Original Article

Comparison of dentofacial findings in sickle cell anemia patients and controls among a tribal population in Kerala – A cross-sectional study

ABSTRACT

Aim: (1) To diagnose by clinical inspection the dental and craniofacial skeletal anomalies in sickle cell anemia (SCA) patients of the particular tribal population. (2) To compare the clinical findings with normal controls of the same population.

Materials and Methods: A study sample of 52 SCA patients and a control sample of 52 non-SCA patients, both belonging to the tribal communities of the area, were selected by nonrandom purposive sampling from the patients attending the Public Health Centre and Dental Outpatient Department of the Government Tribal Hospital of that region. Permission was duly taken through proper channels from the competent authorities. The study design was a comparative cross-sectional study using nonrandom purposive sampling. The study sample consisted of confirmed SCA patients above 18 years of age of both sexes and belonging to any tribe of the region.

Results: Statistical analysis was performed using the Chi-square test for categorical variables and Student's t-test for quantitative variables. Frontal bossing, numb chin syndrome, diastemata, and ischemic pulp involvement were absent in both the study and control groups. Intraoral findings such as the inclination of incisors, overjet, and overbite were tabulated and analyzed statistically.

Conclusion: The data obtained were proposed to be used to formulate a plan for 17 prevention and treatment of the anomalies through timely intervention. Furthermore, the patients need 18 to be made aware of the relationship between these pathologies and SCA. Of the ten features that 19 were examined, only retroclination of incisors showed a significant difference from the control 20 groups. This could be attributed to the lip pressure on the maxilla. However, it did not affect 21 esthetics or function, and therefore, no intervention was required.

Keywords: Dentofacial abnormality, pulpal, sickle cell anemia, tribal population

INTRODUCTION

Sickle cell anemia (SCA) is a genetic hemoglobinopathy which results in morphological changes in the red blood cells (RBCs) and renders the patient susceptible to its consequences. The RBCs undergo transformation from normal biconcave shape to take on a rigid and curved shape which resembles a sickle. The clinical manifestations are primarily the result of ischemia caused by obstruction of the microvasculature by these abnormal RBCs and hence may be seen in any tissue of the body. Thus, a wide range of symptoms involving any organ may be expected.[1] Dental findings occurring in SCA patients have been documented in previous studies.[2] Patients with SCA are found to exhibit specific changes in their dental and craniofacial skeletal complex. These result in loss of esthetics and compromise function.

Some of these bone and dental pathologies can be ameliorated or corrected by proper and timely interventional

Anila Karunakaran, K. K. Sivaprasad, P. Ranjith Kumar, Leena Philipose

Department of Dentistry, Institute of Integrated Medical Sciences (Government Medical College), Palakkad, Kerala, India

Address for correspondence: Dr. P. Ranjith Kumar, Department of Dentistry, Institute of Integrated Medical Sciences (Government Medical College), Palakkad, Kerala, India.
E-mail: drranjithp@hotmail.com

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treatment. Hence, it is imperative that these are diagnosed as being caused by SCA and patients are made aware of the same. Here, we have screened selected dental and skeletal findings in SCA patients in a tribal population of a particular area in Kerala state, India, and compared them with the normal subjects of the same population.

Aims and objectives

Through this study, we aim:

1. To diagnose by clinical inspection the dental and craniofacial skeletal anomalies in SCA patients of the particular tribal population
2. To compare the clinical findings with normal controls of the same population.

The objectives of the study were:

1. To identify the dental and craniofacial skeletal anomalies in SCA patients of the tribal population
2. To assess the extent of severity of the above pathologies in the study population as compared with the normal controls
3. To make the patient aware of the relationship of the anomalies with the underlying sickle cell disease (SCD)
4. To formulate a protocol for treatment for dental and craniofacial skeletal anomalies in the study population.

MATERIALS AND METHODS

A study sample of 52 SCA patients and a control sample of 52 non-SCA patients, both belonging to the tribal communities of the area, were selected by nonrandom purposive sampling from the patients attending the Public Health Centre (PHC) and Dental Outpatient Department of the Government Tribal Hospital of that region. Permission was duly taken through proper channels from the competent authorities. The study design was a comparative cross-sectional study using nonrandom purposive sampling.

The study sample consisted of confirmed SCA patients above 18 years of age of both sexes and belonging to any tribe of the region. Those with systemic comorbidities, craniofacial anomalies, congenital cleft lip/palate, and a history of orthodontic treatment or surgical treatment involving the head and neck were excluded from the study. These confirmed SCA patients are registered in the Government Tribal Hospital/PHCs and are undergoing regular weekly checkups.

Patients of the tribal population who have been duly screened and found to be free of SCA and who have reported to the PHC/Tribal Hospital for other treatments were taken as controls.

All patients were taken up for the study after obtaining informed consent. The patients and controls were examined at the Government Tribal Hospital of the region.

Each patient was examined for the following:

Extraorally

1. Protrusion of maxilla
2. Depression of the nasal bridge
3. Mandibular overgrowth
4. Frontal bossing
5. Numb Chin syndrome.

Intraorally

6. Overjet
7. Overbite
8. Diastemata
9. Retroclined maxillary and mandibular incisors
10. Ischemic pulpal involvement (dental pain in teeth unaffected by caries, periodontal pockets, regressive changes, or trauma).

Extraoral findings were detected by photometric analysis[3] done using NemoCeph software, except for frontal bossing and numb chin syndrome, which were detected by inspection and palpation. Intraoral findings were detected by history taking, inspection, and percussion. Intraoral examination was done using a mouth mirror and probe under proper illumination. The inspection findings such as the inclination of incisors, overjet, and overbite were recorded in the prepared format and tabulated. The patient was positioned in front of a grid using standard procedures used for a lateral cephalogram. Photographs were taken using an Olympus Digital Camera placed at a fixed distance [Figure 1]. The soft-tissue landmarks used in this study are G-glabella, N-nasion, Cm-columella, Sn-subnasale, Ls-labial superior, Pg-pogonion, and Prn-pronasale. Angular measurements of the following were recorded to detect the respective skeletal anomalies. The same procedure was repeated for the controls. The findings of the confirmed SCA patients were compared with the normal controls. Statistical analysis was

Figure 1: Patient positioning in front of a grid for a lateral cephalogram

performed using the Chi-square test for categorical variables and Student's *t*-test for quantitative variables.

RESULTS

Frontal bossing, numb chin syndrome, diastemata, and ischemic pulp involvement were absent in both the study and control groups.

Intraoral findings such as the inclination of incisors, overjet, and overbite were tabulated and analyzed statistically [Tables 1-3 respectively].

Maxillary overgrowth [Table 4] was identified by the decrease in nasolabial angle (Cm-Sn-Ls). Nasolabial angle (Cm-Sn-Ls) decreases with increased maxillary protrusion.

Depression of the nasal bridge [Table 5a and b] was detected by a decrease in the angle of the nasal dorsum (N-Mn-Prn) and nasomental angle (N-Prn/N-Pg).

Mandibular protrusion [Table 6] was assessed by the decrease in projection of the upper lip to chin (N-Pg/N-Ls). Projection of upper lip to chin (N-Pg/N-Ls), decreased in mandibular overgrowth.

Table 1: Inclination of incisors

Group	Retroclined (%)	Straight (%)	Normal/proclined (%)
SCA	4 (7.7)	20 (38.5)	28 (53.8)
NCA	0	0	52 (100)

SCA: Sickle cell anemia, NSCA: Non Sickle Cell Anemic

Table 2: Overjet

Group	*n*	Median	Range	*P*
SCA	52	2	−1-7.5	0.448
NCA	52	2	0-10	

SCA: Sickle cell anemia, NSCA: Non sickle cell anemic

Table 3: Overbite

Group	*n*	Median	Range	*P*
SCA	52	1	0–5	0.579
NCA	52	2	0–4	

SCA: Sickle cell anemia, NSCA: Non sickle cell anemic

Table 4: Maxillary protrusion using nasolabial angle

Group	*n*	Mean (SD)	*P*
SCA	52	97.02 (11.34)	0.29
NCA	52	91.63 (14.02)	

SCA: Sickle cell anemia, SD: Standard deviation, NSCA: Non sickle cell anemia

Table 5a: Nasal bridge depression using angle of nasal dorsum

Group	*n*	Mean (SD)	*P*
SCA	52	177.79 (9.39)	0.579
NCA	52	172.24 (9.92)	

SCA: Sickle cell anemia, SD: Standard deviation, NSCA: Non sickle cell anemic

Table 5b: Nasal bridge depression using nasomental angle

Group	*n*	Mean (SD)	*P*
SCA	52	31.84 (4.78)	0.79
NCA	52	32.26 (2.79)	

SCA: Sickle cell anemia, SD: Standard deviation, NSCA: Non sickle cell anemic

Table 6: Mandibular protrusion using projection of upper lip to chin

Group	*n*	Mean (SD)	*P*
SCA	52	12.91 (3.79)	0.41
NCA	52	11.82 (2.88)	

SCA: Sickle cell anemia, SD: Standard deviation, NSCA: Non sickle cell anemic

DISCUSSION

SCD is a group of inherited autosomal recessive hematologic disorders consisting of SCA, sickle cell hemoglobin disease, and beta-thalassemia. It is an autosomal recessive condition caused by the homozygous occurrence of an abnormal form of hemoglobin (HbS).[1] Unlike normal adult hemoglobin (HbA), HbS polymerizes under low oxygen tension (after transfer of oxygen to cells), imparting a rigid half-moon shape to RBCs and decreasing their normal elasticity. SCA is the most seen form of SCD and is due to the replacement of glutamic acid by valine in position 6 of the N-terminus of the beta chain of globin.[2,4] SCD is the most common genetic disorder affecting the tribal population of Central and Southern India. African Americans and Tribals are more affected by this disease which is endemic in malaria-prone tribal belts worldwide.[4] The sickled RBCs are more resistant to the malarial parasite, and the mutation of HbA to HbS is thought to be an attempt to stall the malarial onslaught.[2] Common oral findings associated with SCD are mucosal pallor, delayed eruption, and dental hypoplasia. Malocclusion (including overjet, overbite, and retroclined maxillary and mandibular incisors) and a decrease in dental caries and diastemata are also observed. Skeletal anomalies seen are protrusive maxilla and forward advancement of the mandible. The sickling crises occurring within the microcirculation of facial bones and dental pulps and small areas of necrosis in the bone marrow cause orofacial and dental pain in the absence of dental pathology.[4] In our study, the only significant finding was retroclination of the upper and lower incisors. In a population where bimaxillary protrusion with proclined incisors is a normal finding, this could be attributed to the lip pressure occurring as a result of rapid protrusive growth of the jaws.[4,5] This helps to compensate for or decrease the overjet caused by the disease. The mandible is poorly perfused when compared with the maxilla, and so there is more chance of the development of osteomyelitis. Frontal bossing and maxillary protrusion are observed. This may be due to bone marrow hyperplasia in SCD patients. Vaso-occlusive crises can cause inferior alveolar nerve neuropathies resulting in numb chin syndrome. Pulpal

changes may also be asymptomatic in SCD patients, and so they may not be aware of pulpal damage. Repeated hemorrhage and fibrous tissue repair cause gingival enlargement in SCD patients.[1,6] Of all SCA patients screened in a study in Brazil, 10.2% were indicated for endodontic treatment.[7]

CONCLUSION

The present study attempted to know if any morbidity was caused to the dentofacial complex as a result of SCA among tribals, among whom it is prevalent. In our findings, only retroclination of incisors was found to be significant. This assumes importance as tribals are a population where the bimaxillary protrusion is seen normally. However, this did not affect esthetics or function, and hence no intervention is required for the SCA patients of the study conducted area. The absence of pathologies may be attributed to early intervention, proper medical surveillance, and adequate treatment provided to these patients by the state. However, studies are recommended on a larger sample for further evaluation.

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Conflicts of interest

There are no conflicts of interest.

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