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SPONTANEOUS REGRESSION OF RETINOBLASTOMA: A CASE-REPORT AND LITERATURE REVIEW

A case-report of the rare phenomenon of spontaneous regression in retinoblastoma which occurred in a Nigerian girl at the age of 2 is reported. At birth, the child had divergent squint of the right eye. This was followed by leukokoria and progressive enophthalmos of the globe 1 and 6 months later respectively. With no history of trauma or any form of treatment, spontaneous involution of the globe occurred at about the age of 2. Six months later, leukokoria was again noticed in the left eye. At presentation, a fusiform mass was noted. Histopathology report of the left enucleated eyeball confirmed retinoblastoma.

Key Words: Retinoblastoma, Retinoma, Spontaneous regression.

This is the third report and the first detailed literature review on the spontaneous regression of retinoblastoma in Nigeria. The first two cases were reported by Ayetey in 1988.

CASE REPORT

N.A., a 3-year-old girl was admitted via the Ophthalmo-\nClinic of the University of Nigeria Teaching Hospital, Enugu with a one year history of left leukokoria and progressive enophthalmos of the globe. In addition, the right globe was phthisical. A history of right divergent squint prior to birth followed a month later by leukokoria, progressive increase in size of the globe four years of age of 6 months and spontaneous "rupture" about 2 years of age was given by the mother. There was no mention of history of trauma or treatment prior to the investigation.

The patient is the fourth of 5 children from healthy parents. All but one of the siblings are alive and well (the second died from unexplored cause around the age of 2). There is no history of consanguinity or similar illness in the family.

Growth examination revealed a well-nourished child who was not pale and in a normal respiratory distress. The central nervous system examination revealed a satisfactory involvement by the tumor otherwise the other systems were clinically normal.

Orbit examination revealed right phthisis bulbi and a left fungating mass. The results of laboratory investigations were as follows:

INTRODUCTION

Spontaneous regression is a rare manifestation of the retinoblastoma gene, the common ones being retinoblastoma and malignant retinoblastoma. Galle et al. have found that spontaneous regression is expected to occur in 2% of individuals with the retinoblastoma gene 1.

This phenomenon was first reported by Fuchs in 1931 and over the years the concept of spontaneous regression of retinoblastoma has been widely quoted and confirmed upon. In the two distinct situations in which spontaneous regression have been identified, rapidity and eyes with minimal residual tumor mass good vision have been noted.

The recent addition of extracapsular invasion in the finding as seen in parents of a child with retinoblastoma. Many mechanisms to explain the finding of phthisis bulbi et retinoblastoma have been put forward, the most popular of which is vascular obstruction to both tumor and eye by tumor growth. In addition to the controversy on the pathogenesis of the type II pattern of spontaneously regressed retinoblastoma, many issues have been suggested for it. They are retinoma, retinocygin, spontaneous cases of retinoblastoma and benign retinoblastoma. Spontaneous cases of retinoblastoma which seems to be the most appropriate name was suggested at an international retinoblastoma Symposium 1.
explain the phthisis bulbi seen in this type I regeneration pattern of retinoblastomas. The most widely accepted pattern of obstruction is the complete occlusion of the eye and the results as a result of tumor growth.

In other words, several retinal alterations occur by a tumor cell resulting in phthisis bulbi as well as severe damage of the eye ball the later being as a result of the systemic necrosis of all the other non-tumor tissue of the organ.

One evidence in support of this view was put forward by Aschan and James "where they found the tumor emerging through central retinal vessels in a pattern with phthisis bulbi and retinal necrosis".

Coffin et al. later confirmed this mechanism.

While a whole round mass of blood supply is present in the phthisis bulbi retinoblastoma occurs beyond its margins. In other words, when the tumor has spread to the optic nerve before the continuous regeneration, since the optic nerve has an extracranial blood supply it continues to grow to the presence of a phthisis bulbi eye.

It has also been suggested that immunological process could be asymptomatic for this spontaneous regression. To support this, Mehl and Reed reported the presence of spontaneous regression in a case of retinoblastoma, which was subsequently cured by a phthisis bulbi in immunologic reasons. This proposal was not confirmed by the pathological features in a phthisis bulbi with retinoblastoma where there was no evidence of an immune reaction. Other suggestions in support of the latter conclusion included the presence in the phthisis bulbi of patients with retinoblastoma of specific systemic elements and the association of spontaneous regression with certain genotypes. Two theories were all related to different works.

One percent of spontaneous regression of retinoblastomas is that most commonly seen in the fusion of a child with retinoblastoma. Even in the absence of a histopathological confirmation these lesions were ascertained to be spontaneous retinoblastomas from the characteristic appearance. Again, in support of this was the fact that a great percentage of the patient also gave a personal history of its occurrence.

The characteristic features of the type II bulb are as follows: transitory retinal moist, "congealed cheese" calcification and disappearance of the pigment epithelium. According to Liogee et al., histologically, these lesions were made up of homogeneous-looking cells with several flowers and no anisocoria or anisokoria activity. There was an evidence of invasion into the choroidal and optic nerve. Also the spontaneous and retinal pigment epithelial hyporeactivity could also be seen. Smith in his series noted this feature was no Phthisis bulbi in the presence of a pattern of chronic retinoblastoma exenteration but these lesions continued of differential neural elements.

On the issue of what these lesions really were, many controversial suggestions have been made. Following, this their retinoblastoma appears to resemble this soon in some retinoblastoma cells after...
In the treatment of lesions Griffiths expresses a form of recognition. However, Gullott et al. noted that in their studies where the only two cases out of 34 patient showed regression did occur 2 years of treatment. Again, in further support of their opinion that this lesion was not a manifestation of regression at all, Griffiths et al. expressed that statistical evidence had also been observed by others and other workers. On the contrary, they had suggested that these lesions were "morbidly" different from the conditions described in the literature, leading to the hypothesis of regression in different conditions, leading to a hypothesis of regression in different conditions. In addition, a suggestion was made that these factors may not easily have multiple possibilities as could be placed at the most benign end of the spectrum of malignant neoplasms.

The treatment of the malignant melanoma was an interesting area of the eyes with such type IIB lesions vascular in character and has not been implicated in this particular situation.

Several names have been suggested for this particular lesion. "Recurrent" was suggested by Griffiths et al. and others. For example, in a paper discussing the lesion it was suggested that this lesion was a "morbidly" different from the condition described in the literature, leading to the hypothesis of regression in different conditions. In addition, a suggestion was made that these factors may not easily have multiple possibilities as could be placed at the most benign end of the spectrum of malignant neoplasms. From the literature of the malignant melanoma, an interesting area of the eyes with such type IIB lesions vascular in character and has not been implicated in this particular situation.

CONCLUSION
Spontaneous regression of malignant melanomas is not uncommon in the literature for the condition. The lesion is vascular and has not been implicated in this particular situation.

REFERENCES

Afterglow, at the time of presentation, there was just a phlegmatic eye on the right, and some for a follow-up of the patients for any other in 4 out of 3 malignant melanomas which has a very poor prognosis.


