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Case Report Open Access

Desmoid Fibromatosis of the Maxilla: A Rare Case Report

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Abstract

Fibromatosis is an increase in the amount of connective tissue due to hyperplasia in connective tissue cells. Desmoid tumor, also called aggressive fibromatosis, is locally aggressive and does not show metastasis. If this tumor is not treated, it continues to grow frequently. Recurrence rate is high after treatment. Due to its aggressive nature, wide resection is recommended. In our case, a 27-year-old male patient had a desmoid tumor in the left upper premolar region. The patient was referred to the surgical clinic and the tumor was removed by wide excision.

Keywords: Fibromatosis; Connective tissue; Hyperplasia

Introduction

Fibromatosis are proliferations of highly differentiated fibrous tissue from musculoaponeurotic formations [1]. Varieties of this soft tissue tumor is known as desmoid tumors (DF), aggressive fibromatosis, or desmoid-type fibrosarcoma. These proliferations are commonly seen in abdominal region, and the occurrence of these tumors in maxillofacial region is a rare entity [2]. Due to locally infiltrative growth and high recurrence rate of DF, it is considered as an aggressive tumor [3]. DF occurs 2 to 4 per 1 million with a female/male ratio of 3:1 [4]. While the incidence of DF in children peaks at about 8 years of age, in adults, peaks in the third and fourth decades [5]. While 25% of all desmoid tumors occur in children under 15 years of age, 12% and 15% of all desmoid tumors are seen in head and neck region [6]. Head and neck lesions are more aggressive than extra-abdominal fibromatosis [7]. Although exact etiology of DF is still unknown, trauma, genetic mutations and hormonal differences are considered as the predisposing factors of this disorder. DF shows low cellularity and contains extracellular matrix [8]. Although it can arise to great sizes and have local aggressive character, malignant transformation is observed less frequently. In this case report, an extraordinary desmoid fibromatosis with different histological features was identified.

Case Report

A 27-year-old man was admitted to the clinic for a routine dental examination. The patient had no history of systemic disease and no similar family findings after genetic examination. On panoramic radiograph (Figure 1), a unilocular radiolucency was detected in the upper left premolar region and root resorption was noticed in these teeth (Figure 2). The patient showed no swelling, discoloration, tingling, or other signs of clinical or radiological findings. The extraoral examination of the patient did not show any cervical or submandibular lymphadenopathy. There was no significant facial or cervical asymmetry. The patient was consulted to the oral and maxillofacial surgery department after clinical and radiological examination. The surgical treatment plan was the enucleation of the lesion. Enucleation limits in the operation area. Desmoid fibromatosis findings were detected in the histopathologic examination was preferred to create significant (Figure 3). In pathological records immature odontogenic epithelial remnants and myxoid matrix in examined mass were remarkable differences in comparison to other known properties of DF.

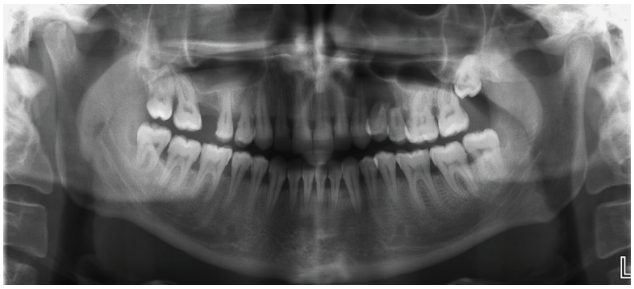

Figure 1: A unilocular radiolucency in the upper left premolar region.

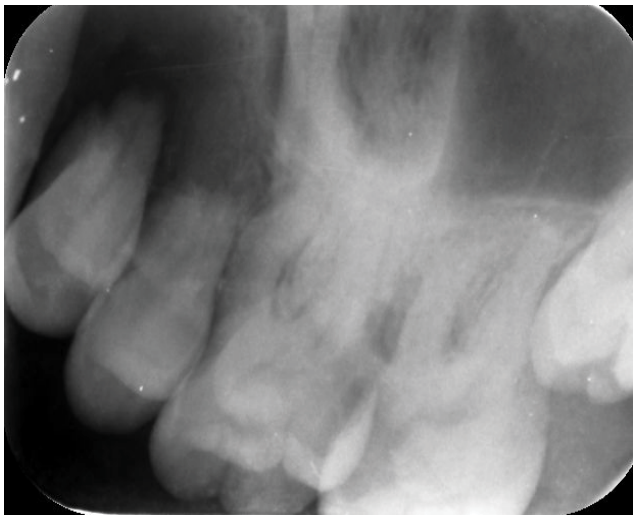

Figure 2: Root resorption in premolar teeth.

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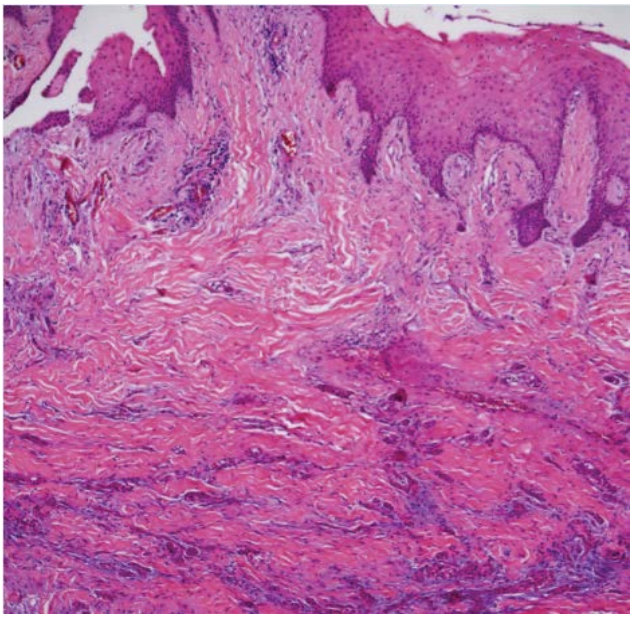

Figure 3: In the mucosal tissue sample with preserved epithelium, increased fibrous connective tissue is seen in the subepithelial area. There are myxoid changes and large amounts of immature odontogenic epithelial residues in connective tissue.

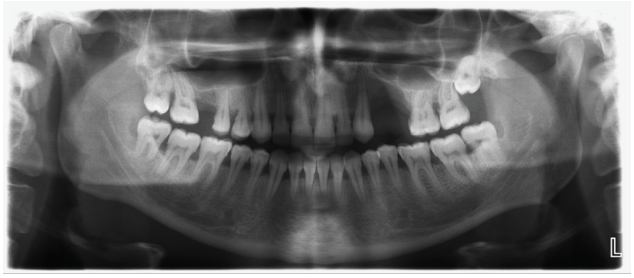

Figure 4: Panoramic view of the operation area one year later.

Discussion

Desmoid tumor was first detected by John Macfarlane in 1832 [9]. According to the WHO classification of soft tissue tumors, desmoid tumors are called as desmoid-type fibromatosis [10]. Desmoid fibromatosis is rare in the maxillofacial area [11]. Although it have a non-metastatic character, desmoid tumors have been classified by WHO as intermediate grade tumors due to aggressive progression and repetitive at very frequent intervals [3]. Only 12% of the extra abdominal fibromatosis develop in the head and neck region [12]. According to Ruan Min et al.'s research, desmoids-type is an extremely high ratio of suspected malignant change, mainly high-grade fibrosarcoma that was found in postoperative paraffinic tissue, including either the first operation or the subsequent operation owing to recurrence. It is a general opinion that desmoid-type fibromatosis is a rare in the oral and maxillofacial region, non-metastatic neoplasm and malignant transformation is very uncommon from the literature review [7-13]. DF is a difficult pathology to treat and control. Surgery, chemotherapy, radiotherapy and hormone therapy are used in the treatment [14,15]. Radiotherapy alone can provide local control up to 75-80% in the presence of large masses or in cases where the surgical margin cannot be maintained microscopically [16]. The primary treatment of the lesion is surgical excision, of which margins end in healthy tissue [17]. The case underwent a surgical excision as suggested. In many cases, it was confirmed as benign desmoid-type fibromatosis by histopathological examination with the appearance of a fibroblastic spindle-celled tumor producing abundant collagen fibers with the cells arranged in interlacing bundles and the tumor tissue was always merged with a small amount of muscle, gland, and bone [17]. In mucosal tissue sample, which has intact epithelial surface, increased fibrous connective tissue was observed in subepithelial area, the histopathological mass included different cells such as immature odontogenic epithelium and myxoid matrix. This case can't be seen like a classic gingival fibromatosis. It was thought that it was fibromatosis (desmoid fibroma) of the bone. The lesion was a progressive lesion under the mucosa. In the histological picture, fibromatosis was seen to progress under the mucosa epithelium. What is interesting is that, unlike classical gingival fibromatosis and desmoid, immature odontogenic epithelial residues are abundant and contain plenty of myxoid matrix.

Conclusion

As a result, desmoid tumor is a benign, local aggressive tumor and does not show metastasis. The primary treatment is surgery with negative margins (Figure 4). Recurrence rate is high after surgical treatment. Therefore, physicians should be aware of desmoid tumor and follow the patients for a long time after treatment.

References

1. Watzinger F, Turhani D, Wutzl A, Fock N, Sinko K, et al. (2005). Aggressive fibromatosis of the mandible: a case report. Int J Oral Maxillofac Surg 34: 211-213.
2. Eagel BA, Zentler MP, Smith IE (1989) Mesenteric desmoid tumours in Gardner's syndrome - review of medical treatments. Postgrad Med J 65: 497-501.
3. Goldblum J, Fletcher JA (2002) Desmoid-type fibromatosis. In: Fletcher CDM, Unni KK, Mertens F, editors. World Health Organization classification of tumors: pathology and genetics of tumors of soft tissue and bone. Lyon: IARC Press 83-84.
4. Georgiades C, Vallianou N, Argyrakos T, Aristodimou A, Kolovelonis G, et al. (2012) An unusual case of desmoid tumour presenting as haemorrhagic shock. Ann R Coll Surg Engl 94: e81-e82.
5. Chen CB, Chiou YY, Chen CH, Chou YH, Chiang JH, et al. (2010) Sonographic and computed tomography findings of intra-abdominal desmoid tumor. J Chin Med Assoc 73: 393-395.
6. Stout AP (1954) Juvenile Fibromatoses. Cancer 7: 953-978.
7. Masson JK, Soule EH (1966) Desmoid tumours of the head and neck. Am J Surg 112: 615-622.
8. Allen PW (1977) The fibromatoses: a clinicopathological classification based on 140 cases. Am J Surg Pathol 1: 305-311.
9. Dalen BP, Bergh PM, Gunterberg BU (2003) Desmoid tumors: a clinical review of 30 patients with morethan 20 years' follow-up. Acta Orthop Scand 74: 455-459.
10. Jo VY, Fletcher CD (2014) WHO classification of soft tissue tumours: an update based on the 2013 (4th) edition. Pathology 46: 95-104.
11. Angiero F, Benedicenti S, Stefan M (2008) Fibromatosis of the head and neck: Morphological, immunohistochemical and clinical features. Anticancer Res 28: 1725-1732.
12. Fowler CB, Hartman KS, Brannon RB (1994) Fibromatosis of the oral and paraoral region. Oral Surg Oral Med Oral Pathol 77: 373-386.
13. Nuyttens JJ, Rust PF, Thomas CR Jr, Turrisi AT 3rd (2000) Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors:A comparative review of 22 articles. Cancer 88:1517-1523.
14. Mullen JT, Delaney TF, Kobayashi WK, Szymonifka J, Yeap BY, et al. (2012)

Desmoid tumor: analysis of prognostic factors and outcomes in a surgical series. Ann Surg Oncol 19: 4028-4035.

15. Mendenhall WM, Zlotecki RA, Morris CG, Hochwald SN, Scarborough MT (2005) Aggressive fibromatosis. Am J Clin Oncol 28: 211-215.

16. Faulkner LB, Hajdu SI, Kher U, La Quaglia M, Exelby PR, et al. (1995) Pediatric desmoid tumor: retrospective analysis of 63 cases. J Clin Oncol 13: 2813.

17. Magro G, Emmanuele C, Lopes M, Vallone G, Greco P (2008) Solitary fibrous tumour of the kidney with sarcomatous overgrowth. APMIS 116: 1020-1025.