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Case Report

Addisonian Hyperpigmentation as Oral Manifestation in Primary Adrenal Insufficiency – A Case Report

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ABSTRACT

Background: Primary adrenal insufficiency is an endocrine disorder which is characterized by aldosterone and cortisol deficiency due to destruction of the adrenal cortex. The purpose of this case report is to present a rare case of 10-year-old child patient who reported with a chief complaint of pain in the upper left back tooth region of the jaw since a week and was diagnosed as Addison disease with hyperpigmentation of dorsal surface of the tongue, perioral structures and skin folds, nails and overall skin of the body. Oral manifestations along with endocrine issues play a crucial role in diagnosis & treatment planning of this disease. Dental infection can be a cause of adrenal crisis in patients with long term adrenal insufficiency, so paediatric dentists should be aware and a multidisciplinary approach is mandatory to ensure adequate medical and dental treatment in children. Limited number of case reports in literature suggest that adrenal crisis is a rare emergency condition in dentistry, and diagnosis is mandatory to rule out any condition before a dental surgical procedure.

Keywords: *Addisonian hyperpigmentation; cortisol; hydrocortisone; primary adrenal insufficiency.*

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INTRODUCTION

Addison's disease is an adrenal gland disease characterized by decreased levels of aldosterone and cortisol.[1] Adrenal insufficiency can be classified as primary, secondary and congenital or acquired.[2] Primary adrenal insufficiency has a typical depleted levels of mineralocorticoid and glucocorticoid.[2]

Cortisol and aldosterone are two important hormones with functions like improvement of synthesis of glycogen in liver, transport of fatty acids from adipose tissues into muscle tissue.[3] Lack of these hormones leads to nausea, vomiting, hypoglycemia, fever, hypotension and shock leading to loss of life.[3,4]

Few etiological factors include autoimmune, idiopathic atrophy, adrenal hemorrhage, pituitary tumors, infectious diseases, etc.[4]

Acute infection which causes tissue destruction and additional stress can raise cortisol levels up to 3-15 times

greater than normal.[5] A rare case of primary adrenal insufficiency with Addisonian pigmentation due to familial glucocorticoid deficiency in a 15-year-old male patient born out of 2nd degree consanguineous marriage was reported by Sunanda Mahajan et al.[6] No relevant history available with respect to the psychosocial and genetic information and any past intervention done. The treatment involves administration of corticosteroids, glucocorticoids, electrolyte and fluid restitution for dental patients. The aim of this case report is to present a case of dental management of a 10-year-old patient who was diagnosed as Addison disease with hyperpigmentation of dorsal surface of the tongue, perioral structures and skin folds, nails and overall skin of the body.

CASE REPORT

A 10-year-old boy visited the Department of Paediatric and Preventive Dentistry Department, with an appearance of generalized hyperpigmentation of the skin and extremities (Fig.1a and 1b) and with a chief complaint of pain in the upper left back tooth region of the jaw since a week. On taking medical history, there was no relevant family history, history of any drug intake or allergy to any other drugs. Patient gave a history of progressive hyperpigmentation since last 4 years. Dental history revealed no prior dental procedures were done and this was patient's first dental visit. On Clinical examination of the oral cavity, hyperpigmentation of the mucosa of the tongue since last 6 months, hard palate, floor of the mouth, gingiva (Fig.2a and 2b) and peri-oral region was noticed. Multiple carious teeth in maxillary and mandibular arch were seen. The left maxillary second molar was grossly decayed and responded painfully to vertical percussion which was indicated for extraction after radiographic assessment of the same (Fig.3a and 3b). Physical examination showed that the boy was underweight (body mass index for age and gender was below the third percentile) and so the patient was referred to the department of paediatrics of the same institute to rule out the systemic diagnosis before the treatment.

Laboratory investigations results revealed significant changes in ACTH Levels (>2000 pg/mL), and Cortisol levels (3.6 mcg/dL). The results of other tests which includes complete blood count, Erythrocyte sedimentation rate, Serum Triiodothyronine, Serum Tetraiodothyronine, Serum TSH and Electrolytes including sodium, potassium were normal and insignificant.

(Fig:1a)

(Fig:1b)

Fig 1a and 1b shows hyperpigmentation of extremities along with nails and diffuse pigmentation seen over face

(Fig:2a)

(Fig:2b)

Fig 2a and 2b shows gingival and mucosal surface hyperpigmentation observed during routine dental examination.

(Fig:3a)

(Fig:3b)

Fig 3a shows intraoral appearance of maxillary arch and fig 3b shows radiographic appearance of carious tooth including 65.

After consulting paediatric endocrinologist, the patient's medical history, examination, and laboratory findings stated that it is a case of adrenocortical insufficiency- Addison's disease. Medications prescribed included oral hydrocortisone tablet (Hisone 7.5mg per day) and CALCIROL sachet (60,000 IU) once in a month with parenteral rehydration during his hospitalization with a 3-month follow-up scheduled for the same. Before minor dental procedure (extraction under local anesthesia with adrenaline 1:80,000), a premedication of hydrocortisone with double the dose of the normal was prescribed one day prior and continued for next 2-3 days as per the paediatric endocrinologist's opinion. Oral antibiotics (Tab. Cefixime 100 mg for 3 days) were prescribed to avoid infection induced adrenal crisis and a follow up after 7 days was scheduled for assessment of wound healing.

DISCUSSION

Adrenal crisis is a condition which demands for emergency medical support. Congenital Adrenal hyperplasia (CAH) is the most common form and autoimmune destruction of adrenal cortex remains the most common cause in adults for both the genders.

Dental infection causes soft tissue pathology has a risk of increasing cortisol levels due to emotional and fear factor and there is greater chances of depletion in the reserved levels of serum cortisol in patients with adrenal insufficiency. Cortisol (hydrocortisone) is the main choice of drug; while other drug preparations like

prednisone, dexamethasone is also used for the treatment. The mechanism of cortisol explained during higher stress is, it compensates for the vascular smooth muscles dilatation and improves permeability and avoids vascular collapse.[7,8]

Risk category procedures are divided into negligible risk, mild risk which includes non-surgical procedures like simple extractions, periodontal surgery, biopsy, etc. and moderate-to-major risk category includes procedures like multiple extractions, bony impaction surgical treatment, osseous surgery, quadrant periodontal surgeries, surgeries associated with blood loss. Comparatively in an age group of children, they have susceptibility to organ hypoperfusion, which can produce severe manifestations of adrenal crisis.[9,10]

The differential diagnosis includes vitamin B12 Deficiency, severe acute infections, various disturbances of the central nervous system and acute poisoning.[11] The clinician should be thorough with the significance of prophylactic steroid doses effect which could be fatal resulting in 'steroid crisis' and thus the practitioner should refer patients to the hospital for the dental treatment. Few authors suggest, the correlation of salivary cortisol levels to circulating plasma cortisol levels, while few suggests accuracy of urinary cortisol levels is high as compared to salivary cortisol levels in diagnosis.[12]

In case of minor dental treatment, the dose should be double that of the daily recommended dose of corticosteroid a day prior, or 2-3 days after the procedure.[13] In the present case, primary adrenal insufficiency was diagnosed with the help of pediatric endocrinologists and pediatricians with the hyperpigmentation of the mucosa of tongue, skin folds and generalized skin of the body. A major contributing risk factor is a dental infection which can severely worsen adrenal crisis and thus thorough knowledge and care if of utmost importance for dental practitioners.[14] The dental care provided to children should also be supported by preventive efforts on the part of parents/caregivers and children. Moreover, general anaesthesia may be considered to be a valid solution if other behavioural management options have been implemented. Ultimately, dental treatment under general anaesthesia will have greater benefits than risks.[15] Establishment of a Dental Home helps in dissolution of certain "barriers" such as financial constraints, lack of provision of professional care, and guidance to the most vulnerable children of the society. Since inception, dentistry followed the concept of "Drill and Fill." However, with greater understanding of the dynamics of dental Caries, greater emphasis is being placed on prevention, and so, a Dental Home can prove to be an ideal place to sow the seeds of primary prevention the benefits of which are reaped throughout the lifetime of the individual.[16]

CONCLUSION

Oral diagnosis in dentistry is the practice of determining all problems inside and outside of the mouth by using scientific knowledge, and thereby helping to make the right diagnosis of oral and systemic health of the patient. A comprehensive dental treatment is essential for improvement of oral health and maintenance of the quality of life in children and high-risk patients. Pediatric Dentists and Pediatricians should be aware of possibility of adrenal crisis and a multidisciplinary collaboration enables the treatment and prevention of exaggeration of systemic condition which can be provoked by dental infections.

DECLARATION OF PATIENT CONSENT

The authors certified that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal.

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