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Case Report

Aggressive Fibromatosis in Lower Body of Mandible: A Rare Entity Report of a Case

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Abstract

The fibromatosis are a rare group of lesions that can infiltrate widely, replacing muscle, fat and bone with fibrous tissue of varying cellularity. They do not develop distant metastases, however, locally they show an aggressive and infiltrative behaviour. Clinically, aggressive fibromatosis manifests as a painless, firm, often rapidly enlarging mass, fixed to underlying bone or soft tissue. It is never encapsulated. Histologically, it is rich in collagen and fibroblastic cells that are devoid of hyperchromatic or atypical nuclei. Treatment is based on either excision of the mass, or radiotherapy and chemotherapy if the condition is inoperable. We describe a case of aggressive fibromatosis of the mandible in a young adult initially misdiagnosed as a case of some centrally originated pathology.

Keywords: *Fibromatosis; Fibromatosis Mandible; Desmoid Tumor*

Introduction

Aggressive fibromatosis (AF or desmoid tumour) is characterised by infiltrating growth of well-differentiated fibroblasts or myofibroblasts, and often recurs, in particular if the resection was incomplete, but it has no metastatic potential [1].

The most common sites of these lesions are the mesentery, the retroperitoneal space, and the abdominal wall, or in extraabdominal sites such as the trunk and the extremities [2]. Its frequency is reported to be between two and four cases per million per year, representing 0.003 - 0.1% of all tumours. AF occurs more often in women, especially between 25 and 35 years [3]. These neoplasms have been described for almost every part of the body, the most common sites being the shoulder, chest wall, back and thigh. AF is a rare neoplasm in the head and neck, constituting approximately 2% of all cases [4,5]. It believe trauma and hormonal changes have been reported to occur in conjunction with the tumour [6]. Genetic disorders such as the Gardner syndrome may also lead to desmoids [7]. Clinical differentiation of desmoids from malignant tumours may be difficult in view of the local invasive growth of the former. The treatment of desmoids is controversially discussed. Most authors suggest surgical radical resection [8-10], while others recommend radiotherapy. In case of unclear tumour margins or incomplete resection, radiotherapy after surgery is advocated [11,12]. Hence, surgical treatment was planned in the present.

Case Report

A 21-year-old adult was referred with a painless hard mild swelling over left body of the mandible. The patient was asymptomatic and his medical history was without any findings. Physical examination showed a firm mass around the left mandible involving the entire body. No mucosal surface ulceration was present, but corresponding extraoral swelling was evident (Figure 1). Radiographically, ill-defined radiolucency was visible over mandibular body with respect to molars, and bone loss in relation to alveolar crest associated with premolars (Figure 2), these findings were not substantial to ascertain any provisional diagnosis. Subsequently, patient underwent cone beam computed tomography scan (CBCT) for the respective area, which revealed a large portion of eroded buccal cortical plate region (Figure 3 and 4). Still in account of the uncertain diagnostic dilemma, an incisional biopsy was planned. On approaching the pathology through standard envelop incision, the abnormally high fibrotic nature of the overlying soft tissue alerted the surgeon of some rare pathological presence. Thereby, the biopsy sample was obtained from the over lying fibrotic soft tissue and also from the eroded bone region, which was sent for further histopathological examination. The histopathological examination revealed a mesenchymal tumor with spindle-shaped tumor cells growing in fascicles. The tumor showed areas of high cellularity, changing with less cellular areas with a myxoid background. Especially in the less cellular areas, a network of collagen fibers was obvious (Figure 5 and 6). Thus, the final diagnosis was aggressive fibromatosis as confirmed by the histopathological examination, was made. distinguished the lesion from fibrosarcoma. The postoperative course was uneventful. Eighteen months postoperatively there has been no evidence of recurrent disease.

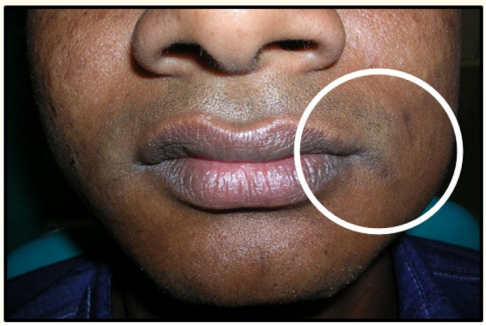

Figure 1: *Extra orally visible mild swelling over left mandible body region.*

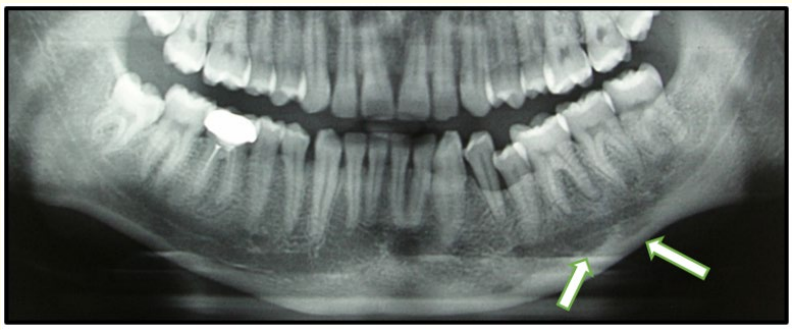

Figure 2: *Panoramic view showing certain inconclusive radiolucent patches.*

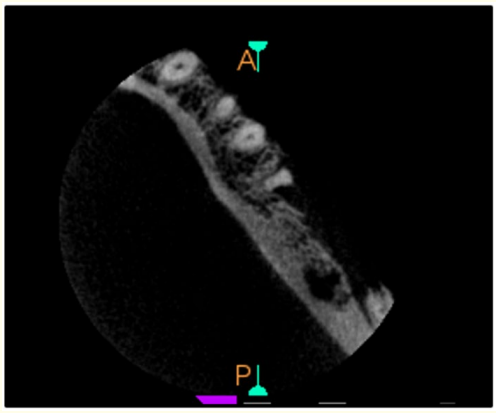

Figure 3: *Axial view of the mandible showing erosive changes over buccal cortical plate.*

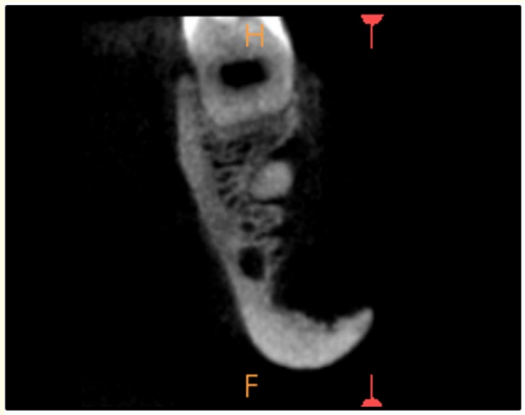

Figure 4: *Coronal view with visible destruction of buccal cortical bone and medullary bone seen.*

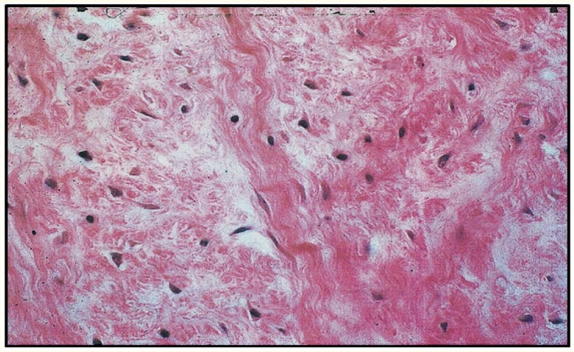

Figure 5: *Histological examination (haematoxylin and eosin, original magnification 200X) showed proliferation of uniform spindle cells in densely collagenized stroma. Cells and collagen fibers were arranged in sweeping bundle that intersected at various angles.*

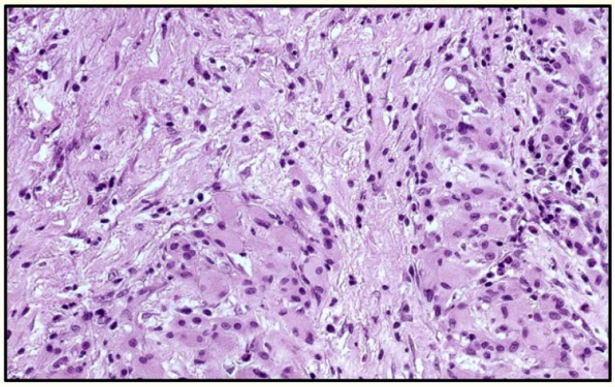

Figure 6: *Histological figure (HE-staining, 50X) showing connective tissue cells infiltrating musculature confirming diagnosis of AF.*

Discussion

Aggressive fibromatosis is a soft tissue neoplasm that presents as a mass within skeletal muscles or in the adjacent fascia or periosteum. Usually, the tumor is oriented with its long axis parallel to the muscle bundles in which it is found. Secondary bony involvement, presumably because of mass effect, can range from cortical remodeling to destructive lesions [14,15]. In present case there were Involvement over left body of the mandible, and there are case reports of infantile fibromatosis involving the submandibular space.

Clinical presentation of the lesion as a rapidly growing swelling was consistent with the reported cases of aggressive fibromatosis. No sexual predilection, most of the reported cases of aggressive fibromatosis of the mandible were young females [9,16]. In the present case, lesion was mainly located in buccal and body of mandible.

Radiologically bony eroded buccal cortical plate was observed in buccal aspect of the alveolar process away from the main tumor mass. Bony destruction could be explained by periosteal extension of the lesion. In study of Roychoudhury., *et al*. [16] and Shuker., *et al*. [17] bone erosion and periosteal bone deposition are reported in only one study, in which there was erosion of the mandibular angle and body of mandible with Destruction of cortical bone with adjacent soft tissue mass were evident.

Histologically they consist of well-differentiated fibroblast, fibrocytes and myofibroblasts with a collagenous to myxoid cells. Atypical mitotic figures or anaplastic cells are most commonly seen [13]. In present case there were mesenchymal tumor with spindle-shaped cells and areas of high cellularity, changing with less cellular areas with a myxoid background were seen.

Surgical excision and radiotherapy are the two principal modalities of treatment [6]. A complete en-bloc resection is the treatment of choice if possible. In head and neck, unless the lesion is small, complete resection with wide margins is technically difficult due to the complex anatomy, presence of important structures and the chances of increased morbidity after excision. In present case the lesion were small and not extensive surgical excision was done with one year of follow-up. Other treatment modalities described are of use of hormonal therapy and chemotherapy [13,18]. There are limited studies on the effectiveness of these modalities [13].

Fibromatosis is benign histologically, but local aggressiveness and high recurrence rate (20% - 70%) may result in associated morbidity and mortality. Recurrence is more commonly reported in the first year, but there are reports of lesion recurrence even 5 years after treatment. A duration of at least 3 years is recommended for follow-up in the literature [13].

Conclusion

Aggressive fibromatosis is a histologically benign soft tissue tumor with nonspecific clinical features, Radiographic appearance of bony involvement, secondary to mass effect, ranges from cortical bone erosion. Histopathological examination plays an important role for definitive diagnosis. Surgical removal with large rim of normal-appearing tissue is advised. Because of local aggressiveness and high recurrence rate, long-term follow-up is necessary.

Sources of Support

Nil.

Conflicts of Interest

Nil.

Bibliography

1. Bertrand JC., *et al*. "Maxillary and mandibular desmoid fibromas (author's transl)". *Revue de Stomatologie, de Chirurgie Maxillo-Faciale* 82.2 (1981): 127-131.
2. Loccufier A., *et al*. "Gardner syndrome and desmoid tumors". *Acta Chirurgica Belgica* 93.5 (1993): 230-232.
3. Mendez-Fernandez MA and Gard DA. "The desmoid tumor: benign neoplasm, not a benign disease". *Plastic and Reconstructive Surgery* 87.5 (1991): 956-960.
4. Leithner A., *et al*. "Treatment of extra-abdominal desmoid tumors with interferon with or without tretinoin". *Journal of Surgical Oncology* 73.1 (2000): 21-25.
5. Enzinger Fm and Weiss Sw. "Fibromatoses". In: E Nzinger FM, Weiss SW, eds: Soft Tissue Tumors. 3rd edition. St. Louis: Mosby (1995): 201-229.
6. Batiani J., *et al*. "Desmoid tumors in adults: the role of radiotherapy in their management". *American Journal of Surgery* 155.6 (1988): 754-760.
7. Fasching M., *et al*. "Desmoid tumors of the head and neck". *American Journal of Surgery* 156.4 (1988): 327-331.
8. Abdelkader M., *et al*. "Aggressive fibromatosis of the head and neck". *Journal of Laryngology and Otology* 115.10 (2001): 772-776.
9. De Santis D. "Fibromatosis of the mandible: case report and review of previous publications". *British Journal of Oral and Maxillofacial Surgery* 36.5 (1998): 384-388.
10. Hoos A., *et al*. "Desmoid tumors of the head and neck". *Head and Neck* 22.8 (2000): 814-821.
11. Mirra M., *et al*. "Aggressive fibromatosis of the larynx: report of a new case in an adult patient and review of the literature". *Pathology - Research and Practice* 197.1 (2001): 51-55.
12. Nuyttens J., *et al*. "Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: a comparative study of 22 articles". *Cancer* 88.7 (2000): 1517-1523.
13. Gupta A., *et al*. "Aggressive fibromatosis of head and neck in a child". *Indian Journal of Otolaryngology and Head and Neck Surgery* 67.1 (2015): 154-157.
14. Alam M., *et al*. "Aggressive fibromatosis in the submandibular region of a child". *Clinical Case Reports* 10.4 (2022): e05761.

15. Lewin JS and Lavertu P. "Aggressive fibromatosis of the prevertebral and retropharyngeal spaces: MR and CT characteristics". *American Journal of Neuroradiology* 16.4 (1995): 897-900.

16. Roychoudhury A., *et al*. "Infantile desmoid fibromatosis of the submandibular region". *Journal of Oral and Maxillofacial Surgery* 60.10 (2002): 1198-1202.

17. Shuker ST and Jabro MH. "A case report of submandibular aggressive fibromatosis in a 16-month-old infant with 8-year postsurgical follow-up". *Journal of Oral and Maxillofacial Surgery* 61.8 (2003): 951-954.

18. Wang CP., *et al*. "Desmoid tumor of the head and neck". *Head and Neck* 28.11 (2006): 1008-1013.

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