Ptosis as an Initial Manifestation of Orbital Lymphoma: A Case Report

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Ptosis on its own is an infrequent initial manifestation of orbital lymphoma. Orbital lymphoma usually presents as a palpable mass with proptosis, diplopia, and conjunctival ("salmon-pink") swelling. We report here a 62-year-old female patient who presented with right eye ptosis. The initial imaging study showed an indistinct enlargement of the superior rectus–levator muscle complex. After 3–4 months, ptosis and upward gazing movement were further restricted. The imaging study revealed a definite soft-tissue mass in the superior orbit surrounding the superior rectus–levator muscle complex. A tumor biopsy through anterior orbitotomy revealed a large diffuse B-cell lymphoma. With the experience of this case, we suggest that orbital lymphoma should be included in the differential diagnosis of ptosis accompanied by impairment of levator muscle function.

Key Words: orbital lymphoma, ptosis
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Ptosis on its own is an infrequent initial manifestation of orbital lymphoma [1]. Orbital lymphoma usually presents with proptosis, periorbital swelling, conjunctival ("salmon-pink") swelling, diplopia, and conjunctival redness and irritation [2–6]. It can remain indolent for a long period of time before the patient notices the periorbital abnormality [2–6]. Often seen are proptotic cases in association with various degrees of ptosis. Here we report an unusual presentation of orbital lymphoma with an initial manifestation of ptosis on its own.

Case Presentation

A 62-year-old woman was sent for consultation for right eye ptosis and mild ocular pain that began about 1 month earlier (Figure 1). Margin reflex distance (MRD) was 1 mm on the right eye and 6 mm on the left. The levator muscle function was 8 mm on the right eye and 15 mm on the left. The supraduction of the right eye was mildly limited. There was neither proptosis nor diplopia. An initial computed tomography (CT) scan (Figure 2) and magnetic resonance imaging (MRI) of the orbit and brain revealed an indistinct mass in the superior orbit close to, or within, the superior rectus–levator muscle complex. The neostigmine test and anti-acetylcholine antibody were both negative. She was treated with prednisolone 15 mg/day and pyridostigmine bromide (Mestinon) by the neurologist who thought the

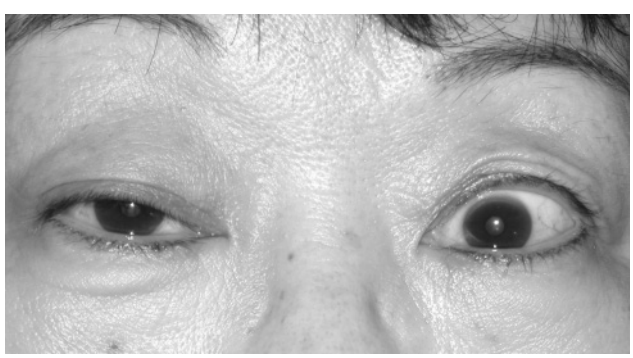

Figure 1. *Patient presented with right eyelid ptosis. Margin reflex distance was 1 mm on the right eye and 6 mm on the left eye. Levator muscle function was 8 mm in the right eye and 15 mm in the left eye.*

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patient was suffering from myasthenia gravis. During the 2-month period of treatment, the ptosis progressed. The MRD measured 1 mm on the right eye. Without strong evidence of an orbital mass lesion, orbitotomy for orbital exploration under general anesthesia was not strongly considered. After discussion with the patient, an exploration of the anterior orbit, in combination with a levator muscle resection to correct ptosis, was performed under local anesthesia. Histopathologic examination showed inflammation only. Five months later, supraduction was further limited, and the ptosis recurred after levator resection. The second CT scan and MRI (Figure 3) of the orbit revealed a definite soft-tissue mass in the superior orbit surrounding the superior rectus–levator muscle complex. A tumor biopsy via anterior orbitotomy revealed a large diffuse B-cell lymphoma. Immunohistochemistry showed that the malignant cells were CD20 positive and CD3 negative (Figure 4). Stage IEA (Ann Arbor criteria) was determined after detailed systemic survey. The patient received a complete therapeutic course of chemotherapy with a regimen of cyclophosphamide, epirubicin, oncovin and prednisolone, and monoclonal antibody therapy (anti-CD20 antibody). A follow-up CT scan showed that the orbital mass had disappeared (Figure 5). However, the right eye ptosis did not improve, with an MRD of –3 mm and very limited levator muscle function. The supraduction was mildly limited.

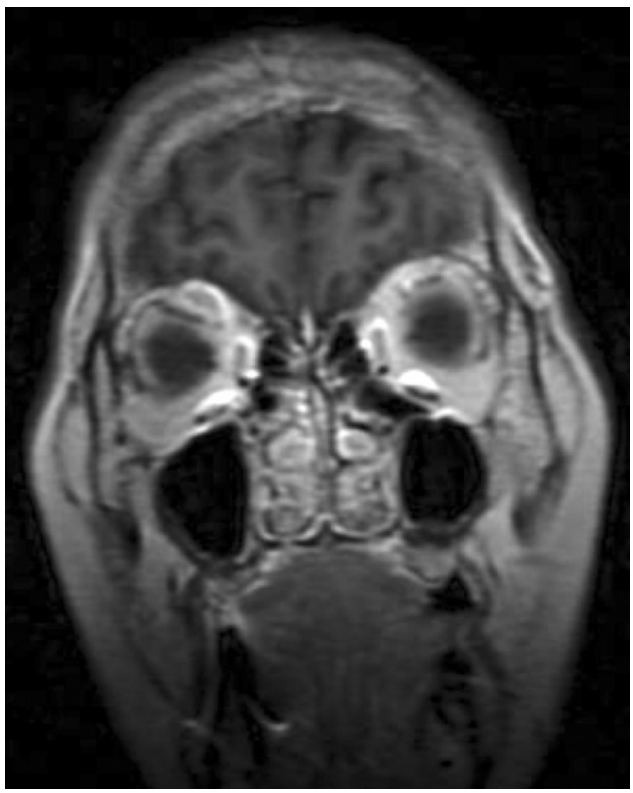

Figure 2. *A soft-tissue mass was indistinctly visible in the superior orbit surrounding or within the superior rectus–levator muscle complex.*

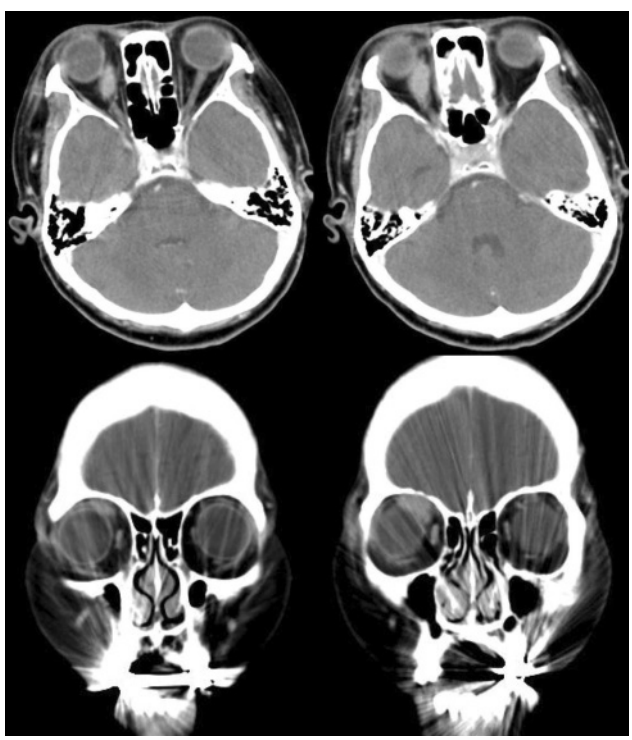

Figure 3. *MRI scan of the orbit revealed a definite soft-tissue mass in the superior orbit surrounding the superior rectus–levator muscle complex.*

Discussion

The patient in this case presented predominantly with ptosis. The imaging study did not show a clear orbital mass, but only a mild enlargement of the superior rectus–levator muscle complex. However, the slight suspicion of an orbital tumor did not justify the performance of an invasive diagnostic exploration of the orbit via orbitotomy under general anesthesia. The lesion could have been an inflammation or a benign reactive lymphoid hyperplasia. In cases of inflammation, the use of immunosuppression or anti-inflammatory treatment might be of help, whereas surgical intervention could be detrimental to levator function of the eyelid or supraduction of the eyeball.

Prednisolone had been tried as the initial treatment. In cases of myasthenia gravis or idiopathic orbital inflammatory syndrome, steroid treatment can be of help [7,8]. However, the ptosis in this patient did not improve, and supraduction limitation developed. Therefore, a follow-up CT scan was performed, and a definite orbital mass was

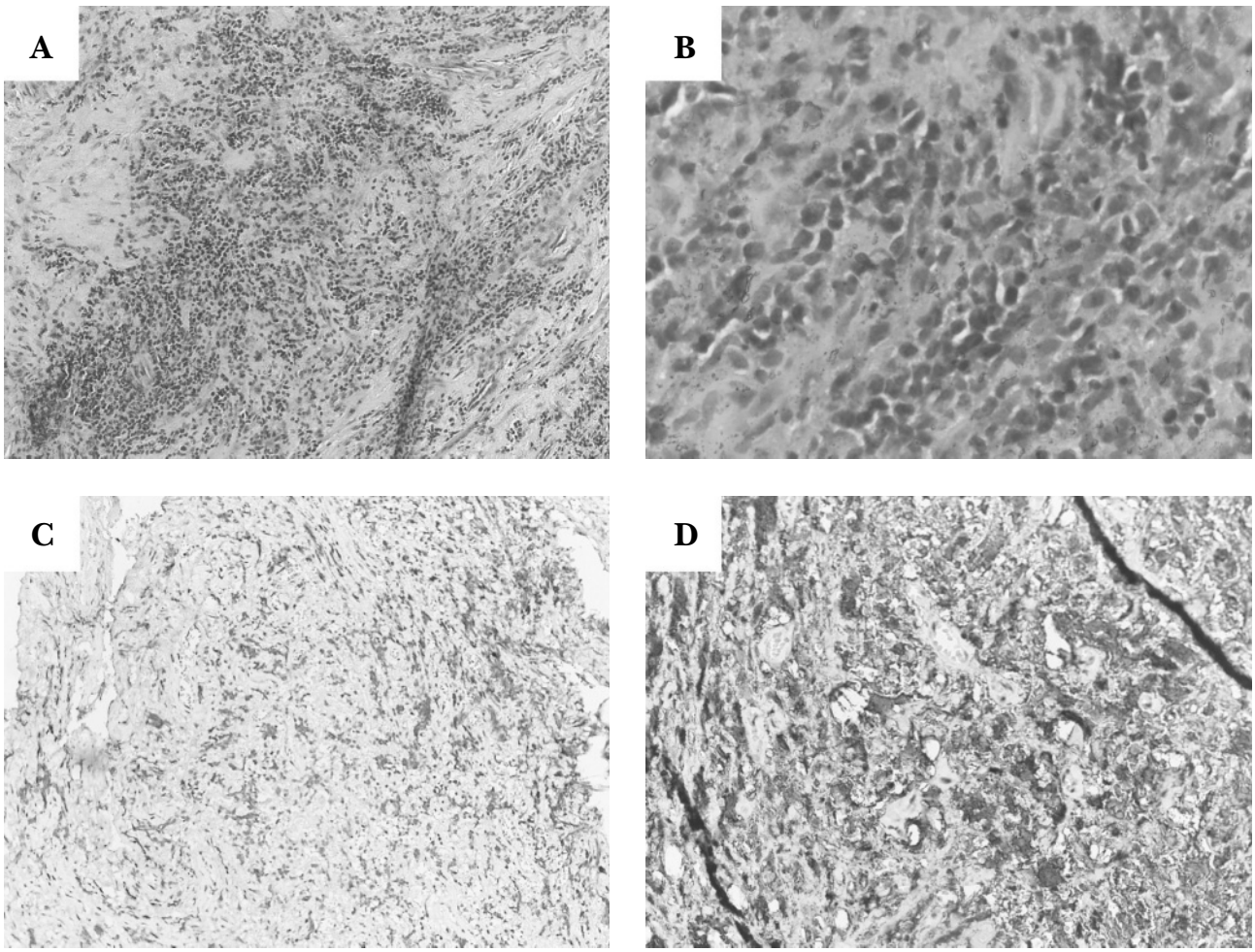

Figure 4. *Microscopically, the soft tissue is infiltrated by large monotonous lymphoid cells diffusely. They have hyperchromatic or vesicular nuclei. There are foci of punctate necrosis and mitotic activity (A, B). The immunohistochemical stain demonstrates: $CD3^-$ (C) and $CD20^+$ (D).*

identified. Only at this point was the ophthalmologist comfortable with advising the patient to have the invasive diagnostic surgery. Nevertheless, initiation of the treatment for orbital lymphoma at that point was not too late. The patient had a complete remission of the tumor after chemotherapy.

On CT scan, orbital lymphoma usually appears as a homogeneous molding mass with sharp margins, most often localized in the anterior portion of the orbit, the retrobulbar area, or the superior orbital compartment [9,10]. The tumor mass rarely erodes bones and does not cause much disturbance of ocular motility, lid excursion, or visual function. More oppressive forms that infiltrate the orbit and erode bone are also possible, but are unusual [11,12]. A case of isolated orbital lymphoma presenting as blindness with an unusual feature of bone erosion has been reported [13].

Orbital lymphomas arising from extraocular muscles are unusual. Hornblass et al reported six cases of orbital lymphoma with similar symptoms [1], which were ptosis or supraduction disturbance overshadowing exophthalmos. The lesions were all located within the superior rectus–levator muscle complex. Lymphoma located within the extraocular muscles (EOM) has been called "EOM lymphoma"[14]. The most common presenting complaints of such lymphoma have been diplopia, ptosis, and lid fullness [1,14–17], whereas pain, conjunctival vasocongestion, or lid erythema are not typical features. Involvement of the superior rectus–levator muscle complex was the most common manifestation [15], followed by involvement of the medial rectus [1,16] and lateral rectus muscles [1]. Involvement of the inferior oblique muscle [14,17] has occasionally been observed as well.

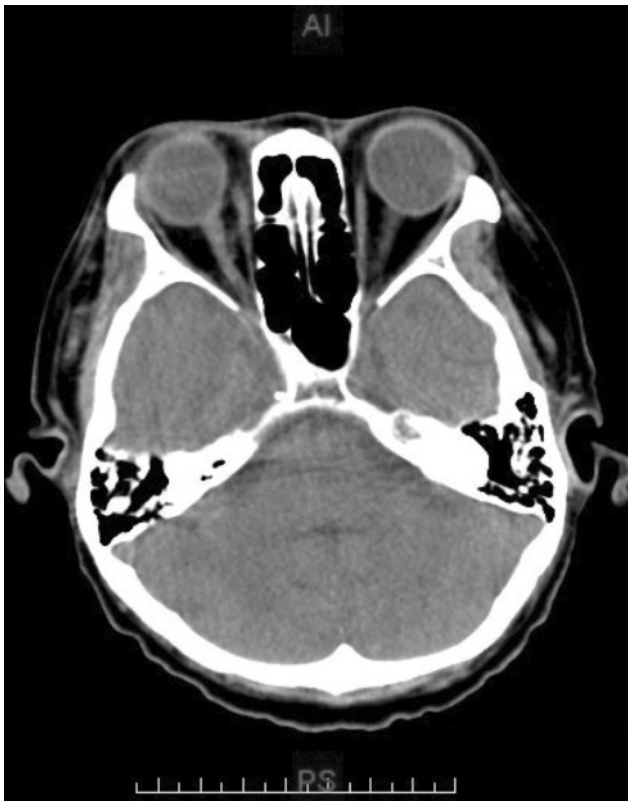

Figure 5. *Follow-up CT scan after chemotherapy showed that the orbital mass had disappeared.*

This case presented a situation of symptom manifestation earlier than confirmed by imaging evidence. The decision to intervene surgically for diagnosis is always debatable. However, close follow-up is the best policy. EOM lymphoma should always be considered within the differential diagnosis of ptosis.

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以眼瞼下垂為最初表現的眼窩淋巴瘤之病例報告

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眼窩淋巴瘤臨床上單獨以眼瞼下垂為表現並不常見，通常以伴隨著腫塊的凸眼、複視、結膜鮭魚樣色紅腫為表現。我們提出一位 62 歲女性病例最初僅以右眼眼瞼下垂為臨床表現，初期影像學檢查僅有不明顯之上直肌及提眼瞼肌肥大，持續追蹤數個月後，眼瞼下垂及眼球向上運動受限進一步惡化，此時影像檢查呈現出明顯眼窩腫塊，手術後病理學檢查確診為眼窩淋巴瘤。根據此病例，我們建議臨床上患者單獨出現提眼瞼肌失能的眼瞼下垂，眼窩淋巴瘤應為可能診斷之一。

關鍵詞：眼瞼下垂，眼窩淋巴瘤

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