Chronic lymphocytic leukemia presenting as unilateral extraocular muscle enlargement and proptosis

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Abstract:
Orbital involvement in chronic lymphocytic leukemia (CLL) is rare with very few published cases. We describe a case of unilateral isolated extraocular muscle enlargement in a patient with CLL. An incisional biopsy was performed from the left medial rectus muscle and histology revealed a lymphocytic infiltrate suggestive of CLL. Complete resolution of signs and symptoms was subsequently achieved with chemotherapy. We would suggest that in patients presenting with atypical clinical features, it is important to consider nonthyroid-related causes of extraocular muscle enlargement and a muscle biopsy should be considered to exclude neoplastic causes.

Keywords:
Chronic lymphocytic leukemia, extraocular muscle enlargement, proptosis

Introduction
Extraocular muscle enlargement is a common feature of thyroid orbitopathy in Graves’ disease. In the literature, the most common nonthyroid causes of muscle enlargement include inflammatory, vascular, and neoplastic processes. In neoplastic cases, the underlying cause is usually either metastatic or orbital lymphoma involving the extraocular muscles. Leukemic infiltration of orbital tissues is rare and comprises <1% of all orbital tumors. Although all types of leukemia may involve the orbit, this is more common in acute leukemias compared to chronic leukemias. A literature search revealed that there are very few published cases of extraocular muscle enlargement due to chronic lymphocytic leukemia (CLL). Orbital leukemic tumors may clinically manifest as proptosis, eyelid edema, pain, motility disturbances, and chemosis.

The authors describe a rare case of unilateral isolated extraocular muscle enlargement in a patient with CLL who responded well to chemotherapy.

Case Report
A 55-year-old Caucasian female presented to the hematologist and was subsequently referred to the oculoplastic clinic, with a 3-month history of worsening left-sided headache associated with pain and swelling of the left upper and lower eyelids. Her medical history included a 4-year history of CLL, which was being managed conservatively without any active treatment and a parathyroidectomy 1 year previously for the treatment of hyperparathyroidism.

On examination, best-corrected visual acuity was 6/6 in her right eye and reduced to 6/18 in her affected left eye. Color vision was mildly affected in her left eye; however, there was no relative afferent pupillary defect. The left upper and lower eyelids were swollen, associated with 3 mm of left-sided axial proptosis and restriction
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A 51-year-old male who developed unilateral leukemic infiltration of the lateral rectus muscle 10 years after a systemic diagnosis of CLL and died 10 months later. The third case described a 53-year-old male with unilateral enlargement of the extraocular muscles; elevated thyroid-stimulating hormone was suggestive of thyroid eye disease, but biopsy of the lateral rectus revealed lymphocytic infiltration suggestive of CLL; he responded well to orbital radiotherapy.

While all previously reported cases were in male patients, our case was in a female patient. This article presents the rare case of a 55-year-old female who developed unilateral enlargement of all the extraocular muscles 4 years after she was diagnosed with CLL. The patient underwent incisional biopsy and subsequent histological analysis confirmed leukemic infiltration. She then made a rapid recovery with chemotherapy.

Conclusion

Although extraocular muscle enlargement is often associated with thyroid-related orbitopathy, it is important to consider other causes, such as inflammatory, vascular, or neoplastic processes. A diagnostic extraocular muscle biopsy should be considered in patients presenting with atypical clinical features.

Acknowledgment

We would like to thank Dr. George R Powell and Dr. Nicholas J Green, Consultant Histopathologists, for interpreting and providing photos of the histological findings.

Discussion

Isolated extraocular muscle involvement secondary to CLL is extremely rare. There have been only three published cases of extraocular muscle enlargement due to CLL. One case described a 72-year-old male who presented with bilateral enlargement of all extraocular muscles due to CLL and responded well to orbital radiotherapy. The second case described a 51-year-old male who developed unilateral leukemic infiltration of the lateral rectus muscle 10 years after a systemic diagnosis of CLL and died 10 months later. The third case described a 53-year-old male with unilateral enlargement of the extraocular muscles; elevated thyroid-stimulating hormone was suggestive of thyroid eye disease, but biopsy of the lateral rectus revealed lymphocytic infiltration suggestive of CLL; he responded well to orbital radiotherapy.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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