have been attempted to identify the best approach for treating fibromatoses. Surgical resection is the primary treatment modality but due to infiltative nature of these tumors the local reccurence rates after wide local excision remain high. For this reason most authors recommend surgical excision as the initial apprroach, with radiotherapy and chemotherapy used adjunctively based on individual basis (8). On the other hand Merchant et al (7) observed that positive resection margins were not predictive of recurrence and selective use of adjuvant radiation therapy did not influnce the rate of local reccurence regardless of the margin status. Here we reported a patient with aggressive fibromatosis of the mandible who underwent 5 surgical resections. The lesion recurred 6 times following resections. The aim of the surgery in each instance was complete excision with tumor negative margins. Unfortunately because the tumor had infiltrated surrounding tissues, microscopic complete excision was not possible. Therefore after the last resection, both types of adjuant therapies were done and she remains well for 8 months.

Özet

Agresif fibromatozis

Agresif fibromatozis metastaz yapmayan ancak lökal invazif özellikte bir neoplastik lezyondur.Henüz tam olarak etyolojisi bilinmeyen bu tümörün tedaviside tartışmalıdır. Aggresif fibromatozis tanısı alan ve tümörü 2 yıl içinde 6 kez nüks etmiş olan hastamız ilgili literatür ışığında sunulmuştur.

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