Lateral Rectus Palsy as First Manifestation of Orbital Lymphoma

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ABSTRACT
Lymphomas are the most common primary orbital tumour in adults 60 years of age and older.

We present a case of a 79-year-old woman complaining of redness in the right eye, tearing and diplopia. She had a history of a thyroid nodule with normal thyroid function, uncontrolled hypertension, and impaired fasting glucose. The Weiss coordimeter showed right eye lateral rectus palsy. Right 6th nerve palsy due to microvascular disease was assumed. At the follow-up visit performed six weeks later, there was a constant esodeviation of the right eye with proptosis and a temporal mass on the right ocular globe. The biopsy was diagnostic of non-Hodgkin’s lymphoma. Differential diagnosis of orbital masses is complex.

Initial manifestations are non-specific and clinical suspicion of an orbital mass may not arise until the considerable growth of the lesion. The relevance of this case is its initial presentation with rapid evolution and the fact that the patient had thyroid disease.

Keywords: Orbital Lymphoma; Oncology; Ophthalmology, Lateral Rectus, Restrictive strabismus.

BACKGROUND
Lymphomas are the most common primary orbital tumour in adults 60 years of age, and older.1 Orbital non-Hodgkin’s lymphoma is rare, comprising only 1% of all non-Hodgkin’s lymphoma and constituting about 8-15% of extranodal non-Hodgkin’s lymphoma.2 Most cases of orbital lymphoma are of low-grade histology.3

CASE REPORT
We present a case of a 79-year-old woman complaining of redness in the right eye...
eye, tearing and diplopia when attending the emergency department. The symptoms had started two weeks before the emergency visit. She had a history of a thyroid nodule with normal thyroid function, uncontrolled hypertension, and impaired fasting glucose. Ophthalmoscopic evaluation revealed visual acuities of 20/25 (Snellen) both eyes, restriction of right eye movement to the right causing diplopia without pain and she had no proptosis. Minor engorgement of the temporal conjunctival vasculature, mild chemosis and an intraocular pressure of 20 mm Hg were the only biomicroscopic findings. The fundoscopic exam was unremarkable. Her blood pressure was 189/119 mm Hg. Neurologic examination was otherwise normal. The Weiss coordimeter showed right eye lateral rectus palsy.

Right 6th nerve palsy due to microvascular disease was assumed, so we scheduled another coordimeter for six weeks later and fogged her glass lens to decrease diplopia.

INVESTIGATIONS

At the follow-up visit performed six weeks later, there was a constant esodeviation of the right eye with proptosis and a temporal mass on the right ocular globe. (Figure 1)

Weiss coordimeter showed an additional superior, inferior and medial rectus involvement.

Ophthalmic ultrasound performed at the follow-up visit revealed a mass shaping the globe reaching to the roof and floor of the orbit. The mass, besides being visible, extended almost to the root of the optic nerve.

The patient also brought previous exams that were performed three months before the emergency visit for the purpose of investigating her thyroid situation (neck CT scan with orbital images). The resulting images led to prompt confirmation of the mass, its location, and apparent rapid growth. (Figure 2)

For better characterization, we ordered an orbital MRI and decided to biopsy the subconjunctival lesion.

Orbital MRI confirmed a homogeneous with well-defined limits mass (isointense relatively to the brain). It was predominantly extraconal and involved the anterior and medial aspects of the right lateral rectus muscle and possibly the right lacrimal gland. The mass moulded itself to pre-existing structures without either erosion of the bone or enlargement of the orbit and without touching the optical nerve.
Histomorphological findings showed densely blue tumour cells. (Figure 3 A)

Immunohistochemistry findings were suggestive of a B-cell chronic lymphocytic leukaemia/small lymphocytic lymphoma: CD20+, Bcl-2+ (focal); ki-67-15%, IgH rearrangement (FR3-JH). (Figure 3 B)

The patient was sent to consultation with haematology and radiotherapy.

Spine MRI and medullar biopsy showed no involvement. She underwent local radiotherapy (28.8 Gy; 16 sessions; 1.8 Gy daily).

Six months after treatment, although there is anatomical (Figure 4) and functional (Figure 5) improvement, the patient has diminished visual acuity of the right eye (20/60 Snellen) and constriction of the visual field. These findings probably correlate to initial signs of radiation retinopathy.

**DISCUSSION**

The most frequent orbital tumours in the adult are lymphoid origin, cavernous hemangiomas, and meningiomas. However, cavernous hemangiomas and meningiomas tend to grow slower. Lymphoma remained as a more plausible diagnosis.

Signs and symptoms of the disease will depend on the lesion location, extension, volume and structure involvement. The most common symptoms and signs of orbital lymphoma are exophthalmos, pain, and ophthalmoplegia.

Imaging (with either CT or MRI) is crucial to characterize the lesion. However, definitive diagnosis is established by biopsy. After the diagnosis, systemic workup for other lymphoma sites is carried out.

The prognosis for orbital lymphomas is closely related to histological type, clinical stage of presentation, and the primary location of the disease.

One study has shown a higher incidence of periocular lymphoma amongst patients with long-standing thyroid disease.

**CONCLUSION**

The differential diagnosis of orbital masses is complex. Initial manifestations are non-specific and clinical suspicion of an orbital mass may not arise until considerable growth of the lesion.

The relevance of this case is its initial presentation with rapid evolution and the fact that the patient had thyroid disease.