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Mycosis fungoides associated with optical neuropathy

Benchekroun Reda*, Arab Lamia*, G. Daghouj*, L. El Maaloum*, B.Allali*, A. El Kettani *

**Pediatric Ophthalmology department , 20 August Hospital , Casablanca , Morocco*

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 is an infiltration of the orbital fat that can lead to a compressive optic neuropathy. Infiltration of the optic nerves and their sheaths by lymphoma remains exceptional.*

Observation *— We report the case of a 38-year-old female patient treated for mycosis fongoide. 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: *LMNH – Mycosis Fungoid – optic nerve infiltration – corticosteroids - chemotherapy*

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I. INTRODUCTION :

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

In LMNH , ocular involvement is rare . Ocular manifestations are usually intraorbital masses, conjunctivo-lacrimal infiltration or posterior uveitis (2). Thus , optic neuropathies are even rarer complications. These last can be the consequence of compression of the optic nerve by a tumoral mass or infiltration of the nerve fibers by lymphoma cells (3). Through this observation 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**)

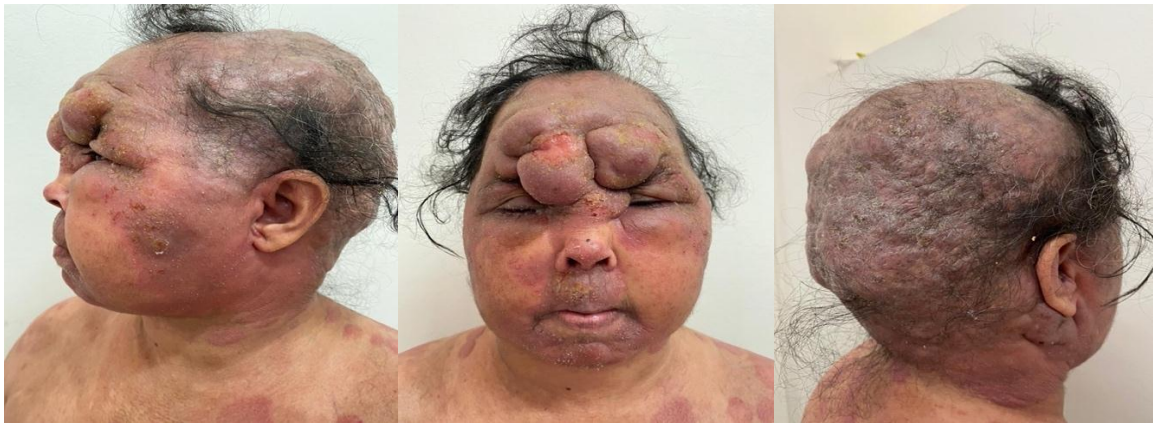

Figure 1 : Photographs showing skin involvement with erythematous scaly swollen patches of the face and scalp.

Cranioorbital 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 , which confirmed the diagnosis .

The patient was treated with 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.

The response to treatment was good initially with recovery of right visual acuity that increased to 2/10 but the patient died 2 months later.

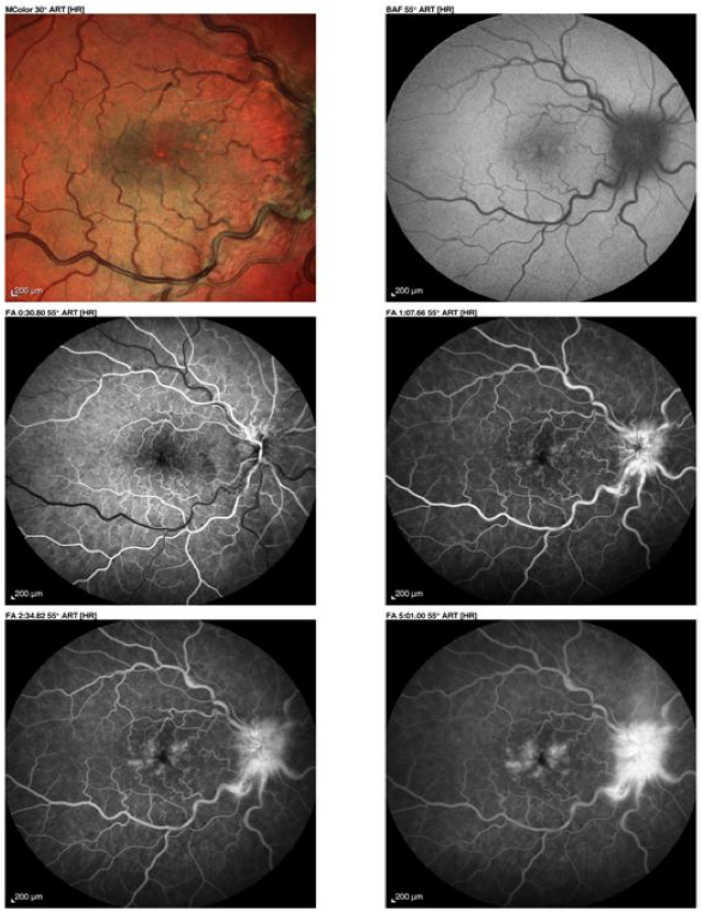

Figure 2 : **Agiographic images showing papillary oedema with papillary fluorescein and macular cubicles**

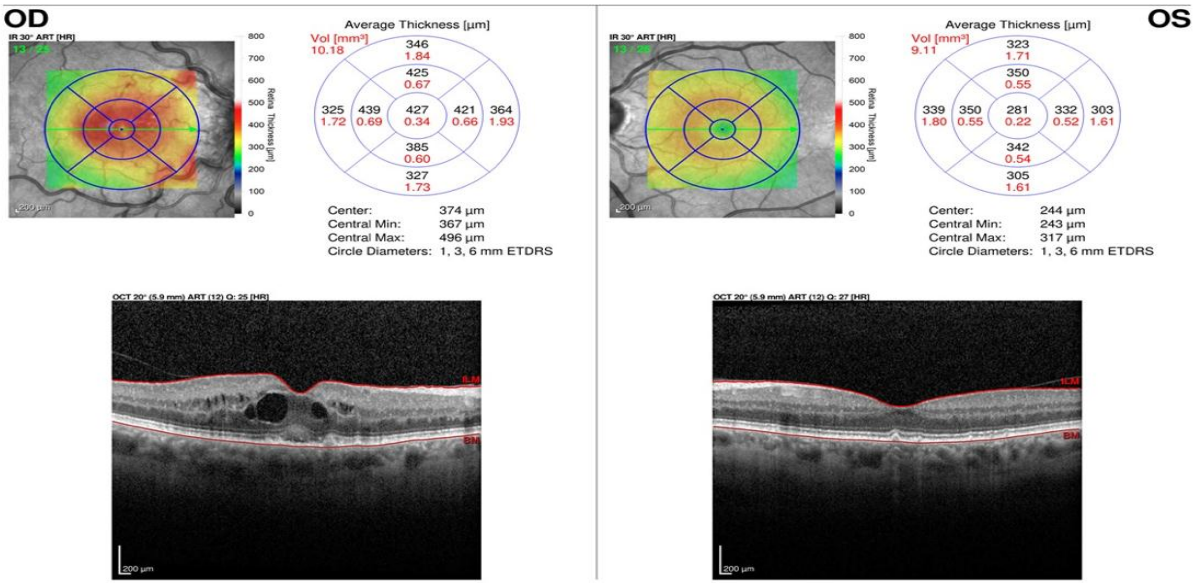

Figure 3 : **Macular OCT showing focal cystoid macular edema on the right.**

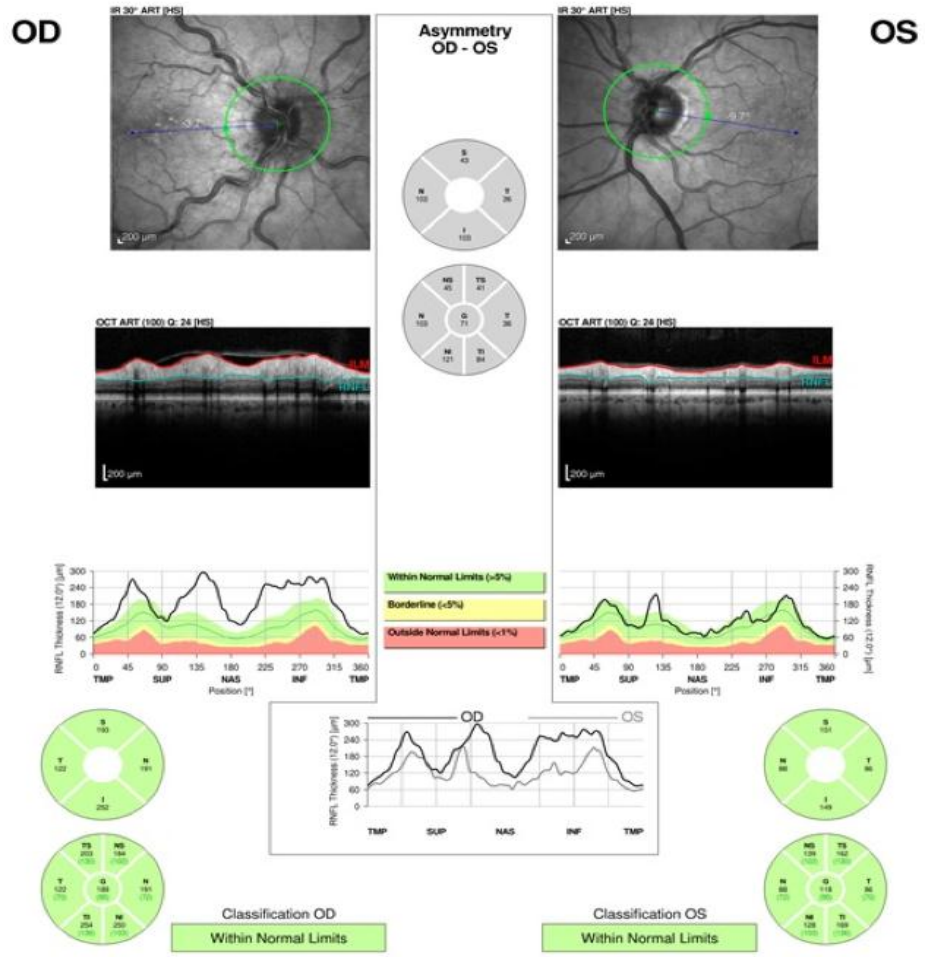

Figure 4 : Papillary OCT showing right papilledema.

III. DISCUSSION:

The optic nerve is more frequently involved by secondary malignancy than by primary malignancy (4) .There are three accepted routes of involvement: direct invasion by ocular melanoma or retinoblastoma; infiltration by lymphoma or leukemia; or blood-borne metastasis from distant malignancies, generally carcinomas. Glioblastoma and other brain tumors have been reported to invade the optic nerve, but late in their course (5).

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 to the eye and orbit.(5–7) . Metastatases to the optic nerve originate most frequently from breast (25-33%) or lung (11-15%) carcinomas, which is consistent with the most common primary tumor metastastatic to the eye and orbit (5,8) . Decreased visual function in non-Hodgkin's lymphoma can have different causes. The most important diagnostic approach is based on CSF examination, combined with the chemotherapeutic test, considering that complications in a period of remission of the disease are probably due to the fact that the blood-brain barrier BBB does not allow the cytostatic product to reach the nervous tissue with the need to consider intrathecal injection of the chemotherapeutic product (9). 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(10) .Case reports highlight compressive optic neuropathy by orbital lymphoma (11), paraneoplastic optic neuritis (12), and central retinal artery occlusion secondary to hyperviscosity (13) . Optic neuropathy may also develop secondary to radiation-related necrosis, vincristine toxicity, or infection (7).

Leukemic or lymphomatous infiltration comprises an estimated 5% of secondary optic nerve tumors(5). Secondary involvement of the central nervous system in systemic lymphoma is less common than primary central nervous system lymphoma (14) however, secondary disease can more often involve the meningeal, perivascular, and spinal epidural areas (7). Tumor cells infiltrate cranial and spinal nerve together with their meninges (15) .The parenchyma of the optic nerve may be invaded by tumor cells (16,17) . 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 (5). 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 (8,18) .

MRI findings in lymphomatous infiltration of the optic nerve include enlargement of the optic nerve and enhancement of the optic nerve sheath(7) . In our case, the infiltration of the optic nerves on MRI, although bilateral, does not cause papilledema on the left side is explained by the predominance of infiltration in the posterior orbital segment of the left optic nerve at a 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 is aimed at minimizing morbidity and limiting disease progression, as cure is rarely achieved.(1) 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 (9).

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 (7) 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 (9). There is no consensus on a standardized treatment given the lack of randomized trials.

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