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Otolaryngological manifestations in sickle cell disease patients in Port Harcourt.

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ABSTRACT

Background: Sickle Cell Disease is the most common hereditary disorder. The complications of the disease are seen in most human organs. Apart from hearing loss, there is paucity of information on other otorhinolaryngological manifestations of sickle cell disease in our environment.
Aim: To determine the common otolaryngological disorders in sickle cell disease patients in University of Port Harcourt Teaching Hospital
Patients and method: This was a three-year retrospective study of sickle cell patients seen in haematology and Ear, Nose and throat clinic university of Port Harcourt teaching hospital from June 1st, 2015 to June 30th, 2018.
Result: Sixty-three sickle cell disease patients aged 0- 29 years had otolaryngological conditions. There were fifty nine (59) HbSS and four (4) HbSC. Thirty-six (57.1%) of the patients had tonsillitis, thirteen (20.6%) had obstructive adenoid and tonsils, three (4.8%) had cerumen auris, one (1.6%) patient had sensorineural hearing and one (1.6%) patient had head and neck lymphoma.
Conclusion: The study revealed that diseases of the tonsils were the commonest otorhinolaryngological disorders seen among sickle cell disease patients in Port Harcourt. Prompt diagnosis and treatment is key to a favourable outcome.

INTRODUCTION:

Sickle cell disease (SCD) is a group of haemolytic disorders in which the erythrocyte assumes "sickle" shape when exposed to abnormal oxygen tension.[1] The morphologic changes in the erythrocyte results from single nucleotide change in the β-globin chain leading to substitution of valine for glutamic acid at position 6 of the amino acid sequence.[1,2] The normal red blood cell flows through the blood vessels because it is discoid and pliable.[3] In contrast, when sickled erythrocytes are exposed to stress like dehydration, deoxygenation, hypothermia or infection may change their mechanical and morphologic properties.[2,3] These changes significantly hinder their flow through small vessels, which in turn leads to obstruction, ischaemia and end organ hypoxia. Complications resulting from vaso-cclusive crisis have been reported in most body

organs/system. [4] There is however, paucity of studies on otorhinolaryngological complications of sickle cells in our environment.

The known otorhinolaryngological complications seen in the SCD patients include sensorineural hearing loss, adenotonsillar disease, priampism of the nasal turbinates, epistaxis and cervical lymphadenopathy.[5-8] The prevalence of sensorineural hearing loss among SCD children in Nigeria is between 3.8-13.4% and as high as 66% in adults. [4,9,10] It is thought to occur to due to vaso-occlusion of the labyrinthine artery which supplies the cochlear. [11] Adenotonsillar disease is quite frequent in SCD disease children. [5, 12, 13] Several hypotheses have been put forward to explain the association. Firstly, the compensatory theory which suggests adenotonsillar hypertrophy to be compensatory to autosplenism, secondly increased upper airway infection due to decrease opsonization of encapsulated bacteria and thirdly a possible haemopoetic activity in the adenoid and tonsil. [5] Epistaxis is not a well-recognized complication of SCD; however, its occurrence among SCD patients has been noted in some studies. [7] It is hypothesized to be due hypersplenism and thrombocytopenia or thombo-infarctive process in the nasal mucosa over Little's area. [7]

The aim of this study is to document the common otorhinolaryngological manifestations among SCD patients in our environment. Thorough understanding of otorhinolaryngological manifestation in SCD will help improve management of these patients.

METHOD:

This was a retrospective study of patients referred from haematology clinic to Ear, Nose and Throat Department of UPTH from June 1st, 2015 to June 30th, 2018. Patients case notes and clinic records were accessed. Information was retrieved from patients' case notes and ward records. Patients age, sex, haemoglobin genotype, presenting complains and diagnoses were analyzed. Retrieved record was loaded into Microsoft excel spreadsheet and analyzed with SPSS version 20.

RESULT:

Sixty-three (63) SCD patients aged between 0-29 years were seen. Thirty-eight (60.3%) were male while twenty-five (39.7%) were female giving male female ratio 1.5:1. Most (93.7%) of the patients had sickle cell anaemia (HbSS) while 6.3% had HbSC. Most (79.4%) of patients studied were between 0-9 years of age bracket. The commonest complain was fever seen 80%, followed by painful swallowing seen 65%. Obstructive adenotonsillar enlargement was seen 20.1% of cases. Recurrent tonsillitis seen 57.1% was the commonest diagnosis. Obstructive adenoid and tonsil was seen 20.6%.

Table 1: Age Distribution of Patients

Age category	Sickle Cell dx n (%)
0 – 9	50 (79.4%)
10 – 19	10 (15.9%)
20 – 29	3 (4.7%)
Total	63 (100%)

Table 2: Diagnosis

Diagnosis	Frequency
Recurrent Tonsillitis	36 (57.1%)
Obstructive adenotonsillar disease	13 (20.6%)
Otitis externa	1 (1.6%)
Otitis media	3 (4.8%)
Cerumenauris	2 (3.2%)
Epistaxis	2 (3.2%)
Pharyngotonsillitis	5 (7.9%)
Lymphoma	1(1.6%)

DISCUSSION

Recurrent tonsillitis was the commonest diagnosis seen in the study. Tonsillitis is a common finding in sickle cell disease. [5, 12, 13] Frequent episodes is said to be associated with painful crisis.[13]Ajulo reported reduced incidence of complications from pneumococcal infections following tonsillectomy.[12]

Obstructive adenontosillar disease was seen in 20.6% of patients. This quite high compared to rate seen in similar age group without sickle cell disease in Nigeria.[14] This also agrees with the findings of Gois et al., in which they found 21% as the prevalence of obstructive adenotonsillar disease in SCD.[15]

Only one case of sensorineural hearing loos was seen in this study. This is quite low compared to the findings of Onakoya et al in Ibadan, Nigeria.[10] They studied 167 adult SCD patient's loss in 66% of their subjects.[10] The difference in prevalence may be attributable to the adult study population.[10] However the prevalence of hearing loss in this study is similar to that reported by Alabi et al in Ibadan.[4] This may be as a result of the similarity in demographics in both study population.

Epistaxis was seen in 3.7% of SCD patients in this study. This is similar to the prevalence gotten among non-sickle cell patient in Kaduna by Kirfiet al.[16] However, this low compared to study similar among sickle cell patients by Nardo-Marino in Uganda.[7]

In conclusion, recurrent tonsillitis and adenotonsillar hypertrophy are commonest otorhinolaryngological manifestation seen in this study. The prevalence of sensorineural hearing loss was very low in this series. We hope that will generate interest to encourage detailed prospective studies on the subject matter.

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Conflict of Interest:

There was no conflict of interest

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