

PROPTOSIS AND BLINDNESS: AN UNUSUAL PRIMARY PRESENTATION OF CHRONIC GRANULOCYTIC LEUKAEMIA

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ABSTRACT

Proptosis and blindness are uncommon presentations of chronic granulocytic leukaemia. We present a 20-year old man with four months history of bilateral swelling of the eyes and deteriorating vision, abdominal swelling and early satiety. Examination revealed massive splenomegaly and bilateral blindness resulting from retinal detachment most probably due to retro-orbital accumulation of malignant myeloid cells. Peripheral blood film confirmed chronic granulocytic leukaemia, CGL. Clinical response to hydroxyurea was excellent but retinal detachment and blindness persisted. We suggest that CGL should be excluded in all cases of painless eye-swelling.

KEY WORDS: Proptosis, Blindness, Chronic granulocytic, Leukaemia.

Pak J Med Sci July - September 2006 Vol. 22 No. 3 326-328

INTRODUCTION

Symptomatic presentations of chronic granulocytic leukaemia, (CGL), are commonly related to anaemia, splenomegaly and increased cells turnover. These symptoms may include left quadrant abdominal swelling/pain, early satiety, night sweat, and weight loss.¹ Very high cell count could result in hyperviscosity which may be associated with stroke, priapism, stupor or visual changes due to retinal haemorrhage.²

Proptosis and blindness or any other ocular presentation is not a common primary presentation of chronic granulocytic leukaemia. When present, however, they are likely to be due to schloroma.³This is a mass of white cells deposited in any tissue of the body. Also called granulocytic sarcoma, it usually consists of

myeloid cells.³The first report of orbital granulocytic leukaemia was published in 1811 by Allen Buns who noticed the greenish tumour in a post mortem specimen.⁴ This is the origin of the term schloroma meaning many colours. In order to make this diagnosis, other causes of proptosis such as haematoma, retinoblastoma and rare malignant causes such as Non-Hodgkin's lymphoma (NHL), and recently a complication of hydroxyurea therapy must be excluded.^{3,5,6}

Proptosis and blindness as the primary presentations of CGL is unknown in our experience and seems uncommon as we could not find a single report in any medical literature, hence this report.

CASE REPORT

A 20 year old young man was referred to us from a religious mission hospital (about a 100km away from our centre), on account of elevated white blood cells count. Four months prior to presenting to our clinic, he had noticed a swelling of the right eye which was shortly followed by a similar swelling in the left eye. These swellings were gradually increasing in size and the vision of both eyes was becoming impaired. About two months following the onset of the bilateral eyes swelling he was seen at a mission hospital

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* Received for Publication: July 5, 2005

* Accepted: December 26, 2005

where the referring Ophthalmologist found retina detachment and blindness of the right eye and impaired visual acuity, (12/6) of the left eye. The attending ophthalmologist initially thought of retinoblastoma. But a routine full blood count, (FBC) however, revealed a leucocytes count of 180,000/ml. He was subsequently referred us on account of the very high leucocytes count.

Further history revealed that he did not present to us until six weeks following the initial referral on financial ground. He was guided into the consulting room by the mother and complained of abdominal discomfort, and early satiety. There was no fever or bleeding disorder. He was unable to see his environment, and had been out of school (First year Higher Diploma student of a Polytechnic in Nigeria) for 4 months. There was no significant medical or surgical history. He is first of four children by his mother, (who is a third wife) and the 7th child of his father. Mother is a petty trader and father is a fisherman. There was no similar history in his family.

His hearing was grossly intact. He could only move around when guided. Physical examination revealed a moderately well fed young man in obvious distress but unable to see his environment. His hearing was grossly intact, and with mental clarity. He was not pale, not jaundiced, and had no peripheral lymph node enlargements. Pulse was 96/minute, moderate volume and B.P was 100/70mmHg. Heart sounds were I & II and had no murmurs. There were no chest signs. The abdomen was full but not tender. The spleen was 20cm below the left costal margin, smooth, firm and not tender. The liver was not palpable and kidneys were not ballotable. No abnormality was found in the musculoskeletal system.

Ocular Examination: It revealed bilateral swelling and ptosis of both eyes, (right > the left); the right measuring about 8cm in diameter while the left measured 6cm in diameter. Both sclerae were completely obscured. Fundoscopy showed bilateral retina detachment.

Investigations carried out revealed the following:

Full Blood Count: Haemoglobin was 13.2 g/dl. Red cell indices were normal. Total white blood

count, (TWBC) was 380,000 × 10⁹/L. Platelets count was 390,000 × 10⁹/L. Blood film showed granulocytes at various stages of maturation and the differential WBC was; myeloblasts 6%, promyelocytes 8%, myelocytes 29%, band forms and matured neutrophils 39%, eosinophils 5%, basophils 9%, and monocytes 2%. Lymphocytes 2%. Bone marrow cytology was essentially similar to that of the peripheral blood film.

Electrolytes, urea and serum creatinine were normal, (K⁺ 4.6mmol/L, Na⁺ 136 mmol/L, Cl⁻ 110mmol/L, Urea 6.67mmol/L, Creatinine 79.74umol). Uric acid was 0.8mmol/dl. Ultrasonography of both eyes revealed extra-orbital deposits of opaque masses.

Based on the above a diagnosis of Chronic Granulocytic Leukaemia in chronic phase with proptosis and blindness was made.

TREATMENT

He was immediately admitted and the nature of his disease explained to him and his mother. The available modalities of treatment including bone marrow transplantation, and the expectations were explained. He was put on capsules hydroxyurea 3G daily, tablets allupurinol 200mg tds with liberal fluid intake. The TWBC was reduced to 20,000/ml in 10 days and spleen also reduced to 10cm. Both eyes swelling were regressing remarkably. By the third week of admission, the WBC had returned to normal (5,100/ml) spleen was no longer palpable and the eye swellings had also completely returned to normal.

Repeat fundoscopy still showed retina detachment and there was no improvement in his vision. Repeat ultra-sound did not show any



Figure-1: Restricted face photograph of the patient before chemotherapy

retro-orbital masses. We therefore reduced his HU to 1g daily as well as allopurinol 100mg B.D. per oral. Repeat FBC; Uric; Electrolytes, Urea and creatinine were all normal. He was discharged one month after admission and given 2 weeks appointment at the Haematology outpatient clinic. The nature of his disease was again emphasised and the need to keep regular appointments stressed. In spite of these admonitions he has since defaulted even without keeping the first post-discharge clinic appointment. All attempts to trace him in the forwarding address in Delta state (swampy and very difficult terrain) have proved futile.

DISCUSSION

Orbital presentation of chronic myeloid leukaemia as the first clinical symptom/sign is uncommon in our experience. A few reports have, however, established such manifestation of chronic myeloid leukaemia which may sometimes precede bone marrow or peripheral blood manifestations.^{3,7,8}

Granulocytic sarcoma could affect any part of the body including the orbit.^{3,7} Orbital manifestation is said to be common in children and young adults as in the case of our patient. None of the above reports mentioned blindness as the first clinical manifestation as seen in our patient.

Late presentation is a recurrent decimal in resource poor communities. This is the norm in our clinical practice. The index patient presented late. It is possible that the initial delay in presentation led to the complete blindness of this patient. It is also possible that early ocular involvement might be associated with central nervous system complications such as cerebro-vascular accident. We cannot be certain of this since patient was lost to follow-up.

Leucocyte alkaline phosphatase (LAP) score was not performed because it is not being done in our centre now. However, the long history with the clinical findings may have sufficiently excluded leukaemoid reaction. Again cytogenetic study is not being done at our centre. We are therefore unable to make any comment on the influence of Philadelphia chromosome on the ocular manifestations of chronic granulocytic leukaemia.

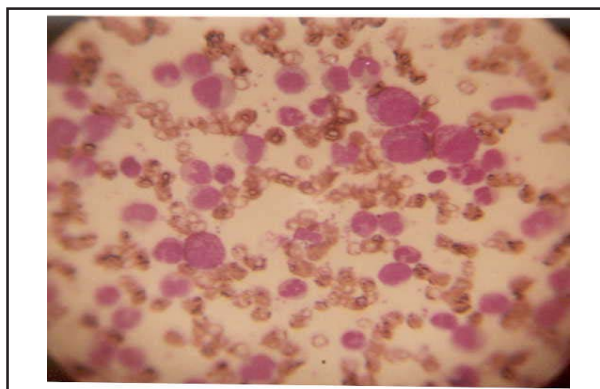


Figure-2: Photomicrograph of the peripheral blood film of the patient before chemotherapy.

We suggest that CGL should be excluded in patients with painless proptosis and blindness. Using the clinical features and basic laboratory investigations like white cells count and blood film the diagnosis of CGL can be made speedily.

ACKNOWLEDGEMENT

We wish to thank Dr. (Mrs) O.T. Edema and her team of the Department of Ophthalmology, University of Benin Teaching Hospital, Benin City, for the ophthalmic evaluation of this patient.

REFERENCES

1. Kantarjian HM, Deisseroth AB, Kurzroch R. Chronic granulocytic leukaemia: a concise update. *Blood* 1993; 82: 691-703.
2. Savage G, Szydlo RM, Goldman JM. Clinical features at diagnosis in 430 patients with chronic myeloid leukaemia seen at a referral centre over a 16-year period. *Br J Haematol* 1997; 96: 111-6.
3. Stockl FA, Dolmetsch AM, Saornil MA. Orbital granulocytic sarcoma. *B J Ophthalmol* 1997; 81: 1084-8.
4. Burns A. Observation on the surgical anatomy of the head and neck. 4th edition. Baltimore, USA: Granville Sharp Pattison, 1823.
5. Verbraeken HE, Hassens M, Hildegaard, Briem. Ocular non-Hodgkin's lymphoma: a clinical study of cases. *B J Ophthalmol* 1997; 81: 31-6.
6. Puri B, Woocock, O'Donnell MP. Localized chronic eyelid disease resulting from long term hydrourea therapy: *B J Ophthalmol* 2001: 85:371.
7. Neimen RS, Barcos M, Bernard C, Bonnen H. Granulocytic Sarcoma: Clinicopathologic study on 62 biopsies. *Cancer*, 1981; 48: 1426.
8. Ziemmerman LE, Font RL. Ophthalmic manifestations of granulocytic sarcoma, (myeloid sarcoma). *Am J Ophthalmol* 1975; 80: 975-90.