



Allgrove Syndrome (AAA Syndrome): Case Report of Dental Management of 15- Year- Old Male

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ABSTRACT

Allgrove Syndrome (Triple A Syndrome) is a rare autosomal recessive condition in which multiorgan dysfunction occurs. It is characterised by the three 'A's: Alacrima, Achalasia, and Adrenal insufficiency (Addison's disease). About 1/3rd of the patients present with neurological and autonomic manifestations, including Parkinsonism, peripheral sensory and motor neuropathy, gait disturbances, muscle weakness, and mental disability.

This is a case of a 15-year-old male who had multiple carious teeth and broken-down roots. The patient was 5 years old when first diagnosed and had a history of absence of tears, difficulty in swallowing, and was on exogenous steroids. The patient had come to the Oral Surgery Department with the complaint of continuous severe, nocturnal pain in the lower right 1st molar for the last 1 week. So, as per protocol, the dose of the steroids was doubled, and the extraction of the tooth was done the following day

Keywords: Allgrove Syndrome, Triple A Syndrome, ACTH-Resistant Adrenal Insufficiency, Achalasia, And Alacrima

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INTRODUCTION

AAA syndrome is caused by a gene mutation on chromosome 12q13, which encodes the ALADIN protein, associated with ALacrima, Achalasia, aDrenal Insufficiency, and Neurological Disorder^{1,2,3}. It involves a combination of ACTH-resistant cortisol deficiency, achalasia, and alacrimia, which lends the name: Triple A Syndrome^{3,4,5}.

The main aim was to report a case of AAA syndrome that came to the OMFS Department of MIHS with the complaint of severe pain due to multiple carious lesions. This is the first case report for dental management in Pakistan

CASE PRESENTATION

A 15-year-old male reported in the Oral Maxillofacial Department at Margalla Institute of Health Sciences with a complaint of continuous, nocturnal pain for a week in the lower right posterior tooth. He also had multiple carious lesions in other anterior as well as posterior teeth. In order to make a definitive diagnosis, patient was referred to the radiology department for intraoral (periapical) and extraoral (OPG) radiographs. On examination of the periapical radiograph, patient was found to have grossly carious lower right first molar with a distinct radiolucency seen in the furcation area of the tooth (Fig 3). The OPG further revealed multiple carious lesions in the anterior segment and broken down roots in the upper and lower right segments (Fig.7, OPG was taken post operatively hence the missing #46). The decision was made to extract the lower right first molar, which was the primary cause of concern to the patient. For this, the patient was appointed for the following day as discussed below.

The patient was on exogenous steroids. Therefore, as per protocol, the patient was instructed to double the dose of hydrocortisone, i.e., from 5mg to 10 mg for the day before the procedure, on the day of the procedure, and the day after, to prevent adrenal crisis.

The tooth was extracted using a No. 87 forcep (mandibular cow horn forcep) after a profound inferior alveolar nerve block. A figure-of-eight suture was also applied to support healing. The patient was given an antibiotic (Tab Amoxicillin + clavulanic acid, 375 mg BD) and an analgesic (Flurbiprofen 100 mg BD) for the following 5 days. He was also given strict oral hygiene instructions.

The patient was on follow-up for suture removal after 7 days, and his other teeth were also removed in the same pattern. He was also referred to the Operative Department for further evaluation of restorable teeth.



Fig. 1(a) Extraoral Frontal View



Fig. 1(b) Extraoral Lateral View



Fig.2(a) Intraoral view: Maxillary arch showing carious lesions



Fig.2(b) Intraoral view: Mandibular arch showing multiple carious teeth

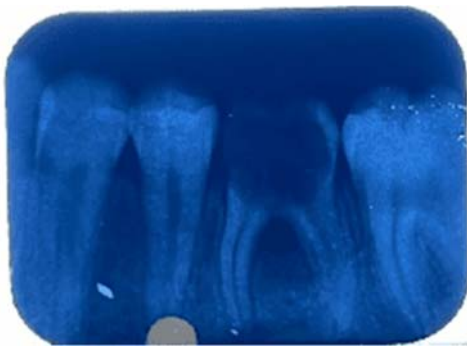


Fig. 3 Periapical radiograph showing grossly carious #46



Fig. 4 Mesial & Distal Roots of #46



Fig. 5(a) Extraction Site



Fig.5 (B) Suturing of Extraction Socket



Fig. 6 Armamentarium



Fig. 7 Post-op Orthopantomogram (OPG)

DISCUSSION

Allgrove Syndrome is an uncommon autosomal recessive endocrine disorder, affecting approximately 1 in every 1,000,000 people^{1,2}. In addition, neurological and autonomic symptoms are observed in about 1/3rd of patients, leading to the syndrome also being referred to as “4A”^{2,6}.

The patient was previously diagnosed with AAA Syndrome and was on exogenous steroids. Due to this he suffered from the inability to increase endogenous steroid production in response to physiologic stress, which in turn increased their risk of suffering from an acute episode of Addisonian crisis. The steroid therapy also increases the patient’s tendency to develop dense alveolar bone, suffer from immunosuppression, delayed wound healing, and emotional lability. The extraction was therefore done surgically to minimize stress to the patient. An anxiety reduction protocol was followed. The patient was given steroid cover in compliance with the protocol by doubling the usual daily dose of steroid on the day before, the day of, and the day after the surgery. On the second day post-surgery, the patient was advised to return to their usual steroid dose [reference from Contemporary Oral and Maxillofacial Surgery by James R. Hupp, Edward Ellis, Myron R. Tucker]. In such cases, the patient can be considered for prophylactic and post-op antibiotics and anti-emetics due to the risk of infections because of immunosuppression and the risk of vomiting^{7,8}.

Extraction should be planned under specialized maxillofacial surgeons who are trained for medical emergencies in a dental office to manage acute adrenal insufficiency.

CONCLUSION

Allgrove syndrome is an uncommon condition, so uncommon that numerous cases remain undetected or are incorrectly diagnosed. Therefore, when a child exhibit any of the three primary symptoms, Allgrove syndrome should be considered, and the child should be monitored closely.

In addition, the dental management is also very important for these patients along with reinforcement of oral hygiene instructions. Early diagnosis and dental management can improve the quality of oral health.

LIST OF ABBREVIATIONS

ACTH: Adrenocorticotropic Hormone

OMFS: Oral Maxillofacial Surgery

OPG: Orthopantomogram

BD: Bi-daily (twice daily)

CONSENT

Informed consent was obtained from the patient for publication of the case report and accompanying images.

FUNDING

None

CONFLICT OF INTEREST

None

AUTHORS' CONTRIBUTION

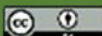
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