THE DENTAL MANAGEMENT OF PAEDIATRIC PATIENT WITH CONGENITAL ERYTHROPOIETIC PORPHYRIA

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ABSTRACT:
With approximately 150 cases reported to date, congenital erythropoietic porphyria (CEP) is an extremely rare, autosomal recessively inherited condition. In few cases this deficiency leads to deposition of red-brown pigments in the teeth. This case report described the microscopic study of the extracted tooth sample along with preventive, restorative dental management and oral health counseling in a rare case of Congenital Erythropoietic Porphyria.

Key words: Erythropoietic porphyria, oral health, pigmentation.

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INTRODUCTION:
Congenital erythropoietic porphyria (CEP) was first described by Günther in 1911. Porphyrias are a mainly inherited group of metabolic disorders which affect the haem biosynthesis pathway as a result of an enzyme defect. This can result in the accumulation or the excretion of porphyrins which are purple-red pigments of a crystalline nature.

Early in life, the patient excretes pink, reddish-brown or burgundy-red urine due to abnormally large amounts of included porphyrins. Deposition of the porphyrin pigments in the skin imparts a photosensitivity. In areas exposed to sunlight, vesiculobullous lesions appear and later heal with variable cicatrization. The skin of these areas becomes atrophic and pigmented and may exhibit hypertrichosis.

Oral manifestations include paleness of the oral mucosa and erythrodontia. The primary teeth are dark maroon, but the discoloration is not uniform. The discoloration of the permanent dentition is usually less intense. The incisors are almost completely colored, whereas in the canines only the cusp tips may show discoloration; the molars vary in discoloration too. The teeth characteristically are fluorescent in near-ultraviolet light (Wood's light).

Apart from other areas of treatments i.e. cutaneous and hematologic, Dental treatment is concerned with providing esthetic improvement with crowns or laminated veneers.

There have been few reports of the dental management of patients with CEP and even less literature relating to restorative and esthetic management of the dentition. This case report described the microscopic study of the extracted tooth sample along with preventive, restorative dental management and oral health counseling.

CASE REPORT:
An 8-year-old girl with CEP was referred by his consultant hematologist for a dental opinion and dental care. Her medical history revealed that the skin is photosensitive to ultra-violet light, resulting in skin lesions with blistering. General body examination showed healed lesions on hands and legs which become atrophic and pigmented. (Figure 1)

The lips were dry and cracked in places. Intraoral examination showed pale oral mucosa, discolored permanent incisors, submerged 55 and multiple caries as she was taking syrups continuously, since she was 1 year old, prescribed from the local practitioners and had a poor oral hygiene. (Figure 2-4)

Biochemical tests confirmed an increased excretion of uroporphyrin-I and copro-porphyrin-I in urine and stool which is a characteristic feature of CEP.

Full mouth rehabilitation was planned for the patient. (Figure 5) Grossly decayed tooth got extracted and used for microscopic study.

Extracted tooth was embedded in a small block of clear, quick-cure acrylic. Sections, approximately 250 microns thick, were cut from selected portions of specimen. Each section was then carefully ground, using abrasive surfaces of decreasing grit size, until very smooth sections of approximately 100 micron thickness were produced. These sections were then mounted and examined in light transmitting microscope and polarized microscope.
Microscopic view showed that there was gross brown discoloration of the dentine in bands or lines corresponding to Owen lines. The enamel tended to show much less pigment. The width of red-brown color bands evident in ground section formed along the incremental lines of dentin reflects the length of time serum porphyrin levels were high or low. (Figure 6 and 7) The dental discoloration is thought to be caused by the deposition of porphyrins in the tooth through its affinity for calcium phosphate.

A preventive program was simultaneously implemented. Oral hygiene instructions and comprehensive dietary advice were given. A 0.05% sodium fluoride mouth rinse was recommended for daily use. Patient was then, recalled for follow-up visits.8,9

Figure 1: Atrophic, pigmented skin of hands and feet

Figure 2: Reddish-brown pigmentation on incisors

Figure 3: Submerged 55

Figure 4: Dental caries with all the four quadrants
Figure 5: Full mouth rehabilitation

Figure 6: Light transmitting microscopic view with 10X magnification: gross brown discoloration of the dentine in bands

Figure 7: Polarized microscopic view with 100X magnification: bands exhibited a reddish brown fluorescence more brilliant than the adjacent calcified tissue
CONCLUSION:
Treatment of patients with porphyria represents a challenge for the dentist. But with appropriate treatment plan a satisfactory outcome can be achieved. The dentist should inform the patient about the harmfulness of strong carbohydrate diets, which are prescribed for such patients, oral health and taking relevant contra-measures with regard to increased hygiene.

REFERENCES:

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