CASE REPORT

FIBROMATOSIS OF THE FACE - A CASE REPORT

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HOW TO CITE THIS ARTICLE:

Rajesh Kumar Singh, Vijayanand Choudhary, Sangeeta Pankaj. "Fibromatosis of the face - a case report". Journal of Evolution of Medical and Dental Sciences 2013; Vol2, Issue 37, September 16; Page: 7155-7159.

ABSTRACT: Desmoid tumours, also known as desmoid fibromatosis, deep fibromatosis and aggressive fibromatosis constitute a rare fibroblastic proliferative disease. Desmoid tumours can occur anywhere in the body and are generally divided by anatomic designation as extra-abdominal, abdominal wall and intra-abdominal. Fibromatosis in the submandibular region is rare. Only 8% of the total desmoid tumors are located in head & neck region. They are locally aggressive and tend to recur after surgery; they also cause considerable morbidity, particularly when they arise in the head and neck. Though they lack malignant nature, their management becomes challenging due to their aggressive infiltrating nature and may even result in impairment or loss of functions of vital organs. Surgery, radiotherapy, estrogen antagonist, chemotherapy or the combinational therapies have been proposed by various oncologists for its management. Chemical ablation, radiofrequency ablation, and cryoablation have been used to treat patients with desmoid tumors which have recurred after surgery.

KEY-WORDS: Fibromatosis, Desmoid tumors, Head & Neck Cancer, Sub-mandibular Region, Radiotherapy, Chemotherapy

Key Message: A combination therapy comprising of radiotherapy and chemotherapy after surgery resulted in tumor regression of around 70 % in the patients without recurrence. However, a well-coordinated, multidisciplinary approach involving the input of surgical and nonsurgical specialists is needed to develop an individualized treatment strategy appropriate for each specific patient.

INTRODUCTION: Desmoid tumors are rare tumors with an estimated incidence of approximately 2-4 cases per million and only 900 new cases are diagnosed in US per year. Since these tumors have a strong potential for local invasiveness, high recurrence rates, tendency toward destruction of surrounding tissue and significant morbidity and potential mortality, they are classified as semi-malignant. These tumors constitute 0.03% of all biopsy analyzed neoplasms and less than three percent of all biopsy-analyzed soft tissue tumors.^[1] Due to its rarity, fibromatosis has been accepted by the National Organization of Rare Disorders (NORD), US, as a rare disorder. Several studies have shown that fibromatosis in females of childbearing age have a greater growth rate than that in pre or postmenopausal women and also greater than in males. The potential morbidity of surgery and radiotherapy and the high local recurrence rates have led investigators to assess the role of pharmacological approaches in settings in which surgery and radiation therapy are either not possible or unsuccessful [11]. There is almost 2 :1 preponderance in females.^[2]

CASE REPORT: On May 5th, 2009, A 55 Year old male came to the oncology OPD, with the history of progressively enlarging left naso-facial mass since last 2 Years. Past history revealed that he was operated 2½ years ago for a mass at the same sight. Complete excision of the tumor was done. Histopathology of the mass showed a desmoid tumor with negative resection margins. He did not

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receive any adjuvant therapy. He was lost to follow up after the surgery and never visited the surgeon again. 2 years back he noticed a mass on the left dorsum of the nose which remained asymptomatic for a month, and gradually infiltrated into the soft tissue of the left cheek overlying the maxilla. The diffused swelling extended up to nasal bridge and down wards up to upper lip. The overlying skin was inflamed but non-tender (Figure1). Plain X-Ray showed haziness in the PNS area. CECT scan showed a mass with heterogeneous enhancement on left nasal cavity and mass effect on the septum extending into the soft tissue of cheek overlying maxilla into nasal cartilage. There was destruction of the left infra orbital wall. There was also infiltration into the ethmoid sinus. Tissue diagnosis was established by incisional biopsy. Histopathology report was diagnostic of desmoid tumor (Figure 2, Figure 3). Presuming it's benign nature and in view of large size of the tumor, radiotherapy was started with palliative intent. Radiotherapy was planned for 50 Gy/ 25# /5 weeks with weekly chemotherapy, with Docetaxel (80 mg). Radiation was given by shrinking field technique. Patient was on radiotherapy from Monday to Friday and chemotherapy on every Saturday. There was tumor regression by around 70 % as assessed clinically. The course of treatment was uneventful. The patient did not agree for further investigation so, post treatment C-T could not be done. The patient came twice for follow up at three months interval with stable disease but did not turn up later. In June 2011 we came to know from his wife that he had died five months back.

DISCUSSION: Desmoid tumors incidence, are most frequently located Intra-abdominally (20%) followed by lower extremity (16 %), abdominal wall (16 %), chest wall (15%), upper extremity (14 %), back (6%) and pelvic girdle (5 %). The occurrence of desmoid tumor in Head and Neck region is around 8 %.^[3] This tumor usually manifests as a painless, slow-growing, deep-seated mass that is fixed to the underlying soft tissue.^[4] These tumors do not metastasize but due to their invasive nature frequently recur within 2 years after the initial excision.^[5]

12% of extra-abdominal desmoid tumors arise in the head and neck. ^[6] Desmoid tumors may recur aggressively after excision and infiltrate other organs. Recurrence rates vary depending on their location with higher rates reported in those arising in extra-abdominal locations. ^[7] Reported local recurrence rates vary between 20% and 60% at five years from surgery. ^[8] Several studies have reported a correlation between a positive tumor margin and the incidence of local recurrence. ^[9] However recent evidence suggests that the status of resection margins is not a reliable predictor of recurrence. Several studies demonstrate a better control of local disease with surgery followed by radiotherapy. ^[10]

Mainstay treatment is surgery particularly for patients with primary resectable and localized tumors where standard goal is complete resection with negative microscopic margins. Potential mutilation and association with considerable functional loss or major morbidity of surgery emphasize that surgical therapy must be performed to what is achievable in terms of margins while preserving functional status for the individual patient.^[12] Radiotherapy as the treatment of aggressive fibromatosis has been used either as adjuvant therapy with surgery with positive margins or as primary therapy for extra-abdominal tumors that are unresectable or where surgery derived functional/cosmetic loss is unacceptable^[12]. In a retrospective analysis of clinical data of 95 patients with aggressive fibromatosis who underwent therapy at the university of Michigan from 1984 -2008 suggest equivalent local control rates between the groups receiving only surgery, only

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radiation, and combination of both.[13] There have been no single arm trials utilizing single - agent therapy in the literature. Several combination chemotherapy regimens are active with doxorubicin, liposomal doxorubicin, dacarbazine, ifosfamide and methotrexate plus Vinca alkaloid producing significant responses, according to RECIST, ranging from 50% to 80% (partial responses or stable disease for at least six months)[11]. One trial has showed that Gemcitabine+ docetaxel is more effective than Gemcitabine alone as second -line chemotherapy, but data are conflicting and toxicity is different.[15] Our patient responded well to radiation and docetaxel.

Aggressive fibromatosis often recur after surgery, particularly after marginal or intralesional excision. Distant hematogenous or lymph node metastasis has not been observed. Head and neck lesions are more aggressive than extra-abdominal fibromatosis elsewhere and are capable of massive destruction of adjacent bone, erosion of base of skull and occasional encroachment on the trachea, sometimes with fatal outcome [2, 14]

Chemotherapy with cytotoxic agents represents an appropriate choice for patients with unresectable, rapidly and aggressively growing tumors or for patients who are highly symptomatic and /or with life threatening desmoid tumor. Preferable regimen includes pegylated liposomal doxorubicin because of lower toxicity rate. The main concern about chemotherapy is cumulative toxicity partly due to the pharmacological characteristics of the agents and partly due to the prolonged period of the application for obtaining treatment response. [12]

In the future, improved understanding of the biology and the biologic diversity of desmoid tumor will likely lead to additional systemic therapy options, and, most importantly, will lead to the identification of patients most likely to benefit from systemic therapies. A well-coordinated, multidisciplinary approach involving the inputs from surgical and non-surgical specialists is needed to develop an individualized treatment strategy appropriate for each patient.

CONCLUSION: Due to rarity of the disease prospective trials and correlative laboratory investigation will require multicentre cooperation. Referral of these patients to expert teams is the only way to gain sufficiently large experience in a prospective way. Such efforts will enable to identify clinical or molecular criteria by which patients can be selected for each single therapy, long term observation, or intense multimodality approaches with the vision of keeping patients with disease alive and preserving their quality of life.

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doi:10.1093/aaonc/mdq209

Figure1: Fibromatosis of face

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Figure 2: Showing fascicular arrangement of spindle shaped cells

Figure 3: Showing spindle-shaped cells with bland features

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Date of Submission: 15/07/2013.
Date of Peer Review: 18/07/2013.
Date of Acceptance: 10/09/2013.
Date of Publishing: 16/09/2013