Case Report

Fibromatosis of the maxillary sinus and muscles of mastication; a case report.

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ABSTRACT

Fibromatosis is a rare and locally aggressive benign tumour arising from the musculoaponeurotic structures of the whole body. Although it may occur in oral and maxillofacial region, is extremely rare in the paranasal sinus and involving the muscles of mastication. A 26 years old Bangladeshi male presented with severe trismus will be reported here to focus attention on this rare disease. Post operative histopathology confirmed the disease as fibromatosis.

Introduction:

Fibromatosis is a locally aggressive, benign tumour arising from the musculoaponeurotic structures[1]. The lesion is rare in head neck region. Between 1968 to 2008, only 179 cases were documented [2].Among the documented cases, only few cases were in paranasal sinuses.In the head and neck region, the lesion is serious because of its local aggressiveness and high rate of recurrences in a relatively restricted area[3].Here, we will report a case of fibromatosis in a 26 years old Bangladeshimale involving the left maxillary sinusand all muscles of mastication on the left side.

The purpose of this study is to focus attention on this rare disorder and also its rare presentationby reporting one case.

The literature review showed that the tumour has been described under a variety of synonyms[4,6]. Based on the site (such as upper and lower compartment), bone involvement and presence of hormone receptors, Kruse et al. proposed a classification [5]. Depending upon its anatomical sites, it may be associated with asymptomatic swelling, deformity of face, sinus congestion, halitosis, trismus, airway obstruction, dysphasia, proptosis, epistaxis[7,8,9, 10,] . Wide local excision is the treatment of choice;however, in the head and neck region, preservation of vital structuresmay impede their objectives[11]. In casewith residual diseaseor where surgery may significantly impair the functional capabilities, adjuvant radiotherapy or and chemotherapy achieves long term control[6, 12-17].

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Case Report:

A 26 years Bangladeshi male patient was referred to the oral and maxillofacial surgery department of Dhaka Dental College Hospital with the complaints of inability to open mouth and pain on his left cheek in an attempt of mastication for the last six months.According to the patient's statement, he was relatively well six months back, and then he developed gradual limitation of mouth opening. He had no other complaints. He had no history of trauma on that region. He denied any positive findings related to his disease in his family members.Drug history revealed nothing significant.

He was ill looking and mild anaemic, all other parameters of general examination was within normal limit. Extra oral examination showed that a 2cm x 2cm depression present on his left cheek just anterior to the masseter (Fig: 1, 2). Overlying skin colour was normal. The area was stiff, fixed to the underlying bone but the skin was free. Mild tenderness was present over the masseter. No Anaesthesia or paraesthesia was found. Both Tempero-mandibular Jointmovementswere present. Multiple lymph nodes were palpable on left level IB and level II.The lymph nodes were firm, free from skin but were attached to the underlying structures.His left nostril was patent, no obstruction or growth was evident. His inter incisal opening was o mm (Fig: 3). on 5^{th} and 6^{th} days of admission, he noticed bleeding through his left nose.

X-ray paranasal sinus (OM View) was orderedwhich showed complete haziness of the whole left maxillary sinus (Fig: 5). OPG showed nothing significant, both TMJ revealed normal (Fig: 4).A CT scan of the oral and Maxillofacial region showed a sizable, in homogenously and mildly enhanced infiltrating soft tissue mass inthe left maxillary sinus with lateral extension to the left infra temporal fossa through the destructed part of the left maxillary antrum. It involves all the muscles of mastication (Fig: 6, 7). All the walls of the antrum were eroded (Fig: 8). Left alveolar process of the maxilla was destructed. No abnormality wasseen on tempero mandibular joints. Mucosal thickening was also noticed on left sided ethmoidal sinus. CBC showed ESR 102 mm in first hour. X-ray chest and ultrasonography of whole abdomen showed nothing abnormal. Incisional biopsy was taken through the labial antrostomy approach but the reports revealed respiratory epithelium.

We performed tracheostomy under local anaesthesia, and thenleft sided radical neck dissection followed by maxillectomy on left side without exenteration of left orbit followed by left sided partial mandibullectomy. Per operative findings was peculiar to us. The lesion was seemed to have a special affinity to the muscles of mastication. The excised muscles were dark grayish in colour. The muscles were too stiff to separate from the ramus. The lesion also involved the temporalis muscle and extended upto the lower part of the temporal fossa. The most laborious part of the operation was to separate the condyle and coronoid process from the lateral pterygoid and temporalis muscle. Mouth opening was possible only after excision of all the muscles and removal of the resected mandible.

Post operative histopathology showed proliferation of fibrous tissue with abundant collagen. The muscles showed degenerative and atropic change. The fibrosis was also present within the inter trabecular space of the bony tissue. No evidence of malignancy was found. Lymph nodes showed reactive hyperplasia. The histopathologist confirmed the case as fibromatosis.The patient was clinically well after 2 months. As the margin was positive, we referred the patient for radiotherapy

Fig -1: Frontal view

Fig-2: Depression on left cheek

Fig-3: Restricted mouth opening

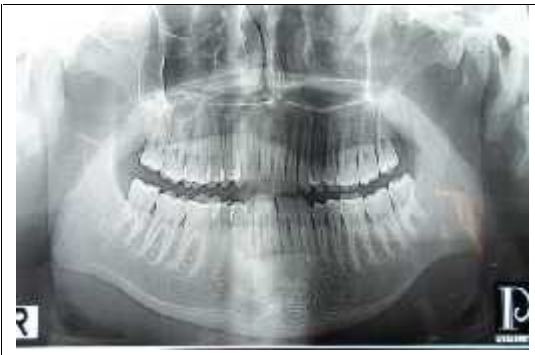

Fig-4: TMJ are normal

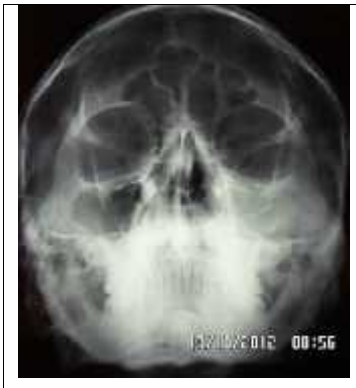

Fig-5: Hazy left maxillary sinus

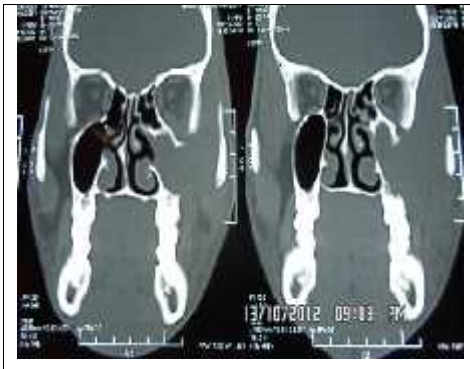

Fig-6: Destruction of left maxillary sinus and extension of the lesion

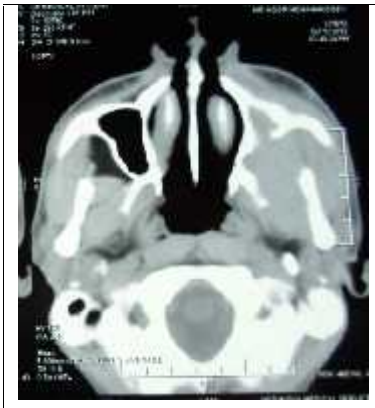

Fig-7: Invovement of left maxillary sinus and all muscles of mastication on left side.

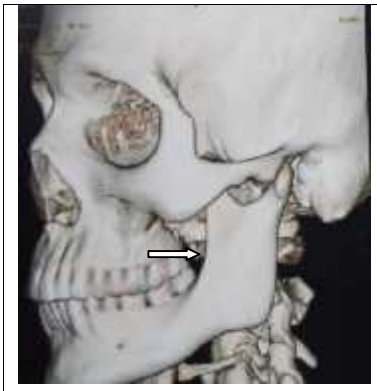

Fig-8: Destruction of buccal cortex

Discussion:

Fibromatosis is a rare benign tumour which arises from the musculoaponeurotic structures throughout the body. In the oral and maxillofacial region, It has been described under a variety of synonyms, including 'extra articular desmoids', 'desmoids tumours', 'grade-1 fibrosarcomas','non metastasizing fibrosarcoma'and 'aggressive fibromatosis'.Fibromatosis remain a fibrous tissue proliferation with an intermediate biologic behavior between a benign fibroma and fibrosarcoma[6]. That is, like fibrosarcoma, they exhibit destructive infiltrative growth and frequently recur, but like fibromas, they do not metastasize[6].

Fibromatosis is uncommon in head and neck region[6.] Masson and Soute[18]cited 12% of 284 cases from all locations of the body while Das Gupta et al.[19] reported an incidence of 11.1% of 72 cases of extra abdominal fibromatosis. The supraclavicular fossa is the most common site for head and neck fibromatosis (40%-85%), followed by face (about 25%)[20]. Other sites were also reported to be affected by fibromatosis, these are mandible[24], nasopharyngeal tract[8], larynx[21], tongue[22], orbit[9], all the paranasal sinuses[23], infratemporal space and para pharyngeal space[10]. Those that occur in the oral and maxillofacial areaare somewhat unique compare to those that occur in the most common locations around the shoulder girdle and trunk. The oral and maxillofacial locations (mostly the mandible, maxilla and mastoid area) show a younger peak age range (5 to 20 years compared to 35 years), more infiltrative and faster growth and a much greater propensity to invade underlying bone or to arise seemingly from bone[6].But Das Gupta et al.[19] found most cases appear between third and fourth decades. Conley[20] in his study showed the age range is between from new born to 70 years. Our patient was 26 years old; it is within the most prevalent age group.

Fibromatosis were found to be more common in female with female and male ratio is in between 3:2[18] to 2:1[19], although both sexes were equally in other reports. Min, R. et al.[25]found male are more affected, the ratio is 3:2.

The exact aetiology of fibromatosis is still unknown. However, trauma, endocrine or genetic factorsare probable aetiological factors, but still no satisfactory explanations present behind them[6]. Fibromatosis in the head and neck region recur from 20% to 70% of cases[11, 27, 28], whether it is due to a more aggressive nature of the tumour or to a technically more difficult excision in this region are unclear[29]. The length of the recurrence is from two months to eleven years[8, 30]. But fibromatosis almost never or in very few cases turn into fibrosarcoma. Reitomo[31] analyzed 1500 cases of fibrosarcoma, a transformation to sarcoma was reported in only two occasions. Min, R et al. [25] found malignant changes in only six cases out of 20 cases, that is 65% of which had abnormally high blood loss of serum alkaline phosphatase.

The usual presentation of fibromatosis is slowly growing asymptomatic swelling[8];the mass is often fixed with the underlying deep muscles or bony structures but not to the skin. This tendency often results trismus.Other features may be nasal obstruction, epistaxis, facial pain[18], dyspnoea, stridor[21], and proptosis[9]depending upon the anatomical sites of the lesion. Pain and paraesthesia is uncommon [6]. The complaints of our patient was trismus and mild pain over the left masseter in an attempt of mastication, he had also history of epistaxis for two episodes; he had no history of paraesthesia or significant pain. So, In general, the clinical features were similar to the other reported cases.

Fibromatosis is grayish white, firm or rubbery in consistency. Its long axis is usually oriented in the direction of muscle bundles in which it is found. It is not encapsulated and the border is ill defined due to its tendency to infiltrate the surrounding structures. It can encase vascular and neural structures without apparent invasion which may account for lack of pain clinically[5, 23]. In reported case, per operatively, it was evident that the lesion had a special affinity to the muscles. The lesion was poorly localized and not encapsulated. The muscles were dark grayish on their cut surface.

Radiographs frequently showpoorly demarcated, irregular bone destruction. If the lesion is located at the surface of the jaws, it may show an irregular resorption of the adjacent cortex only. If the lesion is central, it will usually show a destructive pattern in all directions[6]. Extensive bone destruction of all the walls of the maxillary antrum, involvement of the all muscles of mastication on the left side suggested us an aggressive lesion.

The diagnosis requires a deep incisional biopsy in the centre of the mass. The biopsy should extend to the bone and include periosteum to assess the infiltrative growth pattern. A biopsy at the edge of the tumour will induce scar tissue that is histologically similar to the tumour, thereby confusing the margin at the time of excision[6]. In contrast, Fu and Perzin[8]suggestedtaking biopsy from the margin. They thought that a biopsy from the centre of such lesion may be mistaken for scar tissue. In some cases, only the behavior of the lesion (Infiltration into the adjacent tissue, progressive increase in size etc.) may indicate its true nature. In our case,the incisional biopsy was taken through the labial antrostomy which often leads misdiagnosis. It should be taken through the lateral wall of the nose. We could not take any representative tissue. Incisional biopsy showed it was respiratory epithelium.

Fibromatosis is composed of fibrous tissue which is highly cellular, cells are elongated, splender spindle cells with abundant collagen arranged in broad elongated fascicles. Mitosis is rare and typical. The tumour shows microscopic infiltration into the adjacent tissue particularly striated muscle fibers with the formation of multinucleated giant cells and lymphoid infiltrates[23]. In our case, post operative histopathology showed proliferation of fibrous tissue with abundant collagen along with dense infiltration of acute and chronic inflammatory cells. The skeletal muscles showed degenerative and atropic change. The fibrosis was also present within the inter trabecular space of the bony tissue. No evidence of malignancy was found. Lymph nodes showed reactive hyperplasia. Thus, the histopathology report confirmed fibromatosis.

The extension of the lesion, affinity to the muscles, bone destructive and infiltrative capabilities, the clinical presentation of trismus and epistaxis suggested us about presence of an aggressive lesion. The extensive fibrosis of the muscles in per operative findings, no anaesthesia or paraesthesia, no lymph nodes involvements, no evidence of metastasis,absence of any malignant features and post operative histopathology confirmed us the lesion to be fibromatosis.

The main treatment modality is the wide surgical excision of the lesion and any involved bone with 1 to 1.5 cm. healthy margin with the frozen section biopsy at the time of surgery. Treatment of the lymph nodes is not necessary[6]. But in the head and region, preservation of the vital structures and their function may impede these objectives[11].Therefore, a multi-modality treatment strategy is usually employed to control residual disease. Surgery combined with radiotherapy or and chemotherapy are being reported. Besides, anti estrogen (Tamoxifen) therapy, even NSAIDS (Sulindac) are being used. But their efficacy is not established.

In several reports, Radiotherapy alone (50 to 60 Gy), or combined with surgery in patients with positive margin gives long term controlin 70% to 80% of patientswith desmoids[12-17]. NSAIDS probably acts by inhibiting prostaglandins synthesis which impairs the proliferative capacity of tumour cells and the same time stimulates an immunological response[32]. An anti estrogen (Tamoxifen) therapy is reported on the ground that the speed of the growth of fibromatosis is regulated especially by the female sex hormones. But such treatments have not reached definite results yet[11, 29, 33, 35, 36]. Anti cancer chemotherapy has been reported to produce a partial or complete regression of fibromatosis[29].

Conclusions:

Fibromatosis is rare in maxillofacial region. But it is locally aggressive as it recurs more frequently.In most cases, the lesion is asymptomatic;the patients seek treatment when the lesion becomes a significant size and when the lesion involves the surrounding structures which make the surgery more difficult.Surgery is the treatment of choice with 1.5 cm. healthy margin with the frozen section biopsy. In case of residual disease radiotherapy should be advised. Long term follow up is necessary because re growth or recurrence could occur after a long period of stabilization.

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