Case Report: Tiffany

S- Subjective Phase

Chief Complaint

I have a broken tooth because I had a deep cavity in that tooth.

History of Chief Complaint

Growing up her baby teeth were very small and brown. Friends did not think she had teeth. Her adult teeth were also discolored and caused her not to want to smile fully. Tiffany has had problems in the past with teeth that have had root canals breaking and having to be extracted.

Medical History

She has no known drug allergies and is not currently taking any medications. Her mother's pregnancy with her was normal as far as she knows with no sicknesses or eating disturbances. She had all her immunizations and had chicken pox as a child. The only major medical history was having tubes placed in her ears as a child under general anesthesia.

Dental History

Tiffany had no cavities in her baby teeth but has had significant dental work done to her permanent teeth. She has had her wisdom teeth extracted and experienced dry sockets. She has had multiple root canals with each resulting in a fracture and extraction of the tooth. She reports that her childhood dentist was very helpful and took time to become knowledgeable about her condition. This helped her to achieve a smile that she is very happy with. Treatment included braces for 1-1.5 years, bleaching with take-home gel and trays that was unsuccessful, gum surgery, and veneers on her front teeth. Her condition has had no effect on what she eats except when she has had a dental emergency. She uses orajel and ibuprofen when she has a dental emergency.

Tiffany reports brushing twice a day and flossing 3x a week. She never took fluoride supplements as a child. She reports no problems with grinding or clenching her teeth.

Since going off of her parents insurance, her dental treatment has been more reactive rather than preventive. She would like to have her broken tooth fixed and also have implants placed where she has missing teeth. She would also like to get onto a regular cleaning and exam schedule.

Family History

Tiffany has three sisters and she is the only one affected with this condition on her teeth. Tiffany's mother has the same condition on her teeth. Her maternal grandmother, aunt, 3 uncles and 4 cousins have the same condition.

O- Objective Phase

Clinical Findings

Oral exam showed Tiffany was missing teeth #12, 30, and only had root tips of #19. Tooth #2 has large occlusal caries and broken mesiobuccal cusp. Teeth #7-10 have porcelain veneers. Her lower anterior incisors are grayish in color with translucent enamel. She also has signs of significant wear/erosion on all her molars. Most of her existing molars have large composite fillings. Tooth #18 distobuccal cusp is fractured.

Periodontal charting reveals probing depths between 1-4 mm with minimal bleeding on probing. She has slight recession 1-2 mm in the canine premolar region.

Radiographs show bulbous crowns with normal thickness of enamel. Pulp chambers are small and either small or calcified in the roots. Radiographs must have been taken before clinical photos due to the variations in dental work. BWX still show presence of at least one third molar.

A- Assessment

Differential Diagnosis

Osteogenesis imperfecta: An autosomal dominant condition causing extremely fragile bones. Classic signs include blue tint to the whites of the eye (blue sclera), multiple bone fractures, and early hearing loss. About half of the people who have OI have teeth that appear normal, and their major concerns are routine care. However, the other half has a defect in the teeth called dentinogenesis imperfecta (DI), sometimes referred to as opalescent teeth or brittle teeth. These teeth may be misshapen, may chip or break easily. Besides tooth development issues, people with osteogenesis imperfecta also tend to have a class three malocclusion and open bite.

Dentinogenesis Imperfecta: is a group of inherited defects in tooth dentin that is distinct from odontogenesis imperfecta as only the teeth are affected. It is caused by mutations in *DSPP* (4q21.3), the gene encoding dentin sialophosphoprotein. The teeth are blue-gray or amber brown and opalescent. On dental radiographs, the teeth have bulbous crowns, narrow roots, and pulp chambers and root canals that are small or completely obliterated. Enamel may crack and shear readily from the dentin when subjected to occlusal stress.

Tetracycline Stain: Causes a yellow or grayish appearance to the teeth. Tetracycline has the ability to incorporate into tissues that are calcifying. People who were exposed to tetracycline while teeth were forming either as a child or while they were developing in utero can develop this condition.

Fluorosis: Defect to enamel caused by ingestion of too much fluoride during tooth development. Most cases of fluorosis are mild and will appear as tiny white specks or streaks that are often unnoticeable. However, in severe cases of enamel fluorosis, the appearance of the teeth is marred by discoloration or brown markings. The enamel may be pitted, rough, and hard to clean.

Dentin Dysplasia I: The condition also known as “rootless” teeth has an incidence of ~1:100.000. Clinically both permanent and primary teeth are of normal shape, form, and color in most cases. Radiographically the teeth have short roots with unusual mobility, spontaneous abscess, and early exfoliation. Crescent-shaped pulpal remnants parallel to the CEJ in the permanent dentition and total pulpal obliteration in the primary dentition are common features. There are usually numerous periapical radiolucencies in non-carious teeth.

Dentin Dysplasia II: Dentin dysplasia type II is distinguished from DGI because the permanent teeth are normal in color but show “thistle-tube pulp chambers” and pulp stones on radiographs. The primary teeth have features of DGI. The permanent teeth are normal shape, form and color in most cases. Both dentinogenisis imperfecta and dentin dysplasia conditions are autosomal-dominant and can be caused by mutations in the dentin sialophosphoprotein gene. Depending on where the mutation occurs on the DSPP gene the resulting clinical phenotype may be dentin dysplasia or dentinogenisis imperfecta type II or type III.

Amelogensis Imperfecta: (AI) is a heterogeneous group of inherited defects in dental enamel formation. The malformed enamel can be unusually thin, soft, rough and stained. The strict definition of AI includes only those cases where enamel defects occur in the absence of other, non-dental, symptoms. Isolated enamel malformations are caused by defects in a number of different genes. X-linked, autosomal dominant and autosomal recessive modes of inheritance are possible, with considerable variability in the character and appearance of the resulting enamel. Fourteen subtypes are recognized.

Hypophosphatasia: Hypophosphatasia is an inborn metabolic disorder of the bones characterized by skeletal defects resembling those of rickets. The symptoms result from a failure of bone mineral to be deposited in young, uncalcified bone (osteoid), and in the cartilage at the end of the long bones (epiphyses) during early years. The activity of the enzyme alkaline phosphatase in blood serum and bone cells is lower than normal. Urinary excretion and blood serum concentrations of phosphoethanolamine and inorganic pyrophosphate are abnormally high. Unlike other forms of rickets, hypophosphatasia does not respond to treatment with vitamin D.

Diagnosis

Dentin Dysplasia Type II**,** confirmed by genetic testing.

Discussion

Dentin Dysplasia is an autosomal dominant genetic defect meaning it doesn’t skip a generation. Tiffany’s children will all have a 50% chance of being affected if her husband doesn’t have a history of the disease. Dentin Dysplasia is the least severe form. Dentinogenesis Imperfecta type 2 and type 3 are much more severe.

P-Treatment Plan

Management

Stainless steel crowns on molars and composite strip crowns on canines and incisors during primary and mixed dentition stage. Full coverage casting crowns on permanent molars to

maintain vertical dimension, composite or porcelain veneers on incisors to establish incisal guidance and on other teeth where esthetic is critical Regular oral and radiographic examinations to detect tooth fracture and dental abscesses are important.

Treatment Outcomes

Teeth affected by Dentin Dysplasia cannot support large bulky fillings, and will tend to fracture. Therefore, teeth needing treatment should be covered and protected with a full crown. Veneers are also an option for the esthetics of anterior teeth.

In Tiffany's case, tooth #2 should be checked for vitality, caries removed and a build up and crown should be placed. If needed, she should be referred to an endodontist to treat #2.

If finances are not a concern, implants should be placed to replace missing teeth. This will require a referral to either a periodontist or oral surgeon to evaluate bone width and height to accommodate implants.

The distobuccal fracture on tooth #18 should be closely evaluated to see if she needs a full crown on that tooth as well. Tiffany should be placed on a regular recall, and have BWX taken every 12-18 months.

Resources

American Academy of Pediatric Dentistry, http://www.aapd.org/publications/brochures/fluorosis.asp

National Organization for Rare Disorders, http://www.rarediseases.org/search/rdbdetail_abstract.html?disname=Hypophosphatasia

M.L. Beattie, J.-W. Kim, S.-G. Gong, C.A. Murdoch-Kinch, J.P. Simmer, and J.C.-C. Hu (2006). Phenotypic Variation in Dentinogenesis Imperfecta/Dentin Dysplasia Linked to 4q21 J Dent Res 85(4):329-333.

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University of Michigan School of Dentistry *Jan C-C. Hu, BDS PhD; James P. Simmer, DDS, PhD, http://www.dent.umich.edu/genetics/shared/ai.pdf*

David Bixler Heritable disorders affecting dentin. In *Oral Facial Genetics* pp227-261.