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PEDIATRIC ORBITAL AND OCULAR MALIGNANCIES IN MAKURDI, NIGERIA.

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ABSTRACT

Pediatrics malignancies are not uncommon in Nigeria and like all other childhood malignancies, differ biologically from their adult counterparts. The aim of this review is to highlight the age, sex and morphological characteristics of orbito-ocular malignancies in Makurdi, North Center Nigeria. A retrospective review of all orbito-ocular specimens received in the department of Anatomic Pathology of Benue State University Teaching Hospital, Makurdi Nigeria between September, 2012 and December 2013. Clinical data obtained included sex, age and histological type. Fourteen (14) children were seen. Retinoblastoma 7(50%) was the most common pediatrics orbito-ocular malignancy with an average age of occurrence of 3.89 years and male preponderance of 1.7:1. Embryonal rhabdomyosarcoma was the second most common pediatric cause of orbito-ocular malignancy with 5(36%) cases and an average age of 6 years and a female preponderance of 4:1. One (1) case of small round blue cell tumor was found in a 10 year old male patient. The average age of pediatric orbito-ocular malignancies in Makurdi are relatively high and maybe a result of late presentation. There is a need to improve on awareness with further work into their clinical behavior and management, coupled with increase accessibility of the patient to different management modality.

KEYWORDS: Pediatric, orbito-ocular, malignancies, Makurdi.

INTRODUCTION

Pediatric care may begin periconceptionally and continues through gestation, infancy, childhood, adolescence and young adulthood.[1] The US department of Health and Food and Drug Administration in its guidelines for choosing pediatric experts reference the following phases of life (1) infancy between birth and 2 years of age, (2) childhood from 2 to 11 year of age and (3) adolescence from 12 to 21 years of age.[2] The American Academy of pediatrics has previously published a statement on the age limit of pediatrics in 1988.[3] which was reaffirmed in 2012 and identified the upper limit as 21years.

Pediatrics and especially childhood malignancies differ biologically from their adult counterparts.[4] These are especially amplified in the incidence and type of tumors, a close relationship between abnormal development and tumor induction, a greater prevalence of underlying familial or genetic gerrnline aberrations, a tendency to spontaneously regress or cytodifference and better survival and cure rates with attention increasingly focused on preventing subsequent therapy induced malignancies.[4] Many pediatric cancers tend to have a more primitive embryonal rather than a frankly anaplastic histology with features of organogenesis consistent with the site of origin, such tumors are often collectively labeled as "small round blue cell tumors".[4]

Most ocular and orbital tumors of childhood are distinct from tumor that occurs in adult. Many are congenital with early presentation.[5] The most common orbital malignancy is rhabdomyosarcoma, and the most common intra-ocular lesion is Retinoblastoma.[5]

This study is the first long term study of orbital and ocular malignancies in Makurdi North Central malignancies. It will help in illuminating orbital and ocular pediatric malignancies in Makurdi as it review its histopathological pattern in the Nigeria north central city as it relates to age and sex.

MATERIALS AND METHOD

A retrospective review of records was carried out on all orbital and ocular specimen received in the department of Anatomic Pathology Benue State University Teaching Hosptial, Makurdi, Nigeria between September 2012 and December 2019. All the biopsy specimens were formalin fixed and paraffin embedded sections and stained using hematoxylin and eosin. The reports slides and bloods were kept in the department archives. All section were diagnosed and reported by the pathologist. Information collected included patients age, site of lesson sex and histopathological diagnosis. The slides were not retrieved from review; the index diagnosis was accepted as a final diagnosis.

RESULT

A total of four thousand and hundred and ninety one (4991) specimens were received during the period of study. Sixty (60) of this were orbital and ocular specimen constituting 1.2% of the total specimen received in the department. Of the 60 specimens, pediatric orbital and ocular specimen constitute 14 (23%) of the entire orbital and ocular specimen. Seven (7) of this constituting (50%) were from male patients and 7 (50%) from female patients. The commonest orbital tumor was Retinoblastoma,(8) constituting 57% followed by embryonal rhabdomyosarcoma (5),(36%). Small round blue cell tumors (SRBT), 1, (7%). (Table 1).

The average age of occurrence for Retinoblastoma was 3.89 years and, with a male preponderance (1.7.1) Embryonal rhabdomyosarcoma has an average age of occurrences of 6 years and a female preponderance (4:1). The SRBT was seen in 10 year old male patient. (Table 2).

Table 1: Histopathological diagnosis of pediatrics orbito-ocular malignances in Makurdi.

Types of malignancy	Male	Female	Total (%)
Retinoblastoma	5	3	8 (57%)
Embryonal rhabdomyosarcoma	1	4	5 (36%)
SRBT	1	-	1 (7%)
Total	7	7	14(100%)

Table 2: Age and sex distribution of pediatric orbito-ocular malignancies in Makurdi.

	Total	Male	Female	Average age of occurrence in years(yrs)
Embryonal rhabdomyosarcoma	5	1	4	6yrs
Retinoblatoma	8	5	3	3.89yrs
SRBT	1	1	0	10yrs
Total	14	7	7	6.63yrs

DISCUSSION

Pediatrics cancers differ biologically and histologically from their counterparts occurring later in life. The main differences already alluded to in the introduction. Retinoblastoma was the commonest malignancy in this study constituting 57% of all the malignancies and followed by embryonal rhabdomyosarcoma (36%) and a case of small blue round cell tumor (7%).

Retinoblastoma is the most common primary intraocular malignancy in children.[6,7] and appear to be more common in the African child than the Caucasian.[7] Other studies.[8,9,10,11] done within Nigeria and Africa has also alluded to this. The average age of presentation was 3.89 years which is similar but a little higher to the 3.2 years and 3.25years of presentation in Ibadan.[7] and Zaria.[12] respectively. This is probably due to late presentation. Two mutation involving both alleles of RB gene at chromosome locus 13q14, are required to produce Retinoblastoma.[13,14]

Retinoblastoma and Embryonal rhabdomyosarcoma accounted for the majority of the tumors in this study. This finding differ significantly from similar work on childhood orbito-ocular malignancies done in Northern Nigeria which has Burkitt's Lymphoma as the second major ocular-orbital malignancy in childhood.[15] Rhabdomyosarcoma is the commonest soft tissue sarcoma of childhood and adolescence usually appearing before age 20.[16] Rhabdomyosarcoma is a very rare cancer with only about 250-350 cases of children below 21 years of age seen in USA every year.[17] Based on hospital studies in Nigeria, only about 1-4 cases a year are seen in Nigerian hospitals.[18] It is the most common sarcoma of children ocular tumors.[19] Most ocular rhabdomyosarcoma are from soft tissues of the orbit and on some occasions, arise in the ocular adnexal structures and even within the eye.[19] Embryonal rhabdomyosarcoma occurs in females frequently than males.[20] The female to male ratio in this study was 4:1.

A case of small round blue cell tumors was found constituting about 7% of the cases.Small round blue cell tumors is a collective histological description of most childhood tumors because of their primitive histologic appearance. The differential diagnoses of such tumor include neuroblastoma, Wilms tumor, lymphoma, rhabdomyosarcoma, Ewing sarcoma, medulloblastoma and retinoblastoma.[21]

A combination of chromosome analysis, immunoperoxidase stains, or electron microscopy is often required for diagnosis.[4]

CONCLUSION

Pediatric neoplasm of the orbito-ocular structures are not uncommon in Makurdi, Nigeria. The average age of diagnosis are usually relatively high as a result of late presentation. There is a need to improve on awareness with further work into their clinical behavior and management coupled with increase accessibility of the patient to the different management modalities.

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