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CASE REPORT

Acute sinusitis and blindness as the first presentation of chronic lymphocytic leukaemia

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ABSTRACT

Chronic lymphocytic leukaemia (CLL) is the most frequent form of leukaemia among adults in the Western world, presenting at a median age of 65 years. The diagnosis is usually made incidentally during routine blood examination while the disease is still in its early phase. We report a case of blindness of 24 hours due to acute sinusitis based on CLL localisation in a patient with undiagnosed CLL. Emergency endoscopic sinus surgery and intra- and extra-ocular orbital decompression were performed. The sinusitis resolved after surgery and intravenous antibiotics. Her vision improved within 24 hours and eventually recovered completely after six months. Her CLL remained in an indolent state, needing no active treatment. This case illustrates that blindness from a lymphoproliferative disorder may be treated with emergency endoscopic sinus surgery instead of conventional chemotherapy in order to salvage the vision first, even if the vision is lost for more than 24 hours.

KEYWORDS

Chronic lymphocytic leukaemia, blindness, sinusitis

What was known on this topic?
Chronic lymphocytic leukaemia (CLL) is the most common form of leukaemia in adults. Ocular symptoms are uncommon in CLL. CLL is treated with immunochemotherapy. Blindness from orbital cellulitis usually originates from bacterial sinusitis. Urgent surgical decompression within 24 hours is needed to salvage the vision.

What does this case add?
Patients with undiagnosed early CLL can present with acute blindness due to orbital infiltration and orbital CLL involvement. This has not been reported in the literature. Visual loss in early CLL can benefit from surgical decompression instead of conventional chemoradiotherapy in order to achieve quick decompression of the orbit and hence salvage vision, even when blindness is present for more than 24 hours.

INTRODUCTION

Chronic lymphocytic leukaemia (CLL) is the most common form of leukaemia in adults, in Western countries, representing about 25-30% of all leukaemias.[1] Most patients are diagnosed incidentally during a routine blood investigation when they are still in the early phase of CLL.[2] Frequent presenting symptoms include lymphadenopathy, hepatosplenomegaly and/or cytopenias, and some cases present with unintentional weight loss fever, night sweats, or extreme fatigue.[3]

Ocular symptoms are uncommon and, to our knowledge, especially acute blindness due to fulminant orbital cellulitis has not previously been reported as the presenting symptom in CLL. Moreover, it is unusual to recover vision if there is a delay in intervention. Our patient's vision completely recovered after endoscopic sinus surgery and surgical decompression of the orbital content.

CASE REPORT

A 59-year-old female with left periorbital swelling and blindness for 24 hours was referred to us. She had a three-week history of flu-like symptoms with cervical lymphadenopathy, and four days of left periorbital pain with progressive periorbital swelling. The vision in her left eye rapidly deteriorated to complete blindness during the evening before she sought help. She had bilateral rhinorrhoea, hyposmia and nasal congestion. She had no history of diabetes or any other immunocompromising disease. She was febrile but had no signs and symptoms of sepsis. She had severe left periorbital oedema, proptosis with absent light and red reflex, and no extraocular eye movement. Nasendoscopy showed bilateral purulent discharges and inflamed nasal mucosa. Computed tomography (CT) showed left-sided pansinusitis, left proptosis and diffuse infiltrates within the orbit but no obvious orbital abscess (*figure 1*). She was started on intravenous amoxicillin/clavulanate.

Her laboratory results were as follows: leucocyte count 61.2 x 10^9/l (neutrophils 44%, lymphocytes 48% and monocytes 8%), haemoglobin 12.0 g/l, thrombocyte count 365 x 10^9/l, C-reactive protein 276 mg/l, lactate dehydrogenase 169 U/l, serum glucose 7.5 mmol/l, and the plasma levels of immunoglobulins were normal (IgA 1.75 g/l, IgG 6.9 g/l, IgM 0.47 g/l).

Our primary working diagnosis was a left sinogenic, likely bacterial, orbital cellulitis with blindness. Leukaemia was suspected in view of her leucocytosis. The chance of her recovering her vision was low since it had been more than 24 hours. Nevertheless, surgical drainage to decompress the orbit was the only option to save the patient's vision. Hence, after informed consent, an emergency external lateral orbital decompression and complete fronto-maxillo-ethmoidectomy with medial orbital decompression was performed. Purulent fluid was drained from the sinuses. Unexpectedly, no pus was found during the orbital decompression. No bacteria were cultured. Tissue biopsies from the paranasal sinus and lamina papyracea showed dense infiltrates dominated by a monotonous population of small lymphocytic cells that expressed CD20, CD23 and CD5 (*figure 2*). Molecular analyses showed immunoglobulin heavy chain gene rearrangement. Immunophenotyping of peripheral blood showed expansion of CD5 and CD23 positive B cells. Total number of circulating leukaemia cells (CD5/CD19/CD23 triple positive) was 29.4 cells/ul. Immunoglobulin levels were normal. A diagnosis of CLL was made, clinically staged Rai 0 or Binet A. She improved clinically after surgical decompression. Hence, anti-leukaemic therapy was not started.[3,4]

Figure 1. *Contrast-enhanced CT scan of orbit/sinuses illustrating left ethmoiditis and left proptosis secondary to intra-orbital infiltrates*

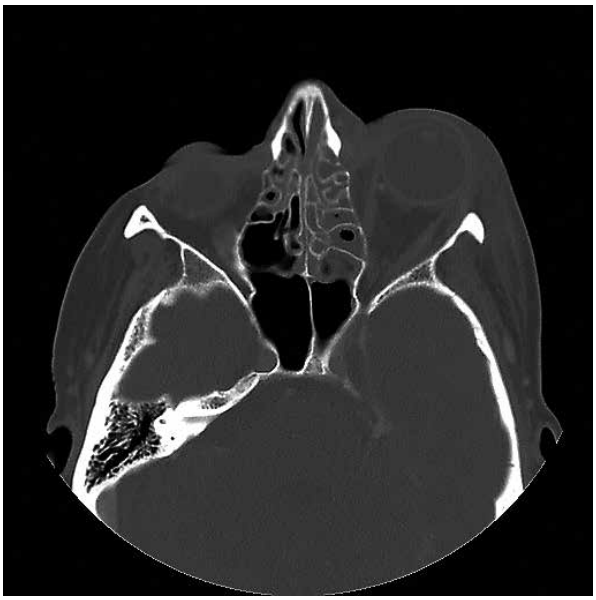
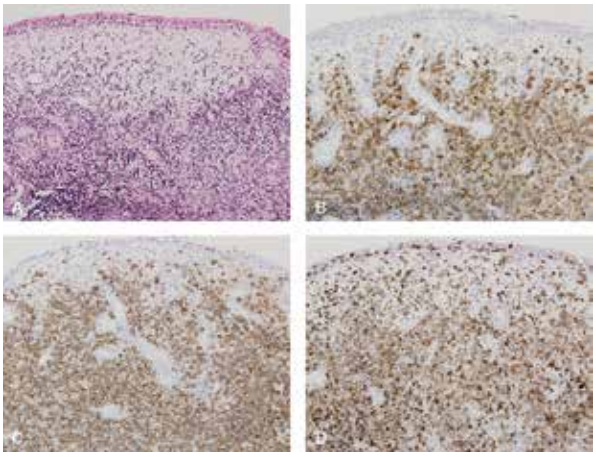


Figure 2. *Mucosal biopsy from the paranasal sinus infiltrated by CLL cells, stained with (A) haematoxylin&eosin and with antibodies specific for (B) CD20, (C) CD23 and (D) CD5. The CLL cells demonstrate moderate expression of CD5 as opposed to the strongly CD5-expressing T cells (100x magnification)*



After surgery, the patient's perception to light improved within 24 hours. Her vision was better two weeks later, and had completely recovered within six months. She had residual diplopia for three months. Besides the first episode, her CLL remains asymptomatic with stable peripheral blood counts and no other signs of disease progression. Therefore, she is currently being monitored without therapy.

DISCUSSION

The clinical presentation of our patient was caused by the infiltration of sinus mucosa and orbital content by malignant B cells displaying a CLL phenotype. This resulted in sinusitis, orbital cellulitis and subsequently blindness.

Sinusitis in early CLL is not common. Sinusitis in advanced stages of CLL is usually due to increased susceptibility of bacterial infections due to acquired hypogammaglobulinaemia or (following T cell depleting treatment) due to an opportunistic infection.[5] The spread of infection from the sinus to the eye is through the lamina papyracea or by venous spread. Complications of sinusitis, if not treated promptly, can lead to visual loss, meningitis, septicaemia and death.[6] Visual loss can occur because of central retinal artery occlusion, optic neuritis, corneal ulceration, pan-ophthalmitis and orbital apex syndrome.[7]

Unlike most sinusitis, our patient's sinusitis and blindness was due to infiltration of CLL in both the sinus mucosa and orbital content. We hypothesised that mucosal infiltration by clonal lymphocytes obstructed the sinus drainage pathway and augmented local inflammation and promoted its spread.

Progressive loss of visual acuity is very rare as an early clinical manifestation of CLL.[8] There were case reports of progressive visual loss in patients who were either in remission or with known CLL in early stage who had not started treatment.[9,10] Gonsalves et al. reported a case of progressive visual loss that was caused by optic neuritis in a previously untreated patient with early stage CLL, in whom a complete resolution of visual defects was achieved with chemoimmunotherapy using fludarabine and rituximab.[10]

Sinogenic orbital cellulitis requires aggressive treatment with intravenous antibiotics, early imaging to exclude abscess and close monitoring.[11] Surgery is required if symptoms do not improve after 24 hours of appropriate antibiotic treatment, or sooner in case of visual loss or orbital abscess. In our patient, contrast-enhanced CT showed diffuse infiltrates in the left orbit without signs of subperiosteal or orbital abscess. Indeed, during surgery no abscesses were found. However, always keep in mind that CT and even MRI have a false-negative rate of 10-15% for subperiosteal abscess.[12,13] Therefore, a reduced visual acuity alone is a good indication for imminent endoscopic sinus surgery and medial orbital decompression.[11,13] External orbital exploration and drainage is required in case of lateral orbital abscesses. Longer duration of blindness predicts poorer visual outcome.[7] Magnetic resonance imaging delineates soft tissue infiltrates better, but there was no time for that since her left vision was lost for more than 24 hours. All fluid and tissue sampled showed features of CLL.

In summary, undiagnosed CLL can present with acute blindness from orbital infiltration which may be best treated with surgical decompression instead of immunochemotherapy in order to save the vision.

DISCLOSURES

The authors declare no conflicts of interest. No funding or financial support was received.

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