IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 22, Issue 4 Ser.7 (April. 2023), PP 59-63
www.iosrjournals.org

Unilateral optic neuropathy and Mycosis fungoide

H.Hamdani; L.Arab; S.Ennaki; G.Daghouj; B.Allali; L.Elmaaloum; A.Kettani
Pediatric ophtalmology unit – Hopital 20 Août 1953 – Casablanca

ABSTRACT
Introduction *— Mycosis fungoides is a non-Hodgkin lymphoma that arise from skin-tropic clonal T lymphocytes. The ophthalmologic complications of non Hodgkin lymphoma are rare. In most cases, it's an infiltration of the orbital fat that can lead to a compressive optic neuropathy. Infiltration of the optic nerve and their sheaths by lymphoma remains exceptional.*
Observation *— We report the case of a 38-year-old female patient treated for mycosis fungoide. She presented with a unilateral decrease in visual acuity in her right eye that was reduced to light perception. Her examination revealed a right afferent pupillary defect and an optic atrophy. Brain MRI emphasized an infiltration of both optic nerves with no other orbital or brain abnormality. Cerebrospinal fluid analysis showed lymphomatous meningitis. She was then considered to have lymphomatous optic neuropathy.*
Conclusion *- The diagnosis of lymphomatous optic neuropathy must be confirmed by brain MRI and cytopathological study of the CSF. Treatment options include high-dose intravenous chemotherapy, intrathecal chemotherapy, radiation therapy and high-dose corticosteroids.*
Keywords: *Optic neuropathy – Mycosis fungoides – Non-Hodgkin Lymphoma*
Conflicts of interest: None

Date of Submission: 07-04-2023 Date of Acceptance: 20-04-2023

I. INTRODUCTION :

Cutaneous T-cell lymphomas (CTCLs) encompass a heterogeneous collection of non-Hodgkin lymphomas that arise from skin-tropic memory T lymphocytes. Among them, mycosis fungoides (MF) and Sézary syndrome (SS) are the most common malignancies (1).

Ocular involvement is uncommon in LMNH. They are mainly intraorbital masses, conjunctivo-lacrymal infiltration or posterior uveitis (2). Optic neuropathies are even rarer complications. They can be the consequence of compression of the optic nerve by a tumor mass or infiltration of the nerve fibers by lymphoma cells (3). We report the case of a unilateral infiltration of the optic nerve in a female patient with mycosis fongoides.

II. OBSERVATION :

Mrs. F.A., 38 years old, was treated in dermatology for mycosis fungoides. She presented with erythematosquamous swollen patches of the face and scalp Figure 1. Histological examination of the skin showed mycosis fungoides in the process of transformation to CD 30-.

She presented to the emergency room with a decrease in visual acuity in her right eye that had been evolving for five days and started to complain of retro orbital pain when moving her eyes, initially interpreted as due to iatrogenic toxicity of vincristine. On ophthalmological examination, the visual acuity in the right eye was light perception LP, with a right pupillary afferent deficit. The anterior segment and the vitreous were normal, and the fundus showed stage III papilledema. The examination of the left eye showed a visual acuity of 6/10 with a normal photomotor reflex, the anterior segment and the fundus were normal. Intraocular pressure measured by applanation in both eyes was normal

Fluorescein angiography and papillary optical coherence tomography were performed, confirming the papillary edema on the right. Macular OCT showed macular edema in the right eye.. **Figure 2-3-4**

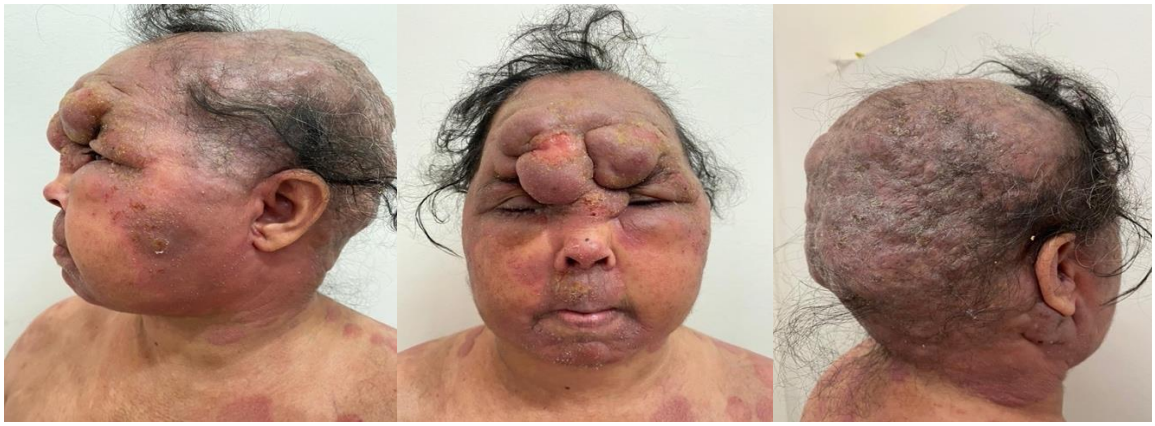

Figure 1 Photographs showing skin involvement with erythematous scaly swollen patches of the face and scalp.

Cranio-orbital CT scan showed enlargement of the 2 optic nerves without brain parenchyma abnormality (fig2).
Cytological analysis of the cerebrospinal fluid confirmed the invasion of the meninges by the lymphoma: hypoproteidinorachy at 0.15 g/L, hypoglycorachy at 0.22 g/L, 988 cells/mm3, of which 95% lymphomatous cells.
The patient received intravenous injections of 1 g of methylprednisolone per day for three days combined with systemic and intrathecal chemotherapy consisting of cyclophosphamide, vincristine, doxorubicin, and high-dose methotrexate.
Initially, the patient responded well to the treatment, the right visual acuity increased to 2/10 but the patient died 2 months later.

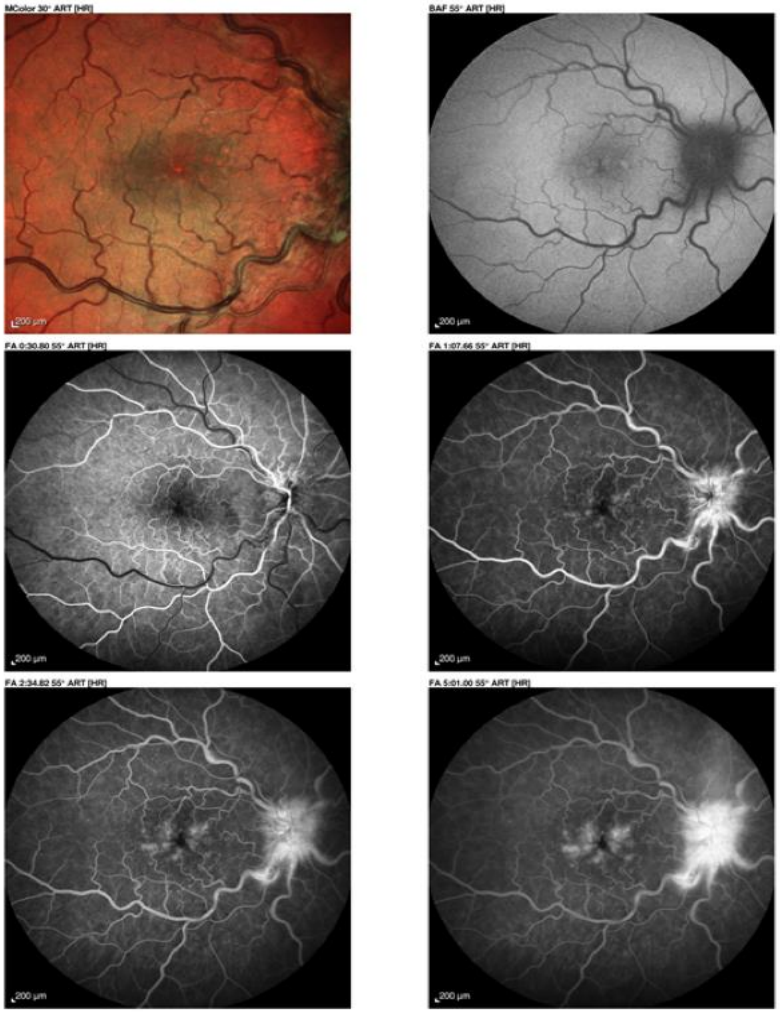

Figure 2 : **Angiographic images showing papillary oedema with papillary and macular fluorescein staining of the right eye**

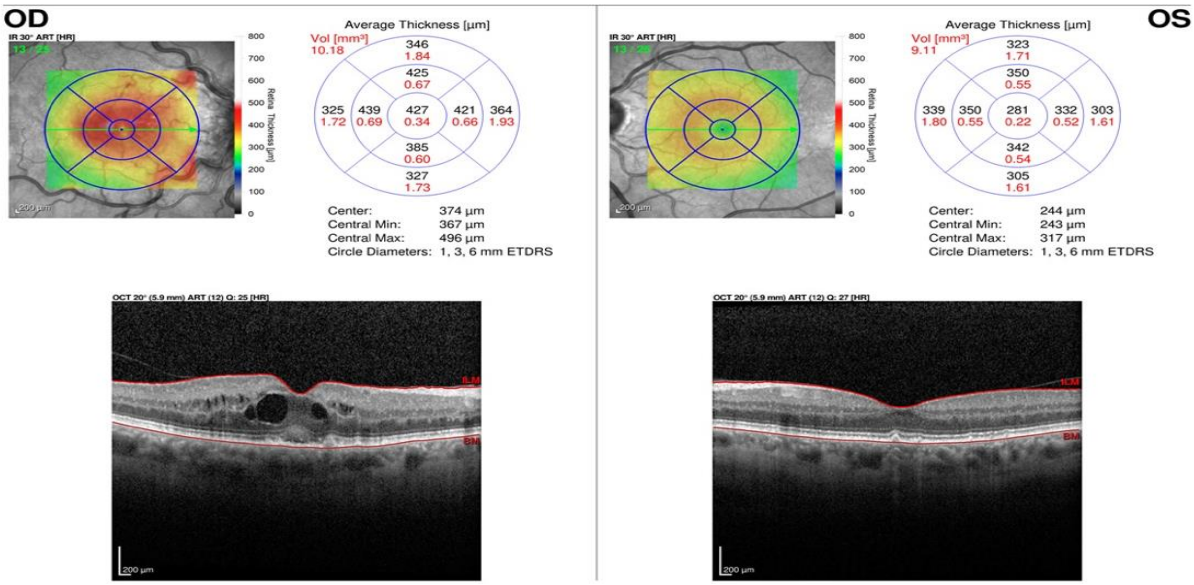

Figure 3 : **OD: Focal cystoid macular edema OS: Normal**

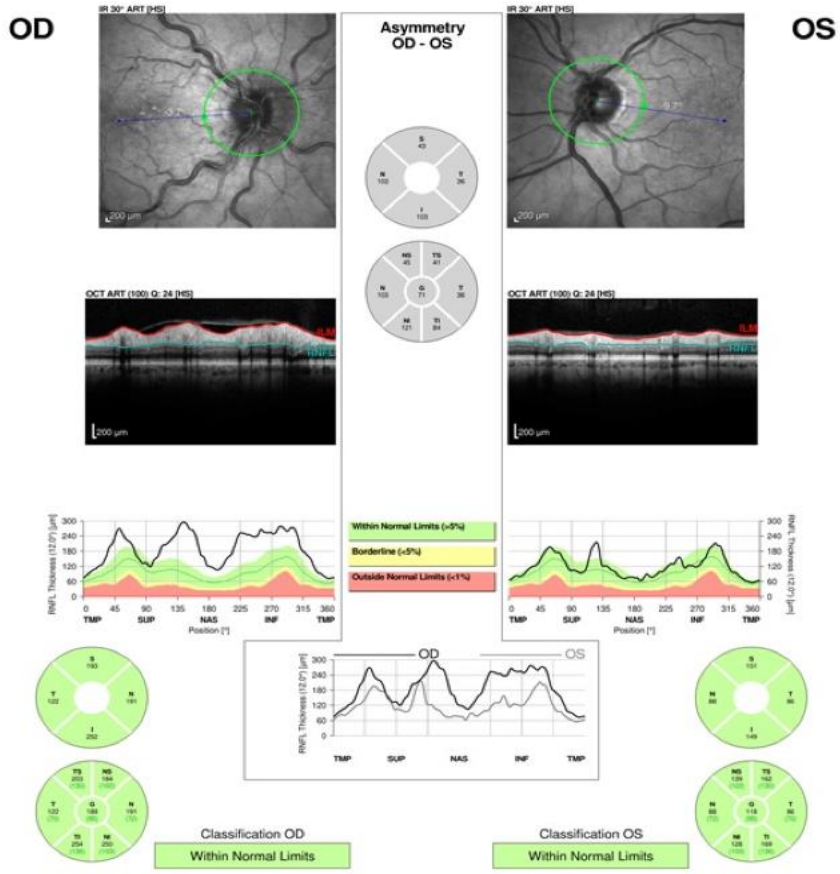

Figure 4 : Papillary OCT showing right papilledema

III. DISCUSSION :

Mycosis fungoides (MF) is a type of cutaneous T-cell lymphoma (CTCL) that primarily affects the skin, and represent 4% of the non-Hodgkin's lymphomas(NHL) (4).

However, ophthalmic involvement can also occur in rare cases and are highly variable. The most commonly involved ocular tissue is the eyelid but intraocular involvement can be found in the advanced stages of the disease.

The optic nerve is more frequently involved by secondary malignancy than by primary malignancy(5). There are three accepted routes of involvement: direct invasion by ocular melanoma or retinoblastoma; infiltration by lymphoma or leukemia; or blood-born metastasis from distant malignancies, generally carcinomas (6).

Metastatic disease to the eye and orbit most frequently involves the choroid, presumably because of its blood supply. In an Armed Forces Institute of Pathology clinicopathologic review, isolated optic nerve metastasis occured in ~1.3-12% of cases of all metastasis of the eye and orbit (6–8).

Lymphomatous optic nerve infiltration is a neuro-oncologic emergency, which often presents a clinical dilemma as other etiologies may be difficult to rule out. Autoimmune inflammation, infection and medication or radiation effects can manifest similar features at presentation, and may be clinically indistinguishable based on standard diagnostic techniques alone(9).

Case reports highlight compressive optic neuropathy by orbital lymphoma (10), paraneoplastic optic neuritis (11), and central retinal artery occlusion secondary to hyperviscosity (12). Optic neuropathy may also develop secondary to radiation-related necrosis, vincristine toxicity, or infection (8) leukemic or lymphomatous infiltration comprises an estimated 5% of secondary optic nerve tumors(6).

Secondary involvement of the central nervous system in systemic lymphoma is less common than primary central nervous system lymphoma(13), however, secondary disease can more often involve the meningeal, perivascular, and spinal epidural areas(8). Tumor cells infiltrate cranial and spinal nerve together

with their meninges (14), and the parenchyma of the optic nerve may be invaded as well (15,16). Optic nerve extension occurs most often in longstanding or recurrent systemic lymphoma, although anecdotal reports have appeared in the literature in which visual loss was the presenting sign of systemic disease(6). Most patients with optic nerve metastases have other known systemic metastases at the time of presentation with ocular involvement, mean survival time after discovery of metastasis to the optic nerve ranges between 6 and 9.3 months (17,18)

MRI findings in lymphomatous infiltration of the optic nerve include enlargement of the optic nerve and enhancement of the optic nerve sheath(8). In our case, the infiltration of the optic nerves on MRI, although bilateral, does not cause papilledema on the left side. It can be explained by the predominance of infiltration in the posterior orbital segment at distance from the globe.

Although there are several recognized therapies for the treatment of mycosis fungoides, there is a dearth of effective therapies that provide durable responses. Treatment aim to minimize morbidity and limit disease progression, as cure is rarely achieved.(1)

Due to the anatomical location and the presence of blood–brain barrier, MF patients with optic nerve invasion respond poorly to treatments. The treatment of lymphomatous optic neuritis, consists in the application, without delay, of high doses of corticosteroids, possibly reinforced by local irradiation, with intrathecal injections of a chemotherapeutic product (19).

Therefore, as illustrated in our case, we should consider intraocular involvement as a warning of CNS involvement in advanced MF patients(CD8), and usually portends a poor prognosis for survival.

IV. CONCLUSION :

Optic neuritis in non-Hodgkin's lymphoma, is a late complication of the central nervous system and has a poor prognosis. Although an uncommon cause of infiltrative optic neuropathy, optic nerve metastases should be considered in patients with a history of lymphoma (8). CSF examination, when positive with the presence of papilledema and a positive therapeutic effect points to malignant infiltration of the optic nerve. More rarely, when CSF examination is normal with the absence of papilledema and a negative effect of cortisone treatment, this suggests paraneoplastic demyelination or iatrogenic optic neuritis due to radiotherapy and/or chemotherapy, the histopathological proof of which will only be provided at autopsy (19).

Bibliography

[1]. Larocca C, Kupper T. Mycosis Fungoides and Sézary Syndrome. Hematol Oncol Clin North Am. févr 2019;33(1):103‑20.

[2]. Meunier J, Lumbroso-Le Rouic L, Vincent-Salomon A, Dendale R, Asselain B, Arnaud P, et al. Ophthalmologic and intraocular non-Hodgkin's lymphoma: a large single centre study of initial characteristics, natural history, and prognostic factors. Hematol Oncol. déc 2004;22(4):143‑58.

[3]. Finke E, Hage R, Donnio A, Bomahou C, Guyomarch J, Sutter C, et al. Neuropathie optique rétrobulbaire et lymphome malin non hodgkinien. J Fr Ophtalmol. févr 2012;35(2):124.e1-124.e4.

[4]. Jusufbegovic D, Char DH. Clinical Variability of Ocular Involvement in Mycosis Fungoides. JAMA Ophthalmol. 1 mars 2015;133(3):341.

[5]. Spencer WH: Optic nerve, in Spencer WH (ed): Ophthalmic Pathology, Vol3. Philadelphia, WB Saunders, 1986, vol 3, pp 2337-2458.

[6]. Christmas NJ, Mead MD, Richardson EP, Albert DM. Secondary optic nerve tumors. Surv Ophthalmol. 1991;36(3):196‑206.

[7]. Ferry AP, Font RL. Carcinoma metastatic to the eye and orbit. I. A clinicopathologic study of 227 cases. Arch Ophthalmol Chic Ill 1960. oct 1974;92(4):276‑86.

[8]. Kim JL, Mendoza PR, Rashid A, Hayek B, Grossniklaus HE. Optic nerve lymphoma: report of two cases and review of the literature. Surv Ophthalmol. 2015;60(2):153‑65.

[9]. Myers KA, Nikolic A, Romanchuk K, Weis E, Brundler MA, Lafay-Cousin L, et al. Optic neuropathy in the context of leukemia or lymphoma: diagnostic approach to a neuro-oncologic emergency. Neuro-Oncol Pract. mars 2017;4(1):60‑6.

[10]. Kline LB, Garcia JH, Harsh GR. Lymphomatous optic neuropathy. Arch Ophthalmol Chic Ill 1960. nov 1984;102(11):1655‑7.

[11]. Coppeto JR, Monteiro ML, Cannarozzi DB. Optic neuropathy associated with chronic lymphomatous meningitis. J Clin Neuroophthalmol. mars 1988;8(1):39‑45.

[12]. Cohen RG, Hedges TR, Duker JS. Central retinal artery occlusion in a child with T-cell lymphoma. Am J Ophthalmol. juill 1995;120(1):118‑20.

[13]. Hedges TR. III: Case records ofthe Massachusetts General Hospital. N EnglJ Med 318:903-915, 1988.

[14]. Olson ME, Chernik NL, Posner JB. Infiltration of the leptomeninges by systemic cancer. A clinical and pathologic study. Arch Neurol. févr 1974;30(2):122‑37.

[15]. Kawakami Y, Tabuchi K, Ohnishi R, Asari S, Nishimoto A. Primary central nervous system lymphoma. J Neurosurg. avr 1985;62(4):522‑7.

[16]. Schaumburg HH, Plank CR, Adams RD. The reticulum cell sarcoma--microglioma group of brain tumours. A consideration of their clinical features and therapy. Brain J Neurol. 1972;95(2):199‑212.

[17]. Arnold AC, Hepler RS, Foos RY. Isolated metastasis to the optic nerve. Surv Ophthalmol. 1981;26(2):75‑83.

[18]. Harbison JW, Lessell S, Selhorst JB. Neuro-ophthalmology of sphenoid sinus carcinoma. Brain J Neurol. sept 1984;107 (Pt 3):855‑70.

[19]. Neetens A, Clemens A, van den Ende P, Bock RD, Neetens I. Nevrite Optique dans le lymphome non Hodgkinien. Klin Monatsblätter Für Augenheilkd. mai 1992;200(05):525‑8.