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Full Length Research Paper

Challenges of adenotonsillectomy in sickle cell disease patients

O. B. da Lilly Tariah[1], A. C. Nwogbo*[1] and E. Ebong[2]

[1]Otolaryngology Department, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers State, Nigeria.
[2]Anaesthesia Department, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers State, Nigeria.

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Sickle cell (ss) diseases, a common haemoglobinopathy are an autosomal recessive disease. Hypertrophy of adenoid and tonsils have been known to persist in ss disease children than in healthy ones. Greater challenges exist during adenotonsillectomy for these patients. It is a retrospective study between January 2010 to January 2013 in University of Port Harcourt Teaching Hospital Nigeria. Case notes of (16) sixteen patients with ss disease, who had adenotonsillectomy were retrieved. Gender, Age, Various indications and challenges were recorded and analyzed. Six (6) patients presented within age range three to five years, Table 1, followed by four (4) patients presenting at age range 6 to 8 years, male/female ratio 3:2. Four (4) patients had post operative bleeding, out of which one died. Persistent low PCV were observed in almost all the patients. Challenges were enumerated in Table 4. Greater challenges are encountered in sickle cell children, undergoing adenotonsillectomy, compared to normal children.

Key words: Sickle cell anemia, Adenotonsillectomy.

INTRODUCTION

Sickle cell (ss) disease, a common haemoglobinopathy is an autosomal recessive disease caused by a single DNA substitution of valine with glutamic amino acid in the beta globin chain (Witting et al., 1988). Hence, HbA is changed to Hbs, which polymerizes under hypoxemic condition leading to sickling of red blood cells, increased haemolysis and adhesion, leading to thrombotic phenomenon. This will result in to ischaemic changes and fibrosis.

There is also neovascularization leading to poor control of bleeding during surgical procedure. They are also prone to infection and the disease is common in Africans.

Hypertrophy of adenoids and tonsils have been known to persist longer in ss disease children than in other healthy children who often times outgrow the phase of lymphoid hypertrophy (Witting et al., 1988; Sidman and Fry, 1988)

Adenotonsillecctomy is a surgical procedure with various indications, however, obstructive upper air way constitute a major indication for which this procedure is done in children, followed by recurrent tonsillitis. Despite various complications that may arise in the surgical process, greater challenges exist amongst patient, with ss disease (Halvorson et al., 1997).

The aim of this study is to look at the various challenges encountered in preoperative evaluation and surgical procedures as well as post operative period in ss disease patients.

MATERIALS AND METHODS

This is a retrospective study between January 2000 to

*Corresponding author. E-mail: nwogbo_augustine@yahoo.com.

Table 1. Age distribution of patients.

Age range of patients	Number of patients
3 - 5	6
6 - 8	4
9 -11	2
12–14	2
15- 17	1
18–20	1
Total	16

Table 2. Gender distribution of patients.

Age range of patients	Gender distribution	
	M	F
3 - 5	4	2
6 - 8	3	1
9– 11	1	1
12- 14	1	1
15–17	-	1
18–20	1	-
Total	10	6

Table 3. Post operative complication.

Post op result of patients	Sicklers	%	Normal Patients
Patients discharged within 3 – 4 days	11	68.7	230
Bleeding 6 – 8 h post operation	3	18.7	5
Readmission for secondary haemorrhage	1	6.2	0
Died due to post operative haemorrhage	1	6.2	2
Total	16		237

January 2013 in Otolaryngology Department of University of Port Harcourt Teaching Hospital Nigeria. Within the period of study 253 Adenotonsilletomies were done. Sixteen (16) were ss patients who had recurrent tonsillitis and obstructive upper air way from adenoid and tonsils. Their case notes were retrieved for analysis. All patients had full blood count and differential, x-ray postnasal space and chest. In all plain x-ray, it showed evidence of postnasal obliteration from adenoid pad. Their gender, age, various indication and challenges were recorded. All ss disease diagnosis, were confirmed through blood analysis. Those without ss were excluded. Haemostasis during surgical procedure was done by pressure packing and bleeding point ligation.

RESULTS

Sixteen (16) patients had sickle cell (ss) disease out of 253 patients that had adenotonsillectomy within the period of study. Six (6) patients presented within age range of 3-5 years (Table 1), followed by 4 patients presenting at age range 6-8 years. Ten (10) patients were male while six (6) were female, with male/female ratio of 3:2 (Table 2). In Table 3, four (4) patients had postoperative bleeding, and out of the four (4), one (1) patients died. Persistent low PCV was observed in all patients, thereby delaying the period of surgery. Late presentation and others recorded in Table 4 were various challenges encountered in the study. About 1/3 of sicklers were discharged four (4) days post operative period.

DISCUSSION

Hypertrophy of Adenoid and tonsils have been noted to persist longer in sickle cell children than in normal children. In Table 1 greater number of children was between ages 3 to 11 years. This was also noted by Raj

Table 4. Various challenges indentified.

S/No.	Challenges identified
1.	Persistently low PCV
2.	Intraporative haemorrhage
3.	Late presentation/Co-morbidity
4.	Post Operative Bleeding
5.	Pre and operative High Oxygen requirement
6.	Blood transfusion

et al. (2010) in their study, tonsillectomy for obstructive sleep Aponea in sickle cell Anaemia (Witting et al., 1988). This also contributes to the tendency of these groups of children presenting late for adenotonsillectomy.

Late presentation places more challenges in surgical management of these patient, due to increased haemorrhage during and after surgical procedure of adenotonsillectomy. Persistence of large tonsils and adenoids may be due to defects in compliment pathway that affects immune surveillance in these children.

Repeated bacterial and viral infection is other potential causes (Sidman and Fry, 1988; Suen et al., 1995). Some presented later as observed in Table 1, these children are faced with other challenging medical conditions; hence attention is not given early to the obstructive symptoms (Halvorson et al., 1997).

There is a male preponderance; this was noted by Warrier and Athale in their study, tonsillectomy and Adenoidectomy for obstructive sleep Apnea in sickle cell Anaemia, involving 28 children in a retrospective study (Raj et al., 2010). However, the reason for this is not very clear. This study recorded a ratio of 3:2.

Persistent low PCV which is associated with sickle cell children was noted in this study, constitute great challenge in pre and operative surgical period. Therefore protocol based comprehensive team oriented management of pre-operative transfusion; hydration, antibiotics, anaesthesia and post-operative care were followed to avoid the risk of hypoxemia, dehydration and possible hypothermia (Raj et al., 2010; Coker and Milner, 1982).

Generally, haemorrhage is a known complication in Adenotonsillectomy, but become a greater challenge in children with sickle cell disease undergoing Adenotonsillectomy (Wali et al., 2003; Duke et al., 2006). In our study three children had postoperative bleeding 6 to 8 h after procedure. One died due to postoperative bleeding, site of bleeding was from the post nasal space, with resulting disseminated intravascular coagulopathy (DIC) with multiple organ failure. One was readmitted following bleeding from operative site nine (9) days post surgery.

High oxygen requirement became obvious in preoperative and operative period, considering persistence in low PCV in these children. It is known that decrease oxygen tension can lead to vasocclusive crisis, prolonged and chronic hypoxia can also predispose to abnormal cerebrovascula flow patterns with risk of CVA (cerebrovascula accident) (Waldron et al., 1999).

More than half of children used in this study had blood transfusions at various stages, depending on when indicated. Challenges associated with blood transfusion were also encountered in this study (Waldron et al., 1999; Bader-Meunier et al., 2007). In normal children blood transfusion is not a mandatory requirement for procedure except when indicated. However, in sickle cell patient preoperative consideration for blood transfusion must be considered before procedure is commenced (Viner et al., 1991; Hill et al., 2006).

CONCLUSION

Adenotonsillectomy, like any other surgical procedure can result into some complications. However greater challenges are encountered in patients with sickle cell disease. These challenges must be properly evaluated before surgical procedure is commenced.

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