RADIO-“LOGICAL” REFLECTIONS

A cocktail of abnormalities (atypical dental dysplasia associated with agenesis, extreme microdontia and segmental ankylosis)

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PRESENTATION OF THE CASE

The patient is a 7-year-old Caucasian girl with no significant medical history, who consulted us with complaint of mandibular crowding, and delayed development of tooth 42. Clinically, there are no peculiarities, both at the facial level (Fig. 1) or at the dental and periodontal levels (Fig. 2a–e). There were no abnormalities or developmental disorders in her family, with the exception of her father who has unilateral deafness since birth.

Figure 1
Frontal face photography.
Cohen-Lévy J., Aloé-Tavernier F., Mer G. A cocktail of abnormalities (atypical dental dysplasia associated with agenesis, extreme microdontia and segmental ankylosis)

DESCRIPTION OF THE RADIOLOGICAL RECORD

In a comparison of the two panoramic shots taken 7 years apart (Fig. 3 taken at the age of 7 years, Fig. 4 taken at the age of 14 years), the patient record reveals anomalies located on the left mandibular alveolar process, distal to canine:

- Agenesis of tooth buds of 37 and 38;
- Extreme microdontia of 34 and 35, whose buds have stopped their development and do not have a coronal pulp or root;
- The persistence of primary teeth 74 and 75; root resorption has occurred in the period between the two panoramic radiographs and only approximately 25% of the root length remains;
- 36 presents only a root outline with a single furcation initiation of approximately 2 mm and an enlarged pulp chamber. Its coronary portion appears normal, in terms of morphology and calcification;
- In the period between the two shots, a left lateral open-bite appeared (Fig. 5) on teeth 74, 75 and 36 (suspicion of segmental ankylosis or a primary failure of eruption (PFE)), the occlusion curve is balanced with the maxilla.

The comparison of the basal edges reveals a slight reduction in alveolar height on the left side when compared to the right side.

**WHAT IS THE DIAGNOSIS?**

The case of this young patient is atypical because of the variety of anomalies it presents, which are limited to the left cuspidal mandibular area, while all the other structures appear normal: it combines agenesis, hypoplasia, dysplasia of the hard tissues, apart from a periodontal segmental anomaly with ankylosis.

Each of these anomalies on their own can be compared with a known condition, but to date, no specific condition can group them all together.

Type-I dentin dysplasia (DDI) is a dental formation disorder, which affects the root portion of the teeth, while the crown appears clinically normal. The formation of root dentin is disrupted, presumably because of the abnormal differentiation of odontoblasts derived from ectomesenchyme. It differs from type-II dentin dysplasia, whose characteristics resemble type-II dentinogenesis imperfecta\(^3\) (not associated with osteogenesis imperfecta). Primary teeth have opalescent crowns, the amber coloration of which is sometimes severe. Isolated type-II dentin dysplasia and dentinogenesis imperfecta would be clinical manifestations of the same illness\(^24,8\).

In this case, the permanent molar 36 would constitute an atypical form of DDI, which would be “focal” (as reported in a previous case report), and which would not present all the characteristics generally found, in particular cameral pulp obliteration (Witkop classification\(^25\), O’Carroll et al.\(^17\) and Scola et al.\(^21\)).

In a recent case report (Rocha 2011\(^19\)), a Brazilian team described an atypical
form which also only affected the left mandibular sector without cameral pulp obliteration or periapical lesions. The team of Kosinski et al.\textsuperscript{12} described yet another variation of DDI, located in the left mandibular quadrant, which they attributed to local dysfunction and cell proliferation abnormalities.

DDI is genetic in origin, affecting one individual in 100,000 with classical autosomal dominant transmission. It affects both primary and permanent teeth. A case of recessive transmission was reported in siblings from consanguineous parents\textsuperscript{7}.

The different authors agree that typical cases, which are used as a basis for classifications, do not state all possible variations. They believe that the current classifications are likely to be modified\textsuperscript{8,24} as long as the molecular pathways in question are not all elucidated. The closest classification of current general advances appears to be the one adopted by \textit{Mendelian Inheritance in Man}, an American database. There are equivalent databases, including the French site Orphanet\textsuperscript{®}, which are constantly being updated.

**DIFFERENTIAL DIAGNOSIS**

In regional odontodysplasia (RO), caused by a malformation of dentin, enamel, and cementum, the teeth are hypoplastic and hypocalcified, and therefore have a \textit{"ghost" teeth aspect}, where the enamel and the dentin cannot be clearly distinguished\textsuperscript{16,18}. RO is not caused by genetics, and in the same way as the patient’s case, it affects only one of the four dental quadrants\textsuperscript{1,6,9,10,11}. It is worrying that a form of RO was associated with perceptive deafness, grouped under the term Robinson syndrome. Only two cases have been recorded to date\textsuperscript{5}, without a genetic anomaly being responsible.

In segmental odontomaxillary dysplasia (SOD), it is the maxilla that is affected, with hypertrophy and is sometimes accompanied by facial asymmetry, with hypertrichosis and agenesis or dental hypoplasia of the affected region\textsuperscript{2}.

**HOW SHOULD WE BEST TREAT THE PROBLEM AND WHAT IS THE POSSIBLE IMPACT ON ORTHODONTIC TREATMENT?**

Treatment seeks to anticipate the loss of dental structures, to maintain masticatory functions, to preserve the mesiodistal space and the vertical dimension of the occlusion, while preventing the erosion of the antagonistic teeth.

A conservative approach is recommended, ensuring regular monitoring of alveolar growth and mobility. A case of late root development has been reported\textsuperscript{22}, but in this case the situation has remained stable over 7 years,
with an early resorption, which makes the prognosis unclear.

A “classic” case, with a phenotype of conical roots (not absent roots), was reported. It could be treated orthodontically, using a multi-bracket treatment and at the same time extracting the premolars. Surprisingly, no radicular resorption or periodontal lesions were caused by this treatment.

The case of our patient is quite different because the affected teeth have no roots and show signs of ankylosis (or primary failure of eruption), which contraindicates the orthodontic movement of the affected teeth. A Class-II correction test was proposed, with a multi-bracket treatment, completely bonded in the maxillary arch and partially bonded to the mandibular arch, using mini-screw anchorage in the left mandibular region (elastic support). As the teeth continue to grow, there is a tendency for asymmetry, which was unfortunately observed in this case, with a recurrence of the left-hand side Class-II malocclusion (Fig. 6). Eventually, we decided to stop the treatment.

Autogenous tooth transplantation may be an alternative option when replacing the affected teeth, especially if the third molars germectomies are performed as planned. The prosthetic replacement, worn as an implant, may be considered once the patient stops growing, possibly supplemented with an onlay bone graft, given the vertical alveolar deficiency.

Figure 6
Frontal face photograph showing the deterioration of facial asymmetry, with the chin deviating to the left.

CONCLUSION

The anomalies exhibited by this patient do not yet fit into a definite category. It is important that such atypical dental anomalies, which are often discovered accidentally during a radiographic examination, are reported.

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BIBLIOGRAPHY


INTERNET LINKS

- Orphanet: French-language website dedicated to rare diseases, updated regularly http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=FR&Expert=83450

RECOMMENDED READING