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An 8-Year-Old Male with Juvenile Aggressive Fibromatosis of The Lower Jaw: A Case Study

[1]Goklani Ankita Amar, Government Dental College and Hospital, Aurangabad

[2]Divya Sharma, Assistant Professor, Department of Oral and Maxillofacial Surgery, faculty of Dental Sciences, Manav Rachna Dental College, MRIIRS, Faridabad

[3]Yajas Kumar, Assistant Professor, Department of Oral and Maxillofacial Surgery, faculty of Dental Sciences, Manav Rachna Dental College, MRIIRS, Faridabad

[4]Ratna Priya, Department of Public Health Dentistry, KD Dental College and Hospital, Mathura

[5]Azmi Malik, Senior Lecturer, Department of Oral Medicine and Radiology, Azamgarh Dental College

Corresponding Author: Goklani Ankita Amar, Government Dental College and Hospital, Aurangabad.

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Abstract

A benign spindle cell tumour known as aggressive fibromatosis is also referred to as desmoid tumour, non-metastasizing fibrosarcoma, and grade I fibrosarcoma in the literature. Here, we describe a case of juvenile aggressive fibromatosis in an 8-year-old boy who had been experiencing loose teeth caused by the resorption of the alveolar and cortical bones for three weeks before to his presentation. Microscopically, bundles of oval cells with fibro-collagenous connective tissue were seen. After conservative surgery to keep the permanent teeth, no recurrence was discovered after eight months of follow-up.

Keywords: Alveolar, Cortical Bones, Panoramic Scan

Introduction

A benign spindle cell tumour known as aggressive fibromatosis is also known as desmoid tumour, grade I fibrosarcoma, and non-metastatic fibrosarcoma in the literature [1,2]. Non-metastasizing fibrosarcoma and grade I fibrosarcoma were discontinued as terms because they both refer to sarcomas with the potential to metastasize [2]. A widely used terminology in the literature for aggressive fibromatosis was desmoid tumor, coined by Mueller in 1838 to emphasize the band like or tendon like consistency of the lesion [2]. Fibromatosis includes an extensive group of benign fibrous connective tissue proliferations with similar microscopic appearance, the biological behaviour of which is intermediate between benign fibrous lesions

and fibrosarcoma [2]. Aggressive fibromatosis of head and neck is uncommon and lesions involving oral cavity and jaw bones are encountered rarely [3]. It is characterized by a locally infiltrative growth potential without metastasis and a remarkably high tendency to recur [3].

Case Review

An 8-year-old child complained of soreness and missing teeth on the left side of the lower jaw for three weeks when he visited the department of oral and maxillofacial surgery at the college of dental sciences, Manav Rachna Dental College, MRIIRS, Faridabad. No previous experience of night sweats, weight loss, or discomfort fever. The face of the patient was found to be symmetrical and had no palpable lymph nodes. On the left side of the lower jaw, around the A, B, C, D, and E deciduous teeth, there was intraoral enlargement of the buccal and lingual cortices with grade 2 motilities of the A, B, C, and D teeth and loss of tooth number D. There was no occlusal constraint and no ulcers on the overlaying mucosa.

Permanent successors were found within the tumour and were pushed downward, and the lower border of the mandible was unharmed on the radiographic image (Figures 1 and 2). This radiolucency extended from the first deciduous incisor to the second deciduous molar in the lower left side of the jaw.

There were two possible diagnoses: dentigerous or periapical cysts. A biopsy was performed, and the results indicated a benign spindle cell lesion. The biopsy showed interlacing bundles of oval cells, adjacent fibro-collagenous connective tissues, reactive bone trabeculae, and no epithelial lining, granuloma, or malignancy. Desman, SMA, CD68, CD34, and S100 were found to be negative, and results that were compatible with fibromatosis were found, therefore additional immunohistochemistry was advised for confirmation and to rule out alternative options.

The treatment strategy was created after having to go over it with the parents. The young kid had conservative surgical curettage and excision of loose deciduous teeth in order to protect the permanent teeth. The patient underwent routine clinical and radiographic follow-up, and a prosthesis was created to replace the extracted teeth and cover the bone defect. The panoramic scan at 8 months after surgery did not reveal any signs of recurrence. Aside from the left lower canine, the permanent teeth in the surgical area appeared to erupt normally (Figure 3). To ensure there is no recurrence and that the permanent teeth emerge normally, the patient is continuously monitored.

Figure 1: Pre operative radiograph.

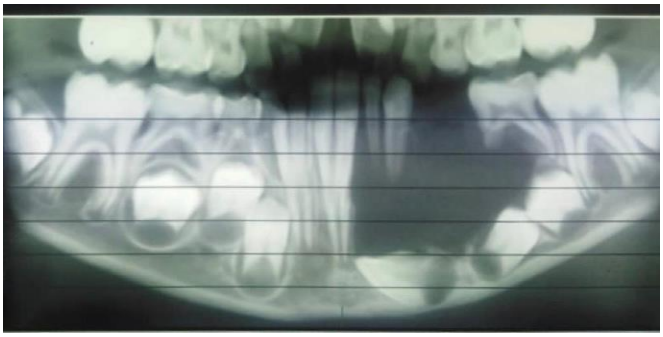


Figure 2: Pre-operative radiograph

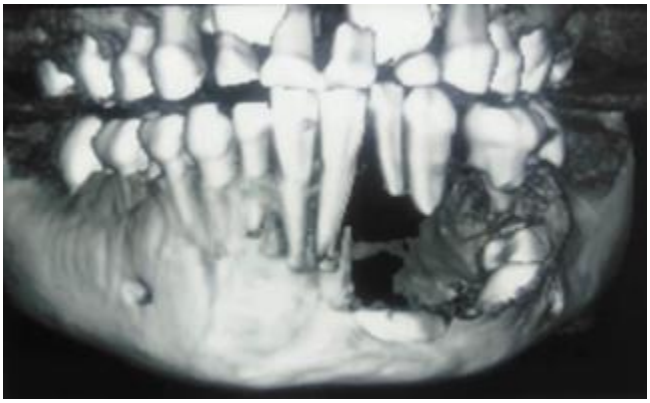
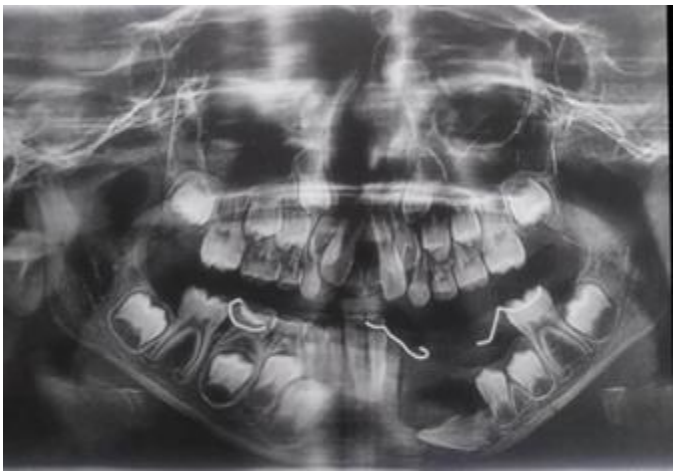


Figure 3: 8 months post-operative radiograph



Discussion

Fibromatosis is a benign tumor, but it is presented as aggressive, and it is etiology and pathogenesis are unclear yet. As noted earlier, fibromatosis has been classified into two types, superficial and deep. The deep type can then be further classified into abdominal or extra-abdominal fibromatosis [4]. Juvenile fibromatosis is more prevalent in patients under the age of 8; similarly to our patient [5]. The reported average age is 9 years, with a range of 8 months to 63 years. Between the ages of 5 and 15 years, 62% of head and neck occurrences occurred, while 21% occurred in children below the age of 5. Studies also revealed a preference for men [6].

Although association of aggressive fibromatosis and Gardner syndrome and familial adenomatous polyposis has been postulated in literature, our case occurred as sporadic condition [7]. The deep variant of fibromatosis which is the most common in the head and neck, occurs in the mandible more common especially in the posterior region specifically the ramous and the angle [8] and similar to this occurrence the presented case had a Mandibular lesion but it occurred in a more anterior region involving the left anterior mandible and part of the left body. In agreement with the results we obtained, which show a unilocular radiolucent lesion with ill-defined diffuse margins on the panoramic view, while the 3 D Con beam CT scan clearly demonstrated a defect in the buccal and lingual cortexes, a plain image of the affected bone displays a well-defined radiolucent lesion that appears as unilocular or multilocular with partially sharp or diffuse border without sclerotic margins [9]. A frequent observation that supports our case is floating teeth without alveolar bone support associated with root resorption. It is preferable to order an MRI to see the full extent of the tumour and to use for surgical planning in order to make a precise assessment of the tumour, which primarily affects soft tissue.[10] Radiographic differential diagnosis of aggressive fibromatosis includes multiple pathological entities which involve ameloblastoma, odontogenic fibroma, aneury small bone cyst, hemangioma, fibrosarcoma, and malignant histocytoma [11], due to these varieties we perfumed the excision biopsy which revealed the classical findings of extrabdominal and desmoid fibromatosis showing, proliferating fibroblast with little mitotic activity of oval cells in fibrocollagenous stromal, the term fibromatosis is indicating well-differentiated fibroblasts and my fibroblasts that develop in the soft tissue [1]. To confirm the diagnosis some studies adopted the concept of using B-catenin marker which is found to be positive in 90% of the histopathological specimen of the cases of fibromatosis. In our case we requested immunohistochemistry for verification and rule out of other possibilities and results were negative for desman, SMA, CD68, CD34, S100 and results consistent with fibromatosis, Those with aggressive fibromatosis usually undergo resection of the lesion with a free margin if it does not cause any impairment or morbidity, but if it causes any impairment or morbidity then the best treatment is curettage plus chemotherapy for patients under 18 years of age, and radiotherapy for above 18-years-old patient [12].

Several authors advised undergoing "segmental resection" or "additional curettage and peripheral osteotomy" in the event of recurrence [13]. Due to the patient's young age and the desire to protect the permanent successor teeth and allow them to fully erupt, we chose to use curettage in this case. As a result, we chose to closely monitor the patient rather than administer adjuvant treatment. Eight months following surgery, a panoramic scan showed impressive bone resorption and an improvement in the line of eruption for the two left lower premolars, but the left lower canine had lost its correct course, so we opted to excise it after confirming the lesion was under control.

Conclusion

The rare, aggressive intrabony tumour known as aggressive fibromatosis typically affects young people and manifests as a painless growth that may disfigure the face. We present a case here that underwent therapy by surgical curettage and flow up, and after 8 months of operation, no indication of recurrence was detected, despite the fact that a few authors had indicated the free margin excision of the lesion and adjuvant chemo or radiotherapy.

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