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Diagnosis and treatment of a rare case of adenomatoid odontogenic tumor in a young patient affected by attenuated familial adenomatosis polyposis (aFAP): case report and 5 year follow-up

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Abstract. – BACKGROUND: The adenomatoid odontogenic tumor (AOT) is a quite rare odontogenic tumor, with an incidence rate of approximately 12 cases/year worldwide. Attenuated familial adenomatous polyposis (aFAP) is a syndrome characterized by a significant risk to develop colon cancer. The aim of the paper is to describe a case never reported before in the literature: an AOT developed in a patient with aFAP; moreover, we want to show how it appears 5 years after surgery and after the regeneration of the eroded bone tissue, using the Platelet-Rich Fibrin (PRF) as filling material.

CASE PRESENTATION: We report the case of a female 18 years old patient, affected by aFAP; she comes to us with a swelling on the right hemi-face. We performed several radiological exams, and they showed a neoformation approximately 2 cm in diameter: this neoformation packed the upper right canine, therefore, we hypothesized a dentigerous cyst.

We decided to proceed to open biopsy and enucleation of the lesion. An intra-operative endodontic treatment on the adjacent partially resorbed teeth was also performed. Finally, we performed a reconstruction of eroded bone tissue, by use of Platelet-Rich Fibrin as filling material.

The samples fixed and embedded in paraffin have led to the diagnosis of AOT. After 5 years from the surgery, we did not find any clear sign of relapse, in addition, the use of PRF has favored an optimal osteogenesis at the surgical site.

CONCLUSIONS: Undoubtedly, a correct diagnosis of AOT allows to have a more performing clinical and surgical approach. Furthermore, this case could document a new manifestation of aFAP in extra-intestinal site.

The onset of an AOT is quite rare in the general population, and this rarity could represent a critical point for its diagnosis; AOT onset in a patient with aFAP is a finding that could represent a new element of diagnosis and, therefore, the starting point to perform a more effective therapy.

Key Words:
Adenomatoid odontogenic tumor (AOT), Dentigerous cyst, Attenuated familial adenomatous polyposis (aFAP), Platelet-Rich Fibrin (PRF).

Introduction

Odontogenic Tumors (OT) comprise a large group of tumors: they differ among themselves for histopathological[1], for radiographic, clinical behavior and for the different localization.

The Adenomatoid odontogenic tumor (AOT) represents 3-10% of all the OT, with about 750 cases described to date in world literature, with an incidence rate which indicates that approximately are discovered 12 cases/year worldwide[2], according to estimates made in the last 60 years[3].

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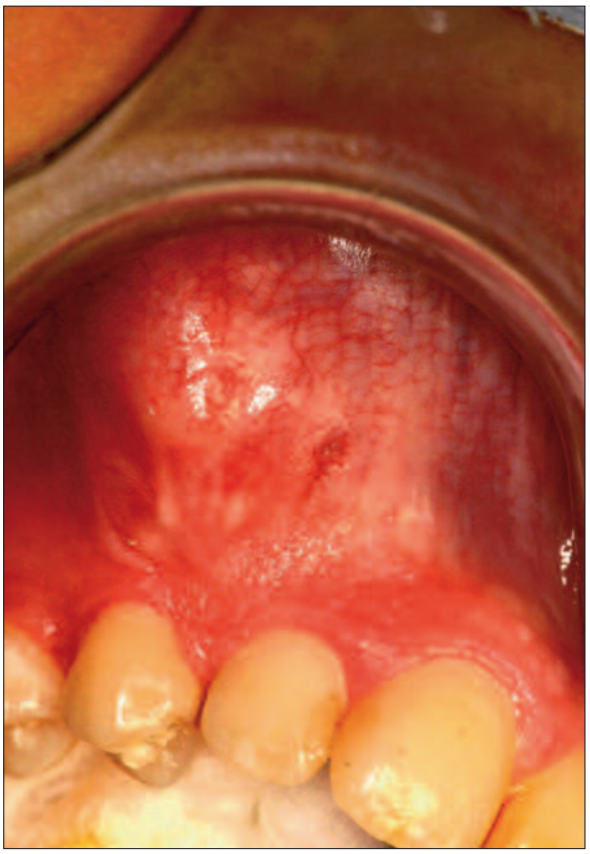

Figure 1. Clinical appearance of the intraoral neoformation.

Attenuated familial adenomatous polyposis (aFAP) is a syndrome characterized by a significant risk to develop colon cancer. The average age of colon cancer diagnosis in affected individuals is 50 to 55 years, that is earlier than when colon cancer typically occurs in the population. In literature, up to now, no case of AOT was reported in patient affected by aFAP, even if the recent researches are more deeply investigating some non-intestine related aFAP findings, such as desmoid tumors and thyroid cancer[4].

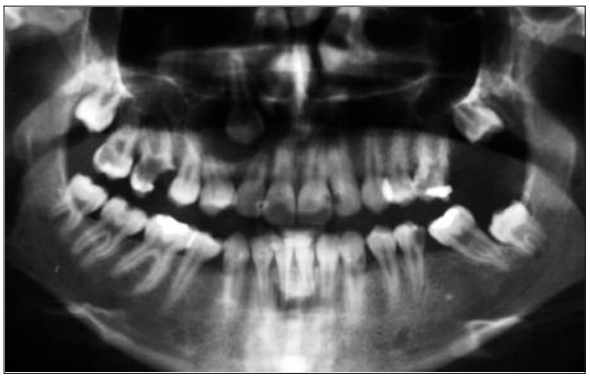

Figure 2. Pre-operatory panoramic X-Ray.

The aim of the paper is to describe a case of AOT developed in a patient with aFAP, moreover, we want to document the use of PRF in the treatment of bone losses resulting from the eradication of tumor and we want to describe the clinical and radiological condition of the patient, 5 years after the surgical treatment.

Case Presentation

We report the case of a female 18 years old patient: she comes to us with a swelling on the right hemi-face, and this swelling persisted for several months, even if the patient referred absence of pain. On clinical examination there was a neoformation with a hard consistency (Figure 1), not painful, at the right side of the vestibular premaxilla. During the anamnesis the patient referred she is affected by attenuated familial adenomatous polyposis (aFAP), diagnosed by means of clinical and genetic analysis.

We performed Orthopanoramic and CT exams (Figures 2, 3): from the images obtained, they

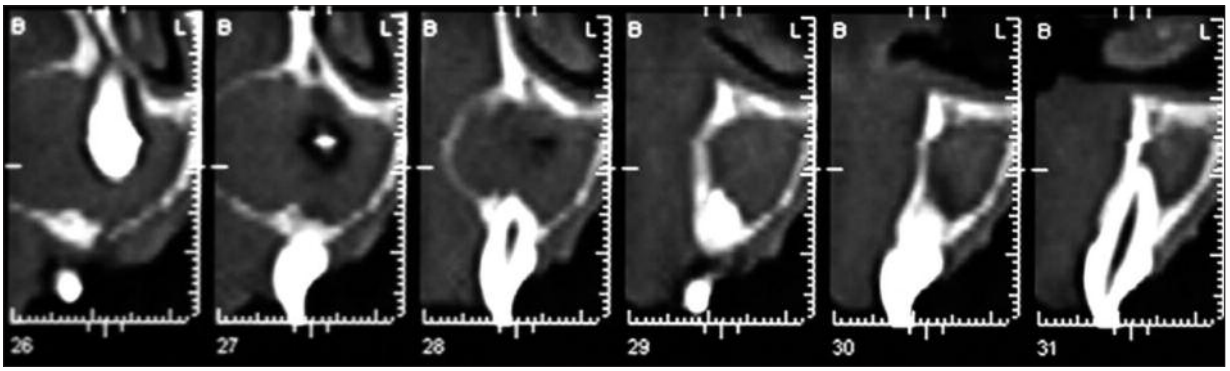

Figure 3. Pre-Operatory CT examination.

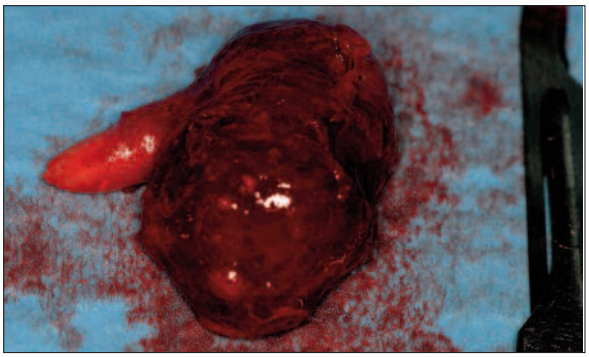

Figure 4. The enucleated neoformation containing an impacted canine inside it.

showed a neoformation approximately 2 cm in diameter, with deformation of the vestibular cortical bone.

This neoformation packed the upper right canine; therefore, we hypothesized a dentigerous cyst. In fact, the clinical and radiological presentation was perfectly conform to a dentigerous cyst diagnosis; however, in the light of the previously referred syndromic condition, we were uncertain about the accuracy of this clinical diagnosis.

The ultrasound examination of the left and right laterocervical region showed no images related to atypical lymph nodes along the course of the vessels of the neck. The chest X-ray was negative. It was decided, therefore, to proceed to open biopsy and enucleation of the lesion. We performed a radical enucleation of the neoformation, and inside it an impacted canine was discovered (Figure 4). Then, we performed an intra-operative endodontic treatment on the adjacent partially resorbed teeth (1.2 and 1.4) (Figure 5) and, finally, a reconstruction of eroded bone tissue, by use of Platelet-Rich Fibrin(PRF) technique[5] (Figure 6). We performed

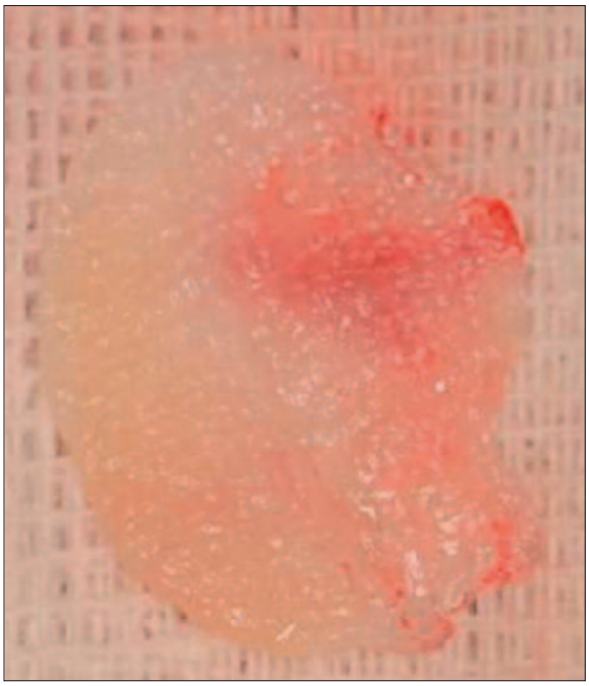

Figure 6. Membranous PRF to be filled inside the surgical site.

a reshaping of the original cortical bone and the subsequent fixing, by means of devices for osteosynthesis (*Utilità, type TRA-100*)[7]. The samples fixed and embedded in paraffin have led to the diagnosis of AOT: in the histological image, was observed, in fact, the presence of odontogenic epithelium, with formation of follicles, and the presence of areas with spindle cells. Psammoma bodies were also present.

The lesion showed complete clinical healing about 1 month later, with the subsequent disappearance of all signs and symptoms.

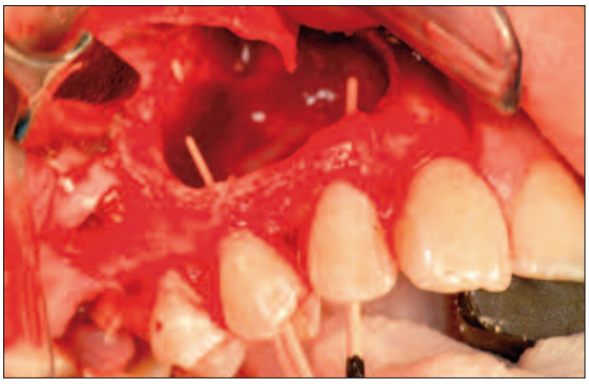

Figure 5. Intra-operative endodontic treatment of teeth 1.2 and 1.4.

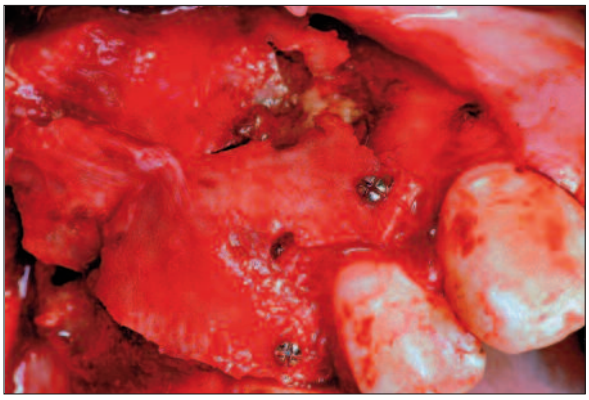

Figure 7. Intra-operative reshaping of the original cortical bone, fixed with surgical screws.

After 5 years from the surgery, we have removed the screws fixed for osteosynthesis: during the clinical and radiological control (Figures 8, 9), we did not find any clear sign of relapse. In addition, the use of PRF has favored an optimal osteogenesis at the site where 5 years before there was a massive osteolysis, resulting from the erosive action of the tumor mass.

Discussion

The clinical and radiographic finding that we found in this case is perfectly comparable to what is stated in the literature regarding the AOT presentation[1,6,7].

Adenomatoid odontogenic tumor is mainly observed in young women, the most frequent site of development is at the maxillary bone in canine region, with a ratio of 2/1 with respect to the mandible[3]. The neoformation can be cystic, it is also generally associated with an impacted tooth, so as to simulate, both radiographically both clinically, a dentigerous cyst. Undoubtedly, a correct diagnosis allows to have a more performing clinical and surgical approach. Furthermore, in those cases with a massive osteolysis is necessary to promote the replacement of tissues by the use of PRF, which has always been used in oral surgery to accelerate and improve the processes of bone regeneration[5,8,9].

This case report elicits a further element of discussion. In fact, the patient is suffering from aFAP; therefore, this case could document a new manifestation of this pathological condition in extra-intestinal site. Already other authors have reported an association between aFAP and desmoid tumors[4], or between aFAP and thyroid cancer[4]. Moreover, some cases have been detected with association between FAP and the onset of osteomas in maxillofacial region. However, even more interesting is what has been observed in a study where it is reported the association between FAP and some important dental abnormalities, such as the presence of unerupted teeth, single or multiple dental agenesis, or, the presence of supernumerary teeth. Finally, it was found that patients with FAP showed an occurrence of dentigerous cysts in approximately 17% of the cases examined, while the percentage of occurrence in the general population stands at around 2%[10].

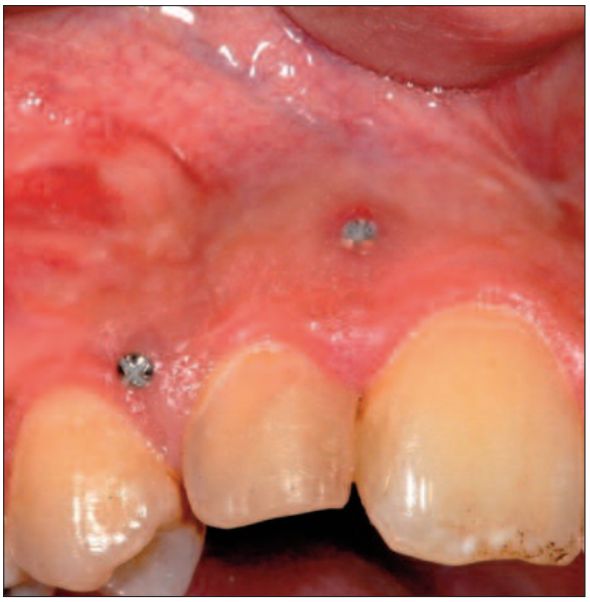

Figure 8. Clinical follow-up after 5 years from surgery.

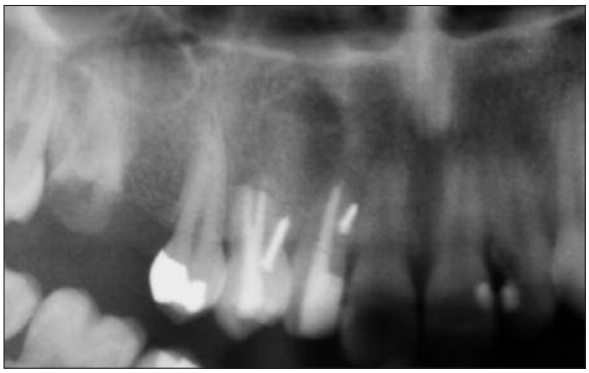

Figure 9. X-Ray follow-up after 5 years from surgery.

Conclusions

The diagnostic approach and treatment of AOT are complicated, primarily because of its rarity and secondly because it always should be considered also all the other OT and cysts of the jaw bones. However, through proper knowledge of the epidemiology of this tumor, through a extensive surgical eradication and through the use of adjuvants of bone regeneration, it may have a "*restitution ad integrum*" without increasing the low risk of relapses that characterizes this disease. Although the onset of an AOT is quite rare in the general population, the onset in a patient with aFAP is a finding that could represent a new element of diagnosis and, therefore, the starting point to perform a more effective therapy.

Consent Statement

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Acknowledgements

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Conflict of Interest

The Authors declare that there are no conflicts of interest.

References

1) Kramer IRH, Pindborg JJ, Shear M. Histological Typing of Odontogenic Tumours. Second Edition. Springer-Verlag, 1992.
2) Neville. Oral and Maxillofacial Pathology (second edition). Saunders Editions, Philadelphia, USA, 2002.
3) Kruse ALD, Zwahlen RA, Grätz KW. New classification of maxillary ameloblastic carcinoma based on an evidence-based literature review over the last 60 years. Head Neck Oncol 2009; 1: 31.
4) Burt RW, Leppert MF, Slattery ML, Samowitz WS, Spirio LN, Kerber RA, Kuwada SK, Neklason DW, Disario JA, Lyon E, Hughes JP, Chey WY, White RL. Genetic testing and phenotype in a large kindred with attenuated familial adenomatous polyposis. Gastroenterology 2004; 127: 444-451.
5) Inchingolo F, Tatullo M, Marrelli M, Inchingolo AM, Scacco S, Inchingolo AD, Dipalma G, Vermesan D, Abbinante A, Cagiano R. Trial with Platelet-Rich Fibrin and Bio-Oss used as grafting materials in the treatment of the severe maxillar bone atrophy: clinical and radiological evaluations. Eur Rev Med Pharmacol Sci 2010; 14: 1075-1084.
6) Holroyd I, Rule DC. Adenomatoid odontogenic tumour in a 12-year-old boy. Int J Paediatr Dent 1997; 7: 101-106.
7) Dayi E, Gürbüz G, Bilge OM, Ciftcioglu MA. Adenomatoid odontogenic tumour (adenoameloblastoma). Case report and review of the literature. Aust Dent J 1997; 42: 315-318.
8) Tatullo M, Marrelli M, Cassetta M, Pacifici A, Stefanelli LV, Scacco S, Dipalma G, Pacifici L, Inchingolo F. Platelet Rich Fibrin (P.R.F.) in reconstructive surgery of atrophied maxillary bones: clinical and histological evaluations. Int J Med Sci 2012; 9: 872-880.
9) Marrelli M, Tatullo M. Influence of PRF in the healing of bone and gingival tissues. Clinical and histological evaluations. Eur Rev Med Pharmacol Sci 2013; 17: 1958-1962.
10) Brett MCA, Hershman MJ, Glazer G. Other manifestations of familial adenomatous polyposis. In: Adenomatous Polyposis and Other Polyposis Syndromes. Edited by Edward Arnold. London, UK, 1994; pp. 143-160.