Case Report

Paediatric Ocular Sarcoidosis in Calabar, Nigeria: A case report

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Abstract

Background: Sarcoidosis is a granulomatous inflammatory disorder of unknown origin that affects multiple organs, most commonly the lungs. Ocular involvement is documented, occurring in 13–79% of patients with systemic disease. However, paediatric sarcoidosis is rare, with an unclear global prevalence. Diagnosis relies on clinical presentation, radiologic findings, and exclusion of differential diagnoses, with biopsy confirmation when possible. We report a case of paediatric ocular sarcoidosis in Calabar, Nigeria with initial symptoms of respiratory disease, presentation and treatment in the paediatric chest clinic one year prior to diagnosis.

Conclusion: Sarcoidosis can manifest in a variety of ways and may be an under-reported disease in Nigeria. Corticosteroids are a useful first line treatment option. This case highlights the importance of

a high index of suspicion, early recognition and comprehensive ocular assessment in paediatric sarcoidosis to prevent and appropriately manage blinding complications.

Key words

Paediatric, Ocular, Sarcoidosis, Calabar.

Introduction

Sarcoidosis can be defined as a granulomatous disorder of unclear origin that affects multiple body systems but most frequently the lungs [1-6]. It is a chronic inflammatory disorder with environmental triggers (proposedly, organic materials like pine leaves and certain seeds), inorganic materials (e.g., aluminium, beryllium and fibreglass) and microorganisms (e.g., Propionibacterium acnes, Mycoplasma and Schistosoma) in addition to predisposing genetic factors. The deficiency of certain nutrients; particularly Vitamin D may increase the risk of developing sarcoidosis. A diagnosis of exclusion; this disease is frequently characterized by non-caseating granulomas present in affected body structures [1, 7]. An idiopathic disease, lung involvement where present, is said to often be associated with extrapulmonary disease [1, 2, 7-9].

While the presentation of sarcoidosis can be non-specific, variable, acute of chronic, self-limiting or widespread, it is reported to affect any part of the ocular structures and ocular adnexa [1, 2, 10]. This makes the assessment of the eyes in this disease important for effective management of ocular sarcoid [1, 2, 4, 8, 10].

Generally, the prevalence of sarcoidosis ranges from 4.7 to 64 per 100,000 population globally while extrapulmonary involvement is said to be the presenting feature in 30-50% of patients [1, 2, 11], with ocular involvement estimated to affect 13 to 79% in those with systemic sarcoidosis [1, 2, 4]. Some reports suggest that eye involvement is more common among Japanese and Finnish patients [12]. In the UK, an estimated prevalence of 20 per 100,000 has been reported. In Europe, a more variable prevalence of 3-50 per 100,000 population with reports from Sweden presenting a prevalence of up to 641 per 100,000 population where autopsy studies are included [1, 7, 13]. In the USA, racial studies suggest that sarcoidosis is more prevalent among people of negro descent with a cumulative incidence estimate of 2.4% for Black people and 0.85% for White people [1, 13]. Sex predilections are less clear. Sarcoidosis has been associated with a 0.29/100,000 morbidity rate [4], and there are no generally accepted diagnostic guidelines [1, 7].

While less common than in the adult population, the global prevalence of childhood sarcoidosis is essentially unknown. It is said to have an approximate incidence in children younger than 15 years of age in Denmark of 0.22-0.25 per 100,000 children per year [1, 4, 13]. Where it is found in children older than 5 years (> 5 years to 16 years), the presentation is said to be likened to that found in adult populations. Patients will typically present with an unexplained fever, malaise hilar adenopathy as well as changes in the lungs radiologically. However, when sarcoidosis occurs in children <5 years of age it is known as Blau syndrome (where there is a positive family history)/early onset sarcoidosis (BS/EOS) [1, 9]. In this set of patients, diagnosis is often delayed as typical lung disease is not seen but the eyes (uveitis), joints (polyarthritis) and skin (rash) are usually involved [1, 14, 15]. Other manifestations (seen in >50% of patients) in this age group include the following: [1, 9]
arterial hypertension, bronchial granulomas, chronic renal failure, erythema nodosum, interstitial nephritis, leukocytoclastic vasculitis, hepatic granulomas, hepatosplenomegaly, pericarditis and pulmonary embolism.

We present a report of paediatric ocular sarcoidosis in Calabar, Nigeria. Diagnosis was made based on clinical, radiologic findings and histopathological findings in keeping with the disease. Other sarcoid-like diseases were ruled out during the clinical work-up including tuberculosis, idiopathic granulomas and cancers including lymphomas.

Case report

The index 11-year-old male patient first presented to the Department of Ophthalmology, University of Calabar Teaching Hospital (UCTH) in February, 2020 with complains of bilateral red eyes of about a months' duration and occasional foreign-body sensation. There was no associated blurring of vision, ocular itching, discharge, eye lid swelling, tearing, history of past ocular trauma or use of eye medication and no history of a similar incident. His family history was unremarkable. He had previously been repeatedly managed in the Children Out-Patients Unit- Department of Paediatrics, for episodes of difficulty in breathing, associated occasional cough and tiredness of unknown cause. His pre-morbid history also included right knee pain, painless nodules on both lower limbs, a poor appetite and weight loss spanning a period of about 1 year prior to presentation in the Eye Department of the UCTH. These symptoms abated temporarily to recurr over the said period of time. There were no other significant contributory histories.

Notable findings following detailed ophthalmic evaluation were; visual acuity (VA) at initial presentation 6/6, N_5 oculus uterque (OU), intraocular pressure (IOP)s was 12 mmHg OU, and he had papillary conjunctivitis and reduced tear break up time. He was then managed as a case of allergic conjunctivitis, commenced treatment and he returned a week later with all symptoms resolved.

After having been symptom free for about 4 months following initial eye treatment, he represented to the Children's Eye Unit with redness, associated eye pain, and tearing affecting his right eye, of one week duration. Significant ophthalmic findings on examination included the following as per **Table – 1, 2**.

A diagnosis of right granulomatous panuveitis (? cause) with uveitic cataract was made. For this, patient's initial medical management was with the paediatric ophthalmology and medical retina units.

The diagnosis was then modified to paediatric ocular sarcoidosis with right pan uveitis and uveitic cataract.

The paediatric respiratory physician and endocrinologist constituted part of his medical management team and following the team review, he was diagnosed with pulmonary sarcoidosis and the decision to commence tablets prednisone as 1mg per kg was made. He received an induction dose and had this medication tapered as appropriate. In addition to the systemic steroid, he received oral calcium, vitamin D, potassium and Omeprazole. He made significant clinical improvement and gradually became significantly less fatigued and his chest symptoms abated. The mediastinal lymph nodes were no longer demonstrable on repeat chest XRAY. He however gained 15kg over 3 months (now 62kg, BMI 30.0) and had in addition, abdominal straie. He was declared to be at risk of impaired glucose control. During this period, the VA of his right eye decreased to PL while that of his left eye remained normal at $6/5^{-2}$.

Over the subsequent 3 months, he received 2 doses of 20mg sub-tenons Depomedrol to control the posterior uveitis and had cataract surgery with IOL on the right eye 6 months after commencing the systemic steroid. Post operatively, he received gutt 1% Cyclopentolate, gutt 0.1% Prednisolone, gutt 0.5% Moxifloxacin on his right eye according to standard protocol. His VA improved to 6/7.5 after a pre-operative VA of PL in the same right eye.

Table - 1: Ocular Findings of Patient.

OD	**Parameter**	**OS**
5/60, N_{48}	**VA**	$6/5^{-2}$ N_5
12mmHg	**IOP**	12mmHg
Mutton fat KPs^{+4} Early band keratopathy	**CORNEA**	Normal
$Flare^{++}$, $cells^{+}$	**ANTERIOR CHAMBER**	Normal
Irregularly round, 360^0 posterior synechiae and APD.	**PUPILS**	Round regular briskly reactive
Cataract (anterior capsular, cortical, posterior subcapsular), pigments on the anterior lens capsule.	**LENS**	Clear
Snow banking	**VITREOUS**	Normal
No clear view	**FUNDUS**	Normal
Supraciliary effusion, blunted ciliary processes, vitritis, thickening of the choroid, no retinal detachment.	**OCULAR B-MODE ULTRA SOUND SCAN USING 10 MHz HIGH-FREQUENCY LINEAR PROBE**	Normal

Table - 2: Summary of findings from other investigations are as listed:

Item	**Value**
Lymphocytes	1 x 10^9/L
Neutrophils	37 (50-80)
Complete blood count	Haemoglobin 13g/dl, Total white cell count 2 x 10^9/L, Neutrophils 37%, Lymphocytes 34%, Monocytes 6%, Platelets 290,000/mm^3, Erythrocyte sedimentation rate (ESR) 10mm/h.
Blood culture	Normal, no pathogen seen
Vitreous tap	No pathogens seen
ELISA for HIV.	Negative
Serology for hepatitis A, B, C	Negative
VDRL.	Non-reactive
Urinalysis	Normal
Serum electrolyte, urea and creatinine	Serum calcium 12.5mg/dl.
Antinuclear antibodies (ANA)	Negative
Anti-neutrophilcytoplasmic antibodies (p-ANCA and c-ANCA)	Negative
Mantoux test	Negative
RF.	Positive 161u/l (61u/l)
Skin biopsy (nodules)	Noncaseating granuloma
Echocardiogram	Normal heart findings
Plain Chest XRAY	Paratracheal opacities- bilateral mediastinal lymphadenopathy, and right upper lobe pneumonitis
Plain XRAY of long bones and skull	Normal findings
Peripheral lymph nodes	No peripheral lymphadenopathy

Gutt 0.1% Cyclosporin A was subsequently introduced to his regimen for his right eye and the topical steroid was tailed off.

His post-operative refraction was OD +1.00DS -0.50DC X 180^0 6/7.5, N_5 . OS Plano 6/5, N_5.

About 5 weeks after cataract surgery, we noticed posterior capsular opacification and VA reduction of 6/48. He was scheduled for NdYAG laser posterior capsulotomy and this improved his VA to 6/7.5. Again about 12 weeks post operatively, intermediate uveitis recurred with macular oedema and best corrected VA also became significantly reduced to 6/48 in the affected eye. He then received a repeat posterior sub tenons depo prednisolone and his best corrected visual acuity (BCVA) improved again to 6/7.5 and has remained stable and IOPs have remained within normal range. His left eye has remained normal and devoid of features suggestive of ocular sarcoidosis. He has since been weaned off the systemic steroid and is in stable general health.

Discussion

Sarcoidosis rarely presents in childhood and its aetiology is largely unclear [1, 4, 9]. A multisystem immune disorder which frequently manifests with granulomas, it has been reported as more common among African American patients and in the 13 years-15 years age bracket [1, 3]. Its incidence may be increasing in Nigeria [3]. Arguably, there appears to be an increasing incidence of several ocular conditions previously thought to be rare in Nigeria [16, 17]. This may be due in part to a change in eye health seeking behaviour, an uptake of the referral system [18] or as a result of an improved diagnostic service.

Generally, presenting symptoms of sarcoidosis are dependent on the structures involved in the disease process [1, 2]. It is found with eye features as initial presentation uncommonly (about 2% to 3% of cases) [1, 2, 8, 19]. Sarcoidosis-associated uveitis is reportedly the most common ocular manifestation and uveitic cataracts have been similarly associated [1, 6, 20]. Dyspnoea typically presents later in the disease, and patients are more fatigued when they have pulmonary and extrapulmonary disease [2, 21]. Fatigue (a manifestation in the index patient) is seen in up to 70% of patients with pulmonary sarcoidosis [1-5].

By way of standard protocol, according to guidelines set by the World Association of Sarcoidosis and other Granulomatous Diseases, the diagnosis of sarcoidosis (in adults and children >5 years of age) is dependent on 3 basic criteria: typical clinical presentation supported with radiologic evidence, histopathologic evidence from biopsies (classically noncaseating granulomas) and importantly, the exclusion of deferential diagnosis. While a definitive diagnosis requires a biopsy of the lesion, comprehensive, meticulous slit-lamp examination and a high level of clinical suspicion are invaluable for making a probable diagnosis of ocular sarcoidosis [1, 4, 21].

The Revised International Workshop on Ocular Sarcoidosis (IWOS) Criteria includes; ‘seven intraocular clinical signs suggestive of ocular sarcoidosis and eight systemic investigation results in suspected ocular sarcoidosis’ [1 9]. Based on these criteria, ocular sarcoidosis can be diagnosed as follows: [1, 19]

Definite ocular sarcoidosis where there is a positive biopsy along with a compatible uveitis.

Presumed ocular sarcoidosis without a biopsy if patient has bilateral hilar lymphadenopathy and a congruent uveitis [2].

Probable ocular sarcoidosis where patient has neither supportive biopsy nor hilar adenopathy, but has at least two supportive investigational tests and a minimum of three suggestive intraocular signs.

Possible ocular sarcoidosis if there are a minimum of four suggestive intraocular signs and at least two supportive investigational tests.

Our diagnosis of sarcoidosis was entertained upon the eye findings supporting a granulomatous panuveitis and blinding uveitic cataract; uveitis reportedly a common manifestation of ocular sarcoidosis [6, 8, 12]. This was additionally supported by the history of lung disease and supporting radiologic evidence. In addition, our patient had positive findings from his skin biopsy, and hypercalcaemia. Panuveitis has also been attributed to delayed presentation [6]. Our patient had a unilateral presentation and this was the less common pattern 10(33.3%) from Nepal [6]. However, their report was in a predominantly adult population (age range 9-62 years). In agreement with our finding, their study also reports radiologically demonstrable mediastinal hilar lymphadenopathy that is similarly reported from other geographic locations [2, 6].

Our index patient was male and had become symptomatic with lungs disease about a year prior to presentation in the eye department. This is in a agreement with other reports [1, 6, 7], however, there was a female preponderance in some reports [3, 5] making sex predilection less clear. Eye involvement necessitated the commencement of systemic and notable clinical improvements occurred after commencement of steroid therapy including improved VA, the resolution of chest findings and weight gain is similarly reported from other centres [2, 4, 6, 14, 22]. However, in consonance to what has previously been reported in Nigeria, he did not have eyelid nodules [22]. Hyporexia, hepatomegaly, nephrolithiasis, palor or vomiting prior to presentation as was seen in Mexico, were not part of his clinical features [4]. His abdominal ultrasound was essentially unremarkable.

No patient from an earlier report from an adult population in Ibadan Nigeria had ocular sarcoidosis and dry eye was the only ophthalmic manifestation in Lagos, Nigeria [3, 5]. However, all patients in their report initially presented in the chest and dermatology clinics [3, 5]. At the time of this report, our patients did not routinely undergo an ophthalmic referral where sarcoidosis was suspected as has been reported in other settings [14]. Importantly, our patient had significant chest features. This has similarly been reported elsewhere in Nigeria [3, 5, 22]. Where there is a high index of suspicion, the referral pathway from Chest/Respiratory Clinics to Ophthalmology Clinics and a multidisciplinary approach is therefore advocated [22]. Timely and appropriate referrals may facilitate the restoration of vision and thereby reduce the burden of avoidable vision loss in the paediatric age group.

Conclusion

Sarcoidosis can manifest in a variety of ways and may be an under-reported disease in Nigeria. In the paediatric age-group can lead to debilitating multi system disease and death. Blinding ophthalmic manifestations are found in this disease. A high index of suspicion and the application of standardized protocol including a comprehensive eye examination for diagnosis and treatment may treat visual disability, systemic manifestations, halt its progression and improve the quality of life. Corticosteroids are a useful first line treatment option.

Ethical Considerations: For the purpose of this study, the authors did not perform any experiments on human or animal subjects.

Confidentiality: The authors declare that patient's data did not appear in this report.

Author Contributions:

The authors confirm contribution to the paper as follows: EDN, GEB, AUE, EDN[2] and UDN: conceived the manuscript, EDN, GEB, AUE, EDN[2] CTA, UDN and DGN did the literature search and interpretation of clinical data. EDN, GEB, AUE, CTA, DGN: patient treatment. All authors contributed to the privately-sourced funding. EDN, GEB, AUE, EDN[2], CTA, DGN, UDN: Writing original draft: All authors were involved in writing, review & editing. All authors contributed to the article and approved

the submitted version. All authors read and approved the final version of the manuscript.

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