Unilateral infiltration of the optic nerve revealing relapse of an acute lymphoblastic leukemia

Infiltration unilatérale du nerf optique révélant une rechute d'une leucémie aiguë lymphoblastique

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RÉSUMÉ

Introduction: L'atteinte oculaire au cours des leucémies peut intéresser l'orbite, l'uvée, la rétine et le nerf optique. Elle résulte soit d'une infiltration oculaire directe par les cellules tumorale soit d'une atteinte indirecte en rapport avec les troubles hématologiques, les infections opportunistes ou les effets indésirables des traitements. Nous rapportons un cas d'une infiltration unilatérale du nerf optique révélant une rechute d'une leucémie aiguë lymphoblastique.

Observation: Il s'agit d'une femme âgée de 48 ans, en rémission d'une leucémie aiguë lymphoblastique B, qui a consulté pour céphalées et flou visuel au niveau de l'œil gauche. L'examen ophtalmologique a retrouvé à l'œil gauche, une acuité visuelle à 20/200, un volumineux œdème papillaire associé à une masse entourée d'hémorragies rétiniennes, d'exsudats et d'un important décollement séreux rétinien. Le Scanner orbito-cérébral a objectivé un épaississement du nerf optique gauche et a exclu un oedème papillaire par hypertension intracrânienne secondaire à une atteinte du système nerveux central. Le myélogramme et la ponction lombaire ont conclu à une infiltration blastique confirmant la rechute de la leucémie aiguë.

Conclusion: L'incidence de l'atteinte oculaire au cours des leucémies aiguës lymphoblastiques a diminué depuis l'introduction d'un traitement prophylactique systématique du système nerveux central. Des contrôles ophtalmologiques réguliers sont nécessaires afin de garantir un diagnostic et un traitement précoces.

Mots-clés

leucémie aiguë lymphoblastique, atteinte oculaire, infiltration du nerf optique

SUMMARY

Introduction: Ocular infiltration of leukemia can involve orbit, uveal tract, retina and optic nerve. It may result from direct ocular infiltration by leukemic cells or indirect ocular involvement resulting from secondary hematologic changes, opportunistic infections and complications of various modalities of therapy. We report a case of unilateral infiltration of the optic nerve revealing a relapse of acute lymphoblastic leukemia.

Case report: forty eight years-old woman in a remission of acute B lymphoblastic leukemia presented with headaches and blurred vision in the left eye. Ophthalmic examination showed a visual acuity reduced to 20/200 in the left eye, and a voluminous disc edema with papillary mass surrounded by retinal hemorrhages, exudates and important serous retinal detachment. CT scan showed a thickened left optic nerve and excluded true papillary edema due to intracranial hypertension secondary to central nervous system involvement. Myelogram and lumbar punction demonstrated blast infiltration and confirmed ocular relapse of the leukemia.

Conclusion: The incidence of ocular involvement lymphoblastic acute leukemias decreased since the introduction of a systematic prophylactic treatment of central nervous system. Periodic ophthalmic examination is necessary to allow early diagnosis and treatment.

Key-words

Acute lymphoblastic leukemia, ocular involvement, optic nerve infiltration

INTRODUCTION

Ocular infiltration of leukemia may occur in 9 to 90% of cases, and can involve orbit, uveal tract, retina and optic nerve (1, 2). It may result from direct ocular infiltration by leukemic cells, indirect ocular involvement resulting from secondary hematologic changes, opportunistic infections and complications of various modalities of therapy (2). However, since the introduction of a systematic prophylactic treatment of central nervous system involvement, their incidence has decreased.

It corresponds mainly to infiltration of the retina and choroid by neoplasic cells. Blast infiltration of the optic nerve head is less common and it is considered as a central nervous system involvement (1,2).

We report a case of unilateral infiltration of the optic nerve revealing a relapse of acute lymphoblastic leukemia.

CASE PRESENTATION

We report a case of forty eight years-old woman with a history of acute B lymphoblastic leukemia associated to a mutation of Philadelphia chromosome that was treated in hematology with chemotherapy. She was considered in hematological remission since 8 months and proposed to undergo allo-transplantation of bone marrow. She was referred to ophthalmologic emergency for acute headaches associated with blurred vision in the left eye. On examination, visual acuity was reduced to 20/200 in the left eye, anterior segment and vitreous were calm, intraocular pressure was normal, and fundus examination revealed voluminous disc edema with papillary mass surrounded by retinal hemorrhages, exudates and important serous retinal detachment (Figure 1). Ophthalmic examination of the right eye was unremarkable.

Fluorescein angiography (Topcon TRC.50IX) showed high hyperfluorescence of the optic nerve head with vascular leakage and peripapillary serous retinal detachment (Figure 2). B ocular ultrasound confirmed serous retinal detachments surrounding the optic disc and optic nerve head thickening (Figure 3). Optical coherence tomography was not performed because of tiredness of the patient. The computed tomography scan excluded papillary edema due to intracranial hypertension secondary to central nervous system involvement. Lumbar punction demonstrated blast infiltration and confirmed ocular relapse of the leukemia.

Infectious cause particularly viral infection (herpes simplex virus and cytomegalovirus) was investigated because of the immunosupression related to chemotherapy and was negative.

The patient returned to hematology for reinforcement of the chemotherapy and central nervous system radiation therapy. Two months later, she underwent allo-transplantation of the bone marrow but she did not demonstrate improvement in her visual acuity.

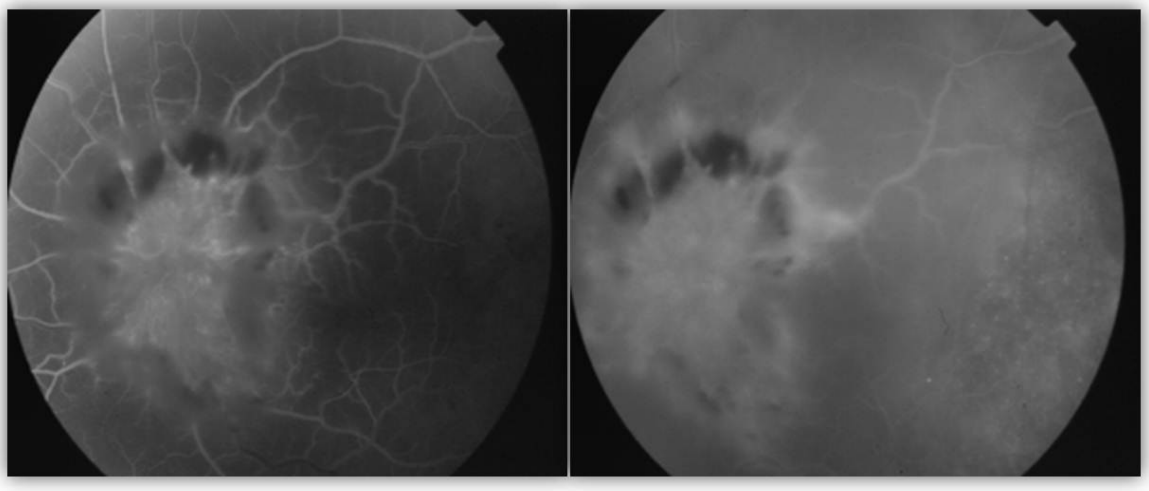

Figure 1: Fundus photography of the left eye showing voluminous disc edema with papillary mass associated with retinal haemorrhages and surrounded serous retinal detachment.

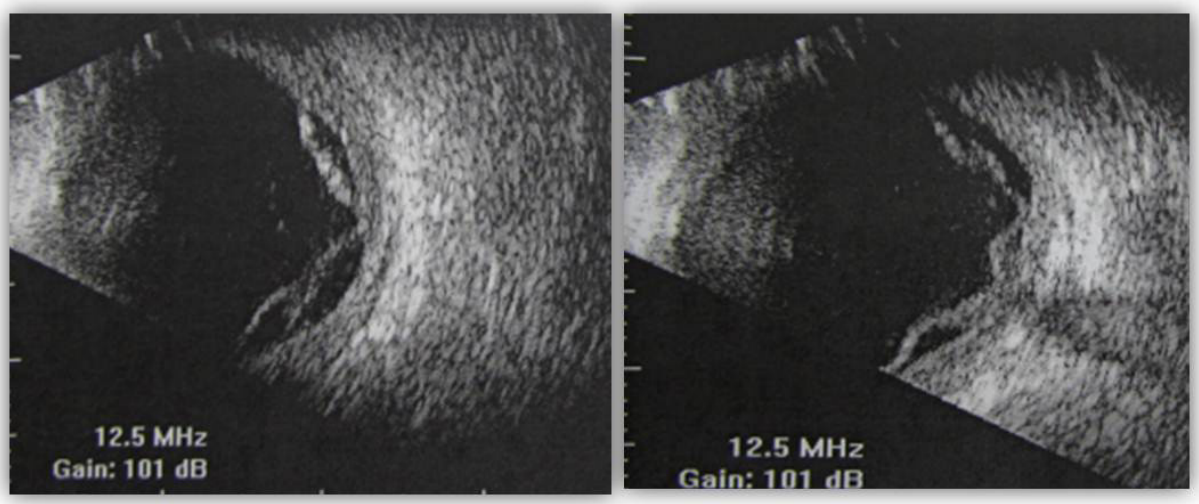

Figure 2: Fluorescein angiography (Topcon TRC.50IX) showing high hyperfluorescence of the optic nerve head with vascular leakage.

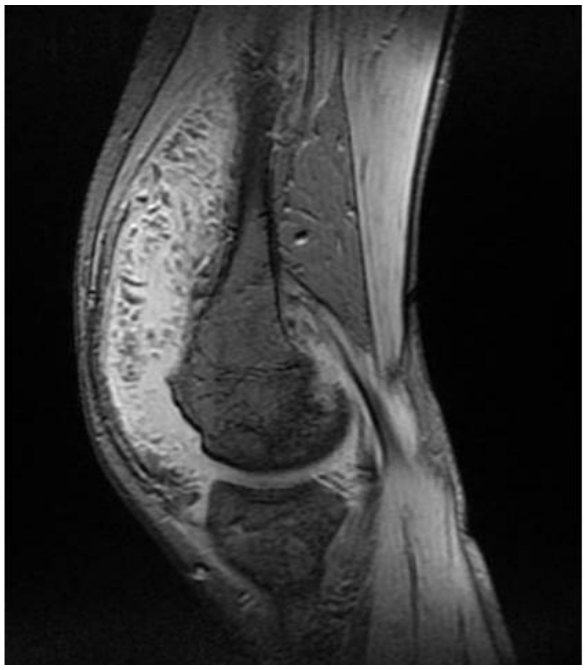

Figure 3: B ultrasound of the left eye showing multifocal serous retinal detachments surrounding disc area associated with optic nerve head thickening.

DISCUSSION

Our case presented leukemic involvement of both the optic nerve head and choroid. leukaemic infiltration of the optic nerve is relatively rare and is considered to be one of the significant clinical findings of central nervous system leukaemia (3). In contrast, the choroidal infiltration with leukaemic cells is common, especially in the case of acute lymphocytic leukaemias, we found diffuse choroidal thickeness on B ultrasound in our patient.

Leukemic ophthalmopathy is mainly reported in children who are known to have the higher prevalence of acute leukemia (3-6). Eze et al reported in a prospective study of 72 adults that ophthalmic leukemic involvement is more common in males, chronic leukemia, and tends to affect

more frequently ocular posterior segment structures than anterior segment (7).

The most common site of infective or neoplastic leukemic ocular infiltration is the choroid as demonstrated by postmortem studies based on histological examination of the choroid. Diffuse choroidal infiltration manifests clinically by serous retinal detachment and leopard spot pattern of the retinal pigment epithelium (8) as presented by our patient.

Optic nerve infiltration is reported in 13 to 18% in necropsic studies (9). This relapse's site may be related to a probable barrier as suggested by Nikaido et al. which interrupts the smooth flow of cerebrospinal fluid making optic nerve unreached by systemic chemotherapy (1) and requiring additional radiotherapy (5). It is however rarely reported as the initial isolated presentation revealing relapse of leukemia in adults with complete remission (10, 11) which may occur even in presence of normal complete blood count and bone marrow examination.

Lin et al. reported three cases of leukemia in adults with a leukemic infiltration of the optic nerve as the initial isolated presentation of disease relapse (10), Siedlińska et al. reported a case of a 60-year old woman with acute T-cell lymphoblastic leukemia who manifested infiltration of the retina and the optic nerve in hematological remission (11). Ocular manifestations of relapse in acute adults leukemia seem to involve more frequently the posterior segment structures compared to relapse in acute childhood leukemia which seems to involve anterior segment. In fact, Chocron et al. reported in three child, two cases of recurrent anterior uveitis and a case of a conjunctival tumor (12).

Ophthalmic involvement in leukemia is associated with vision loss and reduce survival prognosis to less than 24 months. Mandatory regular ophthalmic examination was necessary in patients with leukemia even in absence of ocular symptoms to detect subclinical ocular involvement or subclinical relapse during complete remission.

Our case report describes unilateral optic nerve infiltration in a woman which revealed relapse of leukemia and we insist as many previous authors on periodic ophthalmic examination to allow early diagnosis and treatment.

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