

Ameloblastic Fibrosarcoma - A Review of the Literature

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Abstract

Ameloblastic Fibrosarcoma is an extremely rare odontogenic tumor with unclear etiology, which has clinical and radiological features similar to other odontogenic tumors. The rarity of these neoplasms plus their overlapping features with other odontogenic tumors can make diagnosis challenging. These cases should be discussed and reviewed by oral pathologists in multidisciplinary conferences, dealing with sarcoma, at national and international level.

Keywords: Ameloblastic fibrosarcoma; Ameloblastic dentinosarcoma; Ameloblastic odontosarcoma

Abbreviations: AFS: Ameloblastic Fibro Sarcoma; PCNA: Proliferating Cell Nuclear Antigen

Introduction

The first description of the rare odontogenic tumor, ameloblastic fibrosarcoma (AFS), was in 1887 [1]. Various terminologies like ameloblastic dentinosarcoma and ameloblastic odontosarcoma were employed in the past to denote these types of lesions, which depended upon the presence of enamel or dentin. In the recent past, multiple authors considered these lesions to be the histological variants of the same lesion and were thought to be for a long time. Recently, the World Health Organization classification of the odontogenic tumors, ameloblastic odontosarcoma, and dentinosarcoma are again separately listed. Reported cases of AFS had ages ranging from 3 to 89 years, [2] with a mean age of all the reported cases summing upto 27.3 years [3], which shows that it has a major tendency towards the younger population. The usual clinical presentation is painful, but occasionally painless facial mass. Paresthesia and dysesthesia are the next progressive symptoms. Continuance of the symptomatic episodes is also gradual and has a varied range, from a few days to two years. When analyzing various published articles reporting cases of AFS and literature reviews, it can be seen that more than 32% of the reported cases of AFS were recurrences and this points towards a slightly increased rate of recurrence which further necessitates meticulous removal of the lesion [4].

Radiographic AFS, as per reports, is an expansile destructive mass with ill-defined and irregular borders. The gross tumor appearance varies from cystic or solid with a fleshy whitish to yellow consistency, which is the cause of bone destruction. Histologically, the epithelial component mimics the developing enamel organ,

and the mesenchymal cells vary from hyperchromatic spindle to satellite and exhibit moderate to marked nuclear pleomorphism with a high number of mitotic figures. Fibroblastic presence is seen ultra structurally. Immuno histochemically, multiple reports showed that while ameloblastic fibroma shows negativity towards the p 53 and proliferating cell nuclear antigen (PCNA), AFS show positivity towards the same [4]. Recurrent AFS shows higher labeling indices for Ki-67 as compared with non-recurrent AFS, and the same can be used to identify between the same and can be managed accordingly [5,6].

Management

Since AFS is a rare entity, the reported literature available is very limited in nature. The oddity of the lesion does not make it exempted from the differential diagnosis of multiple lesions until proven differently. Widely acknowledged treatment modality for AFS is a combination of chemotherapy and radiotherapy, accompanied with wide surgical resection. Local control is strongly dependent on the extent of the initial resection, and conservative surgery must be abandoned and replaced by wide surgical resection with the surrounding soft tissue, particularly when the cortical plates have been perforated. As mentioned above, since more than 30-40% [7] of the reported cases is recurrences, the patient should be kept under long-term follow-up as the reported ages show a wide variance.

Previously, curettage, enucleation, and local excision have typically been utilized in the surgical procedure [8] and have failed exponentially with recurrence. In multiple reported cases, it can be

seen that the combination or adjuvant chemotherapy is regarded as having provided complete remission of large lesions without recurrences [7]. The most effective chemotherapeutic regimes which have most effect seems to be a daily oral cyclophosphamide, weekly intravenous actinomycin-D, and vincristine, along with post-operative radiotherapy, with maximum effect reported with 50 Gy doses. Many authors mandatorily recommend it, when the lesion is large or incomplete removal is suspected [1,8]. AFS is considered by many as a locally aggressive neoplasm with a low potential for distant metastasis (4.5%) and with an overall mortality rate of 25.4% [2,7]. Many of the authors recommend a radical resection along with primary neck dissection. However, authors like Kousar et al. [9] suggests that the radical surgical excision of the AFS should not be accompanied by primary neck dissection, only the surgical resection along with adjuvant chemo and/or radiotherapy is needed.

Prognostic prediction for AFS is still under reviews because of the rarity of the cases [10]. Khalili M [11] reported a case of AFS, which underwent radical surgical resection and radiotherapy with no evidence of recurrence or metastasis after two years, which is considered as a favorable outcome. However, many other authors are of the opinion that given the wide range of age of incidence, two years cannot be considered as a safe margin for non-recurrence [9,10,12], and further long-term case studies should be done to reach a favorable conclusion [13].

Conclusion

AFS is an extremely rare odontogenic tumor with unclear etiology, which has clinical and radiological features similar to other odontogenic tumors. The rarity of these neoplasms plus their overlapping features with other odontogenic tumors can make diagnosis challenging. However, from the available literature data, although in limited numbers, we can safely conclude that if found as AFS, it can be managed with a combination of chemotherapy and radiotherapy, accompanied with wide surgical resection. The patient should be kept under follow-up for a long term as long-term reports, and a unified recurrence data is not available.

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