Paediatric ocular and adnexa malformation in Birnin Kebbi, north-western Nigeria

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Original Article

ABSTRACT

Objectives: Congenital eye and adnexa malformations although rare, are however a major challenge to health professionals in terms of diagnosis. There are also emotional, financial and stigmatization challenges experienced by the patients and their relatives. Appropriate management of these patients requires multidisciplinary approach. The study is to determine the pattern and presentation; and also create awareness among health workers and the public on the congenital eye/adnexal malformation.

Methods: A 3-year retrospective review of all patients who presented at our hospital on account of congenital eye/adnexal malformations from 1st January 2011 through 31st December 2013 was done. Information on socio-demographics, laterality, type of malformation, presentation and Visual acuity were extracted from patients' folders. The data was analyzed by SPPS version 18.

Results: 78 cases of congenital eye/adnexal malformations were recorded during the study period. There were equal numbers of males and females (39 each). Most of the patients were 1year and above (57.7%) at the time of diagnosis. Both eyes were involved in 54 (69%) patients and right eye were 17 (22%). The common congenital eye/adnexal defects identified were congenital ptosis 12 (15.4%), glaucoma 10 (12.8%) and esotropia 9 (11.5%). Majority of the mothers were between 21 and 25years of age (65.4%) as at the time of giving birth to these patients.

Conclusion: Congenital ptosis and glaucoma were the commonest types of congenital eye/adnexal defects seen in this environment. These defects were important causes of poor vision and blindness. Parents/caregivers and health professionals awareness on examination, early presentation and treatment is highly required.

Key words: Congenital eye/adnexal malformations, paediatric, parent/caregiver

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Oculaire de pédiatrie et de ses annexes malformation au Birnin Kebbi, du nord - ouest du Nigeria

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Article Original

RÉSUMÉ

Objectif: Yeux congénitales el formations adnexamals bien que rares, sont cependant un défi majeur pour les professionnels de la santé entermes de diagnostic. Il y a aussi, les défis financiers et la stigmatization émotionnelle subis par les patients et les proches des patients. Une gestion appropriée de ces patients nécessite une approche multidisciplinaire. Pour déterminer le modelé et la présentation et de sensibiliser les agents de santé et le public sur le congénital des yeux/annexes malformation.

Méthode: Une etude rétrospective de tous les patients qui se sont présents à notre hôpital à cause de malformations congénitales des yeux/annexcelles de l'er Janvier 2011 jusqu' à 31 Décembre 2013 a été faite. Informations sur socio-démographiques, la latéralité ; le type de malformation la présentation et l'acuité visuelle ont été extraites des dossiers des patients. La donnée était analysée par la version SPPS 18.

Résultat: 78 cas de malformations congénitales des yeux/annexielles ont été enregistrées pendant la période d'étude. Il y avait un nombre égal d'hommes et de femmes (39 chaque). La plupart des patients étaient de un an et plus (57.7%) au les deux ont été impliqués en 54 (69%) patients et l'oeil droit était 17 (22%). Les anomalies congénitales des yeux/annexielles identifies étaient Ptose congénitale 12 (15.40%), et le glaucome 10 (12.8%), l'estropie 9 (11.5%). La majorité des mères étaient entre 21 – 25 ans de l'âge (65.4%) pendant la naissance de ces patients.

Conclusion: Ptose congénital et glaucoma étaient les types les plus courants de l'œil/défauts congénitales annexcelles vus dans cet environnement ces défauts étaient des causes importantes de mauvaise vision et la cécité. Les parents/fournisseurs de soins et la prise de conscience de professionnels de santé à l'examens la présentation et le traitement précoce est fortement nécessaires.

Mots Cles: œil congénital/formations adnexamals, pédiatries, parent/fourisseurs de soins.

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INTRODUCTION

Congenital eye and adnexal malformations are anatomical abnormalities or defects that present at birth. The causes of this malformations may be genetic (familial/sporadic) and non-genetic (environmental). It may present as a single entity or part of systemic syndromes (1). Eye and adnexa development becomes evident at approximately 22 days and 28days respectively (1). During these periods any changes in gene expression or "insults" will lead to disruption in organogenesis and cellular differentiation resulting in malformation with associated visual impairment, blindness, physical or mental derangement in the affected individual. In developed countries the prevalence of ocular malformation from previous studies (2-7) ranges between 0.04 to 6.8/10,000 live births. However studies from Nigeria by Lawan (8) and Adekoya et al. (9) reported prevalences of 1.7% and 9.7% respectively. The management of a patient with this condition is challenging to the parent, patient and health profession. Some of the diagnostic challenges posed by the congenital ocular malformations include; history taken, visual acuity assessment in children, examination under anaesthesia with its complications, laboratory investigations and non-availability of skill manpower to operate on these patients. Many "homes" have collapsed following emotional, financial and stigmatization experienced by the patient, parent and relatives.

To the best knowledge of the authors no study on the pattern of congenital ocular /adnexial malformations has been done previously in Kebbi State therefore, the aim of this study was to determine the pattern and presentation; and also create awareness among health workers and the public on the congenital eye/adnexal malformation.

MATERIALS AND METHODS

A 3-year retrospective study conducted between1st January 2011 through 31st December 2013. The folders of all paediatric patients aged (0-16 years) who presented at our hospital through Accident and Emergency (A&E), paediatric clinic and directly to our eye clinic were retrieved through our hospital health records unit.

Congenital ocular and /adnexa malformation is anatomical eye and or adnexa abnormality detected or present at birth. The Federal Medical Centre is the only tertiary eye centre in Kebbi State and a referral centre for the surrounding states (Sokoto, Niger and Zamfara) and countries (Benin and Niger Republic).

For this study a patient with multiple ocular/adnexal malformations, each abnormality was counted separately. From each folder information on socio-demographics (age at presentation, sex, occupation), laterality, type of congenital eye/adnexal malformation, presenting complaints and maternal age at birth of the baby were extracted. These information were entered into data collection forms. The visual acuity of the patients was assessed by ophthalmologist using various methods according to patient age. These includes: occlusion method, fixation and follow (using pen torch), picture chart and Snellen's chart. The data was analyzed with SPSS version 18 statistical software (SPSS Inc., Chicago, IL, USA).Simple descriptive statistics was done and frequency tables generated for variables. Fischer's exact test was used to test for statistical significant variables. P-value < 0.05 was considered statistically significant.

Federal Medical Centre Health and Research committee gave approval for the study.

RESULTS

Of 1110 paediatric patients seen at our eye clinic during the 3 years study periods 78 (7%) of them presented on account of congenital eye/adnexal malformations.

Males and females were equal in number (39 each) with M:F ratio of 1:1 The mean age was1.58 (SD± 0.5). The minimum age at presentation was a day old and maximum age was 16 years.

Table 1 shows the socio-demographics and laterality in the patients. The oldest patient was a 16 year old girl with bilateral congenital ptosis. Majority of the subjects were 1 year old (57.7%) at present ation to our hospital with twenty three (29.5%) being students and 5 (6.4%) house wives. Both eyes were affected in 54 (69%) patients followed by right eye 17 (22%). Congenital ptosis 12 (15.4%) was the commonest type of malformation seen, followed by congenital glaucoma 10 (12.8%).

Figure 1 shows the type of congenital eye/adnexal malformation seen in the patients. Drooping of the upper lid 12 (15.4%), followed by whitish speck in the eye 10 (12.8%) were the commonest presenting complaints. Figure 2 shows the presenting complaints of the patients at presentation. In about half of the patients, the visual acuities (VA) were not recorded. Among those with recorded VA, most of them had severe visual impairment in one or both eyes.

Table 2 visual acuity of the patients at presentation. Majority of the mothers were between 21 and 25years old (65.4%) at the birth of the patient with congenital eye/adnexal malformation, Figure 3.

DISCUSSION

The prevalence of congenital eye and or adnexa varies from developing to developed countries. Many studies (2-7) from developed countries reported a range of 0.04-6.8/10,000 live birth. In this study congenital eye/adnexal malformations accounts for 7% of 1110 of paediatric patients seen in the 3-year-study-period. This was different from studies from Nigeria by Lawan (8) and Adekoya et al. (9) who reported prevalence of 1.7% and 9.7% respectively,

while KaimboWaKaimbo et al. (10) from Democratic Republic of Congo and Eballe et al. (11) from Cameroon reported 2.2% and 6.7% respectively. The difference in these results may be because of geographic location of the study population. In our study there were equal number of males and females. This was similar to Eballe et al. (11) but different from previous studies in Nigeria (9), Hawaii (12) and China (13) where female preponderance was reported. Several authors have also reported male preponderance (8,10,14,15). In this study majority (57.7%) of patients presented at our hospital at one year and above similar to previous studies from Nigeria (9), Cameroon (11) and United Kingdom (16). But different from studies carried out in Europe where diagnosis rates of 80% were reported during the first week of life (17, 18). The difference might be because of ignorance of the parent/caregiver about the hospital's ability to manage these defects and lack of attention given to suspicious eye malformations by the health professionals.

Congenital ptosis was the commonest malformation seen and accounted for 15.4%, with 2/3 of the cases being unilateral. This was contrary to the study by Adekoya et al. (9) and Eballe et al. (11) where congenital ptosis was 2nd and 5th commonest type of malformation seen. Worldwide the prevalence of congenital ptosis is unknown. Severe unilateral congenital ptosis usually covers the visual axis which may result in Ambylopia and subsequently affect the quality of life of the affected individual. In half of our patients visual acuity (VA) were not recorded in similarity to a previous study (9). This could explain the challenges faced by eye care worker in visual acuity assessment in children. Those with recorded VA had visual impairment. This could explain the deteriorating effect of congenital eye/adnexa defects on VA. In this study Congenital glaucoma was the second commonest type of congenital malformation seen in this study it accounted for 10 (12.8%).

This was contrary to the study by Adekoya et al. (9) and Eballe et al. (11) where congenital glaucoma was 3rd commonest type of malformation seen. The average age of diagnosed congenital diagnosis in this study was 11 months this was a little high than average months reported by Adekoya et al. (9). Congenital glaucoma was diagnosed in 60% of cases within one year after birth in this study. This percentage was less than previous studies from other African countries (11, 19, 20, 21) and Lyon in France (17) in which the diagnosis was made in 73%-82% of cases within 1 year after birth and 80% before one month old respectively. The delay in diagnosis of congenital glaucoma in African might be because of lack of knowledge about the signs of the condition among the parents and caregivers. In our study, it was observed that mothers with ages between 21 and 25years had more babies with congenital ocular malformations, this was in agreement with Dutta and Bhattacharjee (22) who reported maternal age of 24 years - 29 years for congenital cataract in India but difference from a study in northern Nigeria by Onankpa and Adamu (23) who reported majority of congenital malformation in maternal age of below 20 years and above 35 years. However previous study by Nishimura (24) showed no statistically significant association between birth maternal age and congenital ocular malformation.

CONCLUSION

Congenital ptosis and glaucoma were the common types of congenital eye/adnexa defects seen in our environment. These defects were important causes of visual impairment and blindness. Health education on neonatal screening and examination, early presentation and management options is an important tool that can raise awareness and increase early hospital presentation.

Conflicts of interest: No conflicts of interest declared.

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Table 1: Socio- demographics and laterality of the patients

Variables	N (78)	%
Age range 1 day - 16years		
Age of the patient at the presentation		
Within 1year	33	42.3
1 year	45	57.7
Sex		
Male	39	50
Female	39	50
Occupation of the patients		
Dependant (child)	50	64.1
Student	23	29.5
House wife	5	6.4
Laterality		
Both eye	54	69.2
Right eye	17	21.8
Left eye	7	9

Table 2: Visual Acuity of the patients

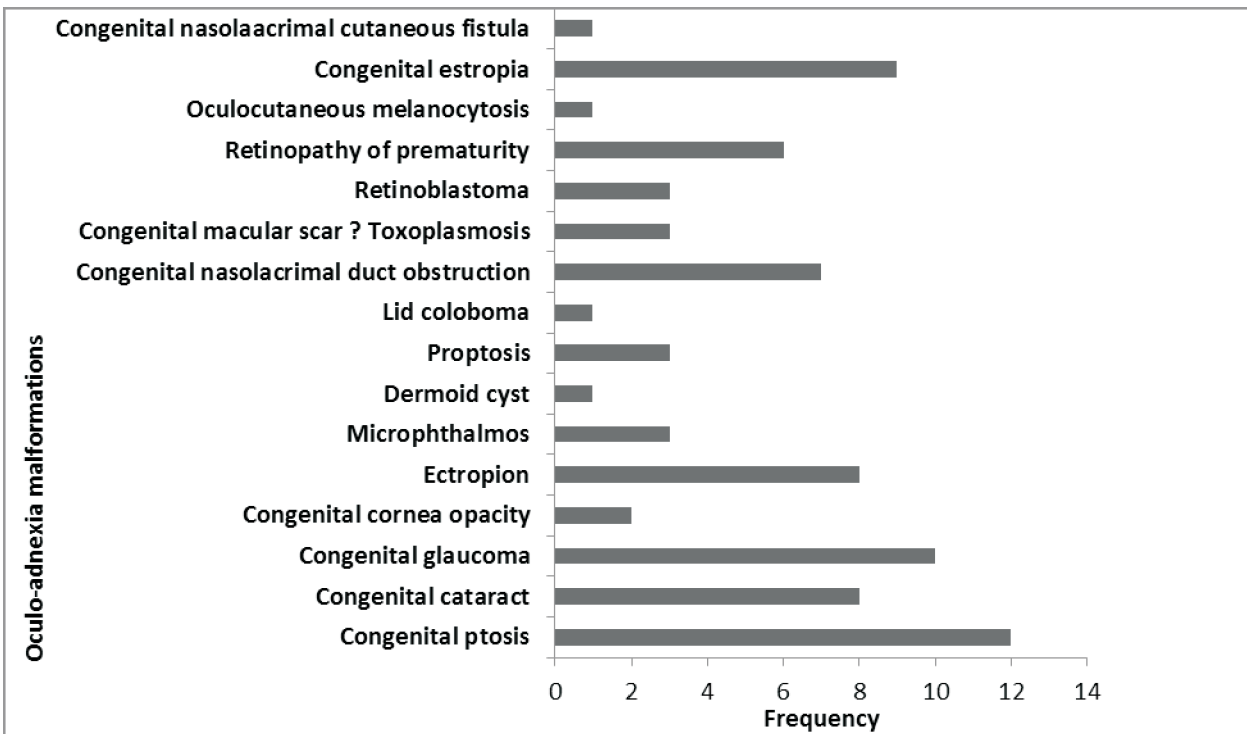| Visual Acuity (VA)	Right eye frequency	Left eye frequency
PL	18	11
HM	18	16
CF	10	19
6/60	7	6
NPL	7	7
Not asses	18	19
Total	78	78

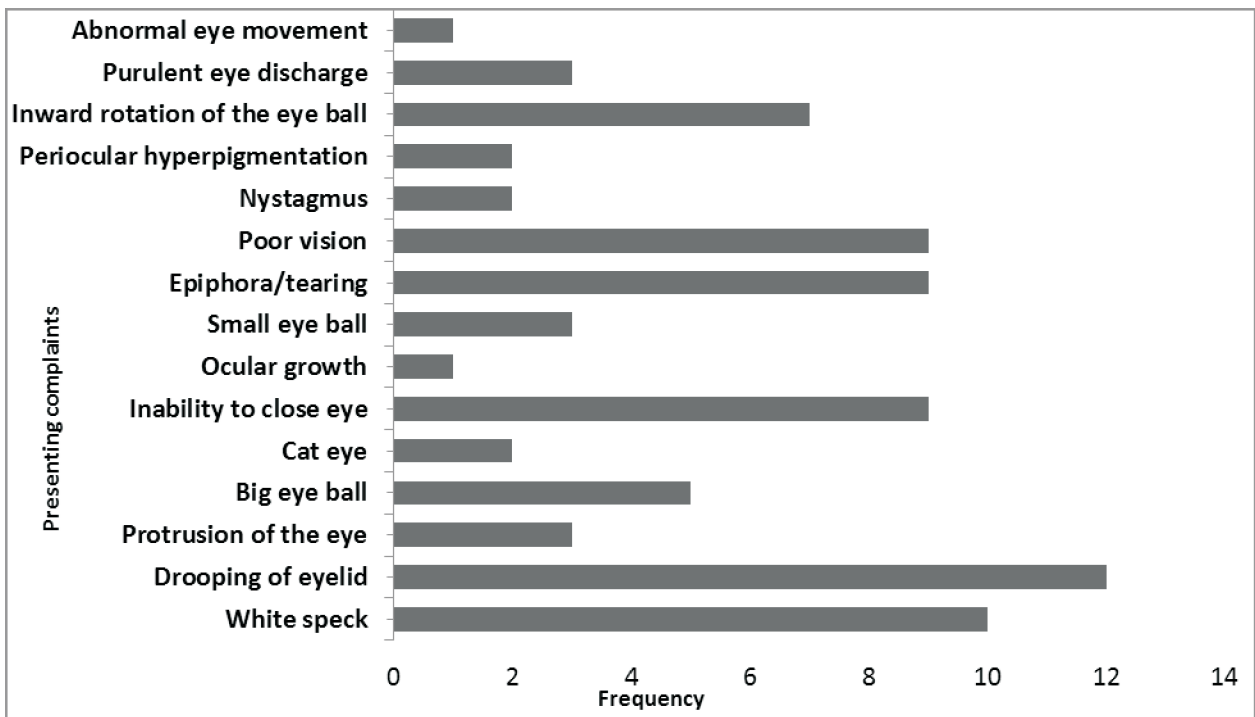Figure 1: Types of oculo-adnexia malformations presented by the cohort (patients)

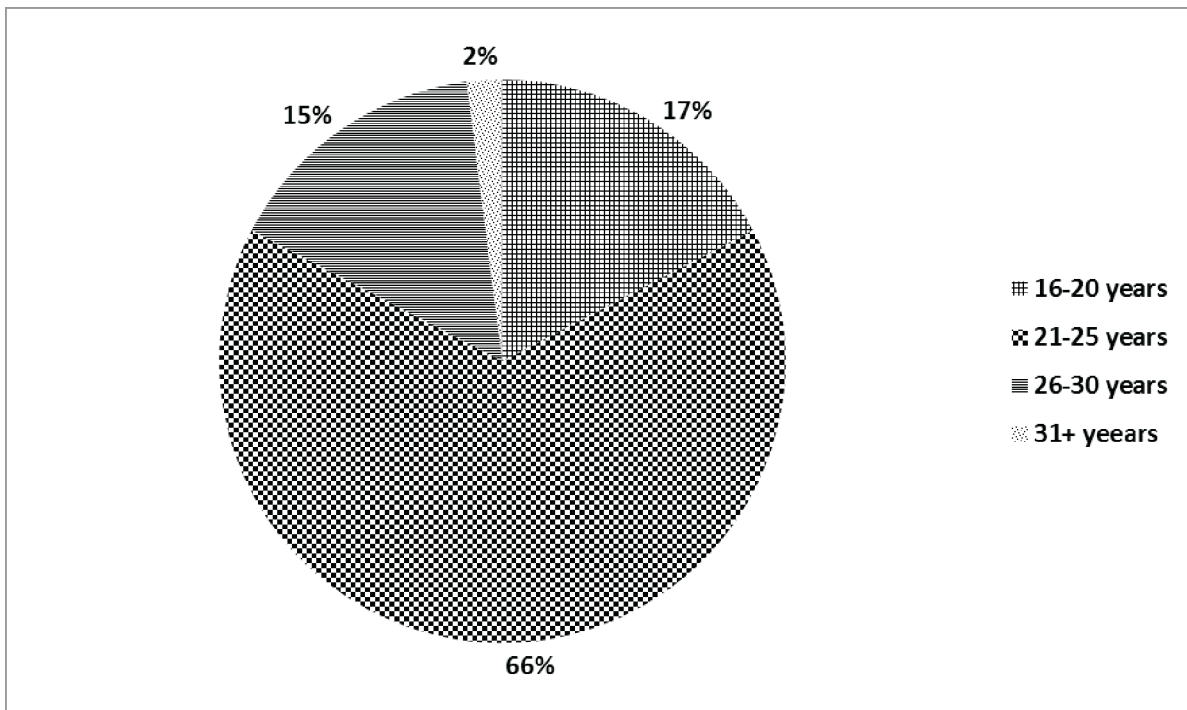Figure 2: Presenting complaints of the cohort (patients)

Figure 3: Distribution of maternal ages at birth of the cohort (patients)