Aggressive fibromatosis in the upper lip: A case report

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ABSTRACT

Aggressive fibromatosis involving the facial bone and facial soft tissue is rare. We present a case of aggressive fibromatosis of the upper lip in a 72-year-old male, who was treated by aggressive resection with 16 months follow up and no recurrence.

Key words: Aggressive fibromatosis; Fibromatosis; Lip

Introduction

The term fibromatosis refers to a group of fibrous tumor or tumor like lesions of soft tissues that share similar microscopic characteristics and possess an intermediate biologic potential between benign and malignant lesions. Fibromatosis was perhaps first described in 1839 by Dupuytren in reference to nodules seen in the palmar fascia of children. Following this little was published concerning this condition until the middle of the twentieth century, when Stout, Mackenzie and others described this tumor. We present a case of aggressive fibromatosis involving the upper lip where the tumor was widely excised with buccal corticotomy with a sixteen month follow-up.

Case Report

A 72-year-old male patient had a painless swelling on the upper lip that had been present for a few months. The swelling was firm, fixed and was non-tender. The mucosa over the swelling appeared normal (Figure 1). Patient gives history of trauma to the middle 1/3rd of face few years back. Incisional biopsy revealed as aggressive fibromatosis. The tumor continued to grow after the incisional biopsy suggesting more aggressive lesion. The computed tomography showed a low-density soft tissue image with uneven boundaries (Figure 2). There was no bone erosion in the anterior maxilla. A wide excision of the lesion and buccal corticotomy with less than 1cm of free margin was performed. Histological diagnosis from paraffin sections confirmed the diagnosis. He has been followed-up regularly, and there has been no recurrence so far.

Discussion

Aggressive fibromatosis is a histologically benign yet clinically invasive lesion that involves the jaws. The hallmark of this neoplasm is its high incidence of recurrence (up to 63%). Patients with aggressive fibromatosis often present with a deep, firm mass with or without pain. Mandible is more frequently involved than the maxilla. Females are more commonly affected than males.

The cause and pathogenesis are unknown although studies have implicated trauma, endocrine and genetic factors in tumor development. There are high incidences of estrogen and anti-estrogen binding sites in the disease and collagen can be produced by culture of cells from the tumor in response to estrogen. Aggressive fibromatosis show is varying degrees of potential for local infiltration, although the histological and clinical criteria for classification as sarcoma may be absent. Although the lesion may cause death by local invasion, it does not metastasize. These lesions exhibit secondary erosion or invasion of bone, which demonstrates the locally infiltrating nature of these lesions.

The treatment of aggressive fibromatosis is traditionally surgical resection with an area of the tumor-free tissue. Radiation therapy has been used alone when resection might lead to serious morbidity and loss of function and when tumor is inoperable or there is gross residual disease after operative debulking. Chemotherapy alone does not appear to be curative, but it may be effective in controlling unresectable tumors, or in reducing tumor size before surgery. Chemotherapy in combination with enblock resection has also been effective in managing recurrent disease.

The role of systemic treatment includes administration of drugs such as tamoxifen, toremifene, non-steroidal anti-inflammatory drugs, or biological agents such as interferon or retinoic acid. 1,25-(OH)2-vitamin D3 has also been shown effective in cases of progression and local recurrence. The prognosis depends entirely upon the initial treatment; if this is inadequate, the tumor will recur one or more times and progress.

The incidence of recurrence following resection and radio-chemotherapy is 20-35% and recurrence following operation alone is reported to be around 40%. Until now, only 16 cases have been reported (including the present one) from 1980 to now.6 We have achieved a good result with wide surgical resection with a 1-year follow-up. Adjuvant radiochemotherapy is valuable if the tumor recur.

Conclusion

In this article, we are adding another case of this rare tumor to the literature database. Early diagnosis and prompt surgical treatment in Oral fibromatosis are of prime importance. Patients should be kept under periodic observation because of reports of its high recurrence.
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How to cite this article

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Source of Support: Nil
Conflict of Interest: None Declared