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ORIGINAL ARTICLE

Histopathological Features of Tumours of the Orbit and Adnexia seen in Korle-Bu Teaching Hospital

Les caractéristiques Histopathologiques des Tumeurs de l'orbite et Adnexia vu Dans Korle-Bu Teaching Hospital

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ABSTRACT

BACKGROUND: **Reports of histological types of orbital and adnexal tumours are few in the West African Sub-region and non-existent in Ghana.**

OBJECTIVE: **This study was to determine and report the histological types of orbital and adnexal tumours seen in the eye unit in Korle-bu Teaching Hospital**

METHODS: **This was a retrospective study of orbital and adnexal tumours in a referral orbit and oculoplastic clinic in a teaching hospital in Ghana. Biopsy specimen taken from the orbit, conjunctiva and eyelids of patients referred to the orbit and oculoplastic clinic of the eye unit between November 2005 and October 2009 were mailed to Royal Hallamshire Hospital Sheffield, UK for histological evaluation. Histology reports of these biopsy specimens were reviewed and analysed for types of *tumours,* frequency of occurrence, sex and age of patients.**

RESULTS: **A total of 190 histological specimen were taken. One hundred and four (54.7%) were malignant tumours, 53 (27.9%) benign tumours and 12 (6.3%) inflammatory lesions. Eighteen (9.5%) of biopsies taken were mostly degenerative conditions.**

CONCLUSION: **Our findings show, as reported from other centres, that squamous cell carcinomas are the commonest cancers of the orbit and adnexia, while malignant melanomas account for only 3.8% of cancers. WAJM 2012; 31(1): 58–62.**

Keywords: **Orbital tumour, squamous cell carcinoma, malignant melanoma.**

RÉSUMÉ

CONTEXTE: Les rapports de types histologiques de tumeurs orbitaires et annexielle sont quelques-uns dans le ouest-africaine sous-région et inexistante au Ghana.

OBJECTIF: Cette étude était de déterminer et de signaler les types histologiques de tumeurs orbitaires et annexielle vus dans l'unité de l'œil dans Korle-Bu Teaching Hospital

MÉTHODES: Il s'agissait d'une étude rétrospective de tumeurs orbitaires et annexielle sur une orbite de référence et de la clinique oculoplastie dans un hôpital d'enseignement au Ghana. La biopsie échantillon prélevé dans l'orbite, de la conjonctive et les paupières des patients visés à l'orbite et de la clinique oculoplastie de l'unité de l'œil entre Novembre 2005 et Octobre 2009 ont été envoyés à l'Hôpital Royal Hallamshire Sheffield, Royaume-Uni pour l'évaluation histologique. Rapports d'histologie de ces biopsies ont été examinées et analysés pour les types de *tumeurs* , la fréquence d'occurrence, le sexe et l'âge des patients.

RÉSULTATS: Un total de 190 échantillons histologiques ont été prises. Cent quatre (54,7%) étaient les tumeurs malignes, 53 (27,9%) des tumeurs bénignes et 12 (6,3%) des lésions inflammatoires. Dix-huit (9,5%) des biopsies prélevées étaient pour la plupart des conditions dégénératives.

CONCLUSION: Nos résultats montrent, tel que rapporté dans d'autres centres, que les carcinomes spino-cellulaires sont les cancers les plus fréquents de l'orbite et adnexia, tandis que les mélanomes malins ne représentent que 3,8% des cancers. **WAJM 2012; 31 (1): 58–62.**

Mots clés: Tumeur orbitaire, le carcinome spinocellulaire, le mélanome malin

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Abbreviations: CLL, Chronic Lymphocytic Leukaemia; **EBV,** Ebson Barr Virus; **SCC,** Squamous Cell Carcinomas

INTRODUCTION

The incidence and distribution of orbital and adnexal tumours vary among races. It is known for example that squamous cell carcinomas are more common among Africans than Caucasians[1,2] and melanomas are more common among Caucasians.[3,4] Clinical presentations also vary, with tumours in Africa presenting much later and in advanced stages. Accurate histo-pathological diagnosis and grading are essential in the management of these tumours.

The orbit and oculoplastic clinic of the eye unit of Korle-Bu Teaching Hospital, Accra, Ghana, was established in 2005. The clinic receives referrals from other clinics within the eye unit of the hospital and from eye clinics all over Ghana. Retinoblastomas are managed by the paediatric ophthalmologists. Intraocular tumours were not referred to this clinic.

In 1996, Ntim Amponsah *et al*[5] reviewed 56 cases of ocular tumours in five years but only 30 of these were histologically confirmed. There are however a few studies in the sub region, mainly from Nigeria[6,7,8] defining histologically confirmed studies of orbito-ocular tumours. All of these reports combined ocular and adnexal tumours. No study describing histological features of orbital and adnexal tumours has been reported in Ghana to our knowledge.

This was a retrospective descriptive study of all tumour masses referred to the orbit and oculoplastic clinic with the objective of determining the different types of histologically confirmed orbital and adnexal tumours between November 2005 to October 2009.

SUBJECTS, MATERIALS AND METHODS

Histology reports of biopsy specimens taken from patients referred to the clinic between November 2005 and October 2009 were reviewed.

Inclusion Criteria

All patients seen in the orbit oculoplastic clinic of the Korle-Bu Teaching Hospital, Accra with tumours of the eyelids, conjunctiva or orbit had biopsies of their tumous taken. Biopsies were either excisional or incisional depending on the clinical presentation. Only lesions suspected to be neoplasms were biopsied.

Exclusion Criteria

Retinoblastomas and Burkit's lymhpomas were excluded from the study as these were managed in the paediatric eye clinic. Tumours suspected to be of intracranial or maxillofacial extension into the orbit were excluded as these were referred to be managed by the maxillofacial surgeons. Eye lid tumours suspected to be neurofibromatosis were also referred to plastic surgery without biopsy. Obvious degenerative lesions were excluded.

Histological Examination of Specimen

Specimen were preserved in formalin, packaged in water-tight containers and sent by regular mail to the National Specialist Eye Pathology Service, Department of Histopathology, Royal Hallamshire Hospital Sheffield (UK), for histopathological analysis. Briefly, ocular tissue was fixed in standard 10% buffered formalin and shipped to the Ophthalmic Histopathology Service in Sheffield. On receipt, the tissue was described macroscopically, cut, representative pieces taken and these processed to paraffin wax. 4 micron sections were cut and stained with haematoxylin and eosin using standard methodology. If required, ancillary investigations, including tinctorial stains, immunohistochemistry and electron microscopy were conducted. The report was then sent by email and later the hard copy was sent by post to Accra.

Clinical Data

Other data collected include age, sex, laterality and location of tumour.

RESULTS

The total number of biopsies taken from tumour masses were 190. There were a total of 89 (46.8%) males and 101 (53.2%)females.

Of the 190 masses, 104 (54.7%) were malignant tumours, 53 (27.9%) were benign tumours, and 12 (6.3%) were inflammatory lesions. Eighteen (9.5%) were suspicious lesions biopsied but found out to be mostly degenerative conditions. Of the total of 169 tumours masses, N 104 (61.5%) were malignant, N53(31.4%) benign and 12 (7.1%) inflammatory.

Of all the biopsies taken, 87(45.8%) were from the conjunctiva, 68(35.8%) from the orbit, and 35(18.4%) from the eyelid. Tables 1, 2,and 3 show the tumour types from the conjunctiva, orbit and eyelid respectively.

Table 1: Histological Types of Conjunctival Tumours

Tumour Types	No	%
Squamous cell carcinoma *in situ*	34	39.2
Invassive squamous cell carcinoma	15	17.4
Pterygium/pengueculum	14	16.2
Viral pappiloma	5	5.7
Naevus	4	4.7
Malignant melanoma	2	2.3
Sabaceous carcinoma	1	1.1
Lymphoma	1	1.1
Lipodermoid	1	1.1
Nodulr fascitis	1	1.1
Limbal dermoid	1	1.1
Cysts	3	3.4
Granulation tissue	4	4.5
Cartilagenous choristoma	1	1.1
Total	**87**	**100**

Table 2 Histological Types of Orbital Tumours

Tumours	No.	%
Invasive squamous cell ca	25	36.2
Lymphomas	10	14.7
Lacrimal gland tumours	5	7.5
Benign adenoma 2		
Adenoid cystic carcinoma 3		
Rhabdomyosarcoma	5	7.5
Alveolar 2		
Alveolar + embryonal 2		
Embryonal with anaplasia 1		
IgG4 disease	3	4.5
Angular dermoids	5	7.4
Haemangiomas	5	7.4
Complex 4		
Angiolymphoid hyperplasia 1		
Vasculitis	3	4.5
Malignant melanoma	2	2.9
Others	5	7.4
Total	**68**	**100.0**

Others: **schwanoma, mesenchymal chondrosarcoma, orbital fat prolapse rosai dorfman disease, non caseating granulation tissue**

Table 3: Histological Types of Eyelid Tumours

Tumour	No.	%
Apocrine Hydrocystomas	10	28.7
Invasive Squamous Ca	3	8.5
Papillomas	8	22.8
Viral-6		
Basal Cell-2		
Chalazion	2	5.8
Naevus	3	8.5
Haemangiomas	3	8.5
Capillary 1		
Complex 1		
Lymphangioma 1		
Xenthelasma	3	8.5
Sabaceous Ca	1	2.9
Vasculitis	1	2.9
Sarcoidosis	1	2.9
Total	**35**	**100.0**

Figure 1 shows the age distribution of all malignant tumours. There was no sex predilection for malignant tumours (Male:Female is 1:1). Fifty - two (5.2%) of the malignant tumours were in the paediatric age group of 0–15 years. In the adult age group (15+ years), of the 96 cases, 61(64%) were between the ages 26 and 55 years with only 16.7% being above 65 years. Squamous cell carcinomas formed the majority of malignant tumours. About 74% (77 of 104) of all malignant tumours and 45.6% (77 of 169) of all tumours were squamous cell carcinomas. Of the 77 squamous cell carcinomas, 34 were in situ carcinoma of the conjunctiva, 15 were invasive carcinomas limited to the conjunctiva, 25 invaded the orbit from the conjunctiva and three were limited to the eyelids.

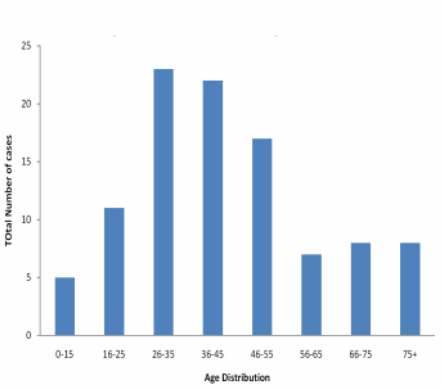

Fig. 1: Age Distribution of Malignant Tumours

Twenty-four (24) pediatric patients were included in the study: five haemangiomas, six papilomas, three rhabdomyosarcomas, five angular dermoids, one limbal dermoid one lymphoma, one Rosai-Dorfman tumour and two conjunctival naevus.

There were eight vascular tumours, five were females and three males. Five were in the paediatric age group, between 7 and 14 years. The other three were 25, 33 and 41 years. Two were limited to the eyelids, one was from the eyelid extending into the orbit while five were limited to the orbit.

There were five rhabdomyosarcomas occurring in patients aged 2,5, 3, 13, 18 and 20 years. Four were males and one female. There was one malignant mesenchymal chondriosarcoma in a female 20 years old.

There were four malignant melanomas; one was from the bulbar conjunctiva, one from the palpebral conjunctiva and two were orbital, both extending from the uvea.

There were 11 lymphoproliferative tumours; one conjunctival and 10 were orbital sometimes with eyelid involvement. One of these with a high grade T cell lymphoma of anaplastic type was in a 23 year old patient. The rest were of ages ranging from 47 to 69 Years. Of these, eight were B cell non-Hodgkin's lymphoma – extranodal marginal zone lymphoma of MALT type (MALToma – WHO classification). Three of these were bilateral. The other two were peripheral T-cell lymphoma: Ebson Barr virus (EBV) positive extra-nodal NK/T-cell lymphoma, of nasal type (WHO classification) and chronic lymphocytic leukaemia (CLL)/ chronic lymphocytic lymphoma - WHO classification. The CLL presented as a conjunctival lesion in a 65 year old male patient.

Inflammatory lesions include granulation tissues, vasculitides (Wagener's granulomatosis), IgG4 sclerosing disease, sarcoidosis and nodular fasciitis (Tables 1, 2,and 3).

DISCUSSION

General Overview

This study was done in a centre where referrals were from all over the country. With assistance from an ocular histological centre the variety of tumours recorded has increased compared to an earlier study in the same centre.[5] Coupled with the fact that all suspicious lesions are biopsied, diagnosis of ocular surface and adnexal tumours increased. Nine and a half percent (9.5%) of all lesions biopsied were negative for tumour. These were mainly conjunctival lesions suspected to be carcinomas.

The previous study[5] in this eye unit reported only 58 tumours over a five 5 year period and this included 29 cases of retinoblastomas but our series do not include retinoblastomas. It is interesting to note that few squamous cell carcimomas (SCC) were reported then. In this series SCC are in the majority forming about 74% of all malignant tumours. These differences may be accounted for by the establishment of a subspecialist centre encouraging referrals from all over the country. The aggressive histological diagnosis of suspicious lesions may have contributed in part. In a four and a half year review of histologically confirmed orbito-ocular tumours in a teaching hospital in Enugu, Nigeria in 2005[7] 43 tumours were reported. Of these 26 % were retinoblastomas and only five squamous cell carcinomas were reported. In Ibadan, a 9 year review in 1971 reported 191 histologically proven tumours.[6] Of these, 59 were from the orbit and adnexia (excluding burkits lymphoma). This series reported 15 cases of squamous cell carcinomas. Several other studies show a similar trend of records of few ocular surface and adnexial tumours in African blacks.[9–12] Our study recorded 169 tumours (104 of which were malignant) of the orbit and adnexia. There were 43 cases of SCC and 34 cases of SCC in situ. These results are similar to a study done in Uganda between 1961 and 1966[13] where there were 188 orbital and adnexal tumours with 57 SCC of the conjunctiva and eyelids and nine SCC *in situ.*

Squamous Cell Carcinomas

The incidence of SCC has seen an upward trend with the AIDS pandemic.[14–16] The high numbers of ocular surface carcinoma in situ and squamous cell carcinomas recorded was possible due to our policy of taking biopsies of any suspicious lesion. This has resulted

in some negative biopsies. However the gains outweigh the inconveniences. Orbital extension of conjuctival SCC was found in 25 cases. Reports of orbital invasion of conjunctival SCC are scanty.[2,17] In a study done in Turkey over a 10 year period, SCC accounted for 17 of the conjunctiva tumours invading the orbit.[18] In Saudi Arabia, over a period of ten years, 28 conjunctival SCC were found to invade the orbit.[2] Although conjucntival SCC is known to be relatively non aggressive tumour especially in Caucasians[3] a few studies from tropical and subtropical regions suggest that in these areas, this type of cancer behaves in an aggressive fashion and tends to affect younger individuals.[1,2] Our study shows similar trends. This stresses the need for early diagnosis and management of earlier lesions.

Malignant Melanomas

Ocular malignant melanomas are rare in all races and even less common among the black race.[3] Orbital extensions are even rarer. In a study in South Africa[3] over a 25-year period, out of 153 cases only eight were in blacks. In the blacks, one patient had uveal melanoma, three had extensive orbital extension and four were limited to the conjunctiva. In our series of 4-year review, there were two uveal melanomas both extending into the orbit and two limited to the conjunctiva. There has been no record of histologically confirmed uveal melanomas from the retina clinic in our unit. Other studies in Nigeria[11] and Uganda[13] confirm the rarity of uveal melanomas. The late presentation of tumours in developing countries accounts for the common occurrence of metastasis and wide extension of these tumours.

Age Distribution of Tumours

The age distribution of ocular and adnexal malignant tumours is known to be bimodal with peaks occurring during early childhood and again during adulthood.[19] The peak in childhood is usually accounted for by retinoblastoma and burkits lymphoma. Since our study did not include these tumours, the age distribution is uni-modal ranging between 26 years and 55 years with a peak at a relatively young age group of 26–35 years. Almost all of this is accounted for by invasive SCC and SCC *in situ*. This is consistent with other studies where conjunctival SCC is seen to be an aggressive disease in tropical and subtropical regions, and tends to occur in younger patients.[3,14]

Rhabdomyosarcomas

There were two cases out of five rhabdomyosarcomas occurring at ages 18 and 20 years. Most orbital rhabdomyosarcomas occur before the age of 9 years.[20,21] The majority are embronal and the alveolar type caries the worst prognosis.[22] The pleomorphic type are rare in the orbit. Four of our cases were mainly alveolar (two were mainly alveolar with areas of embryonal). Only one was embryonal with anaplasia. These were thus atypical histological presentations consisting of histologically aggressive types.

Rare Tumours

In our series, there were a wide variety of cases seen, some uncommon. One case of Rosai-Dorfman disease in a 10year old involving the orbit and paranasal sinuses, a case of malignant mesenchymal chondrosarcoma and three cases of IgG4 orbital diseases were seen. These cases are rare in the literature. There were also a wide variety of histological types within the different types of tumours.

Conclusion

We have reported histopathological types of orbital and adnexal tumours seen in Ghanaians in our referral centre. The most common were SCC. Routine histological diagnosis of orbital and adnexal tumours uncovered the some rare tumours and varied histological types of some tumours. This improves the management of these patients.

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Conflict of interest – None

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