A Vanishing Lymphoma in the Cheek

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Abstract

Background: We describe the unusual case of a 66-year-old woman who presented with a cheek mass that completely abated with oral steroids. Case Report: Multiple separate biopsies of the mass were negative or inconclusive. MRI revealed a large mass, yet after a short steroid course, this mass was completely undetectable on clinical examination. A repeat biopsy eventually revealed follicular lymphoma. Discussion: Lymphomas are known to be steroid sensitive; the medication is an essential component of the common CHOP therapy. While known to occur in the central nervous system, to the best of our knowledge, the presence of a ‘vanishing’ lymphoma has not been documented in the head and neck. We discuss the likely physiology of the vanishing lymphoma, and the diagnostic difficulty it presents. Conclusion: When a lymphoma is suspected, patient care may be optimized if biopsy is delayed until steroids have been discontinued.

Introduction

Short courses of corticosteroids have been documented to cause clinical and radiologic regression in central nervous system (CNS) lymphomas and may cause inconclusive biopsies [1–4]. This behavior creates a diagnostic challenge [5–7]. To the best of our knowledge, these ‘vanishing’ lymphomas have not been reported outside of the CNS and specifically not localized to the head and neck. In this report, we document a lymphoma in the cheek, which disappeared with corticosteroid therapy obstructing its diagnosis. We also briefly review the literature on disappearing lymphomas.
Case Report

A 66-year-old woman presented to our Otolaryngology Clinic with a 2-year history of left cheek swelling and a recent CT scan that showed an ill-defined soft-tissue process in her left malar region. She noted that the swelling would regress when taking oral steroids. A Mohs surgeon had recently performed a biopsy of the area, but remained suspicious due to the persistent mass. The patient denied any systemic symptoms. Her medical history was significant for asthma and sinus allergies as well as previous Mohs resection of a basal cell carcinoma of her left alar crease. Examination revealed mild fullness over the left cheek. A trans-facial biopsy was performed. Microscopic examination revealed fibroconnective tissue with dense lymphohistiocytic infiltrate and associated fibrosis. The lesion was thought to be an inflammatory mass and was followed with repeat scans.

The patient continued to complain of recurrent swelling. Nine months later, a follow-up MRI showed a large $3 \times 4 \times 5$ cm mass in the same location (fig. 1). The patient presented to the clinic 2 weeks later after having taken a short course of oral steroids for unrelated asthma symptoms. Surprisingly, she now did not have a palpable or visible mass. Ultrasound examination was also negative. She reported that the facial mass as well as an enlarged cervical lymph node disappeared after initiating steroids. Biopsy was repeated after steroid discontinuation. Microscopic evaluation now revealed atypical follicular proliferation with invasion of surrounding structures suggesting a diagnosis of follicular B-cell lymphoma (fig. 2).

The patient was referred to Oncology Services for further management. Currently she is doing well with rituximab and methylprednisone for therapy.

**Fig. 1.** MRI shows a left cheek mass (arrow) at follow-up during a period with no steroid use.

**Fig. 2.** High-power HE stain demonstrates lymphocytic infiltration into the muscle, adipose, and nervous tissue. This pattern of infiltration resembling a lymphoid follicle but without marginal zone differentiation is consistent with follicular lymphoma.
Discussion

Lymphoma of the cheek is an uncommon disease [8–10]. Extramedullary lymphoma is thought to represent only 20–30% of lymphomas; it most commonly presents in the GI tract, followed by the head and neck [11]. In the head and neck, it most commonly presents in Waldeyer’s ring, and then in the paranasal sinuses [12]. Lymphoma of the cheek is thought to account for only 2.5% of malignant lymphomas [13]. Interventions may involve surgical excision, chemotherapy, and radiotherapy.

To our knowledge, this is the first case of a vanishing lymphoma reported in the head and neck. In our patient, the mass would regress with steroids and enlarge between courses; moreover, initial biopsy after recent steroids was negative. In fact, our patient underwent 3 separate biopsies before a diagnosis was made. Lymphomas that completely regress with glucocorticoids are known to occur in the CNS [1]. It also has been suggested that glucocorticoids induce some regression in one third of CNS lymphoma cases [2, 3].

Negative biopsies for lymphomas in patients taking steroids have also been documented in the CNS [3, 5, 6]. Consequently, it is recommended that patients with a possible lymphoma discontinue steroids for at least 5 days before undergoing biopsy [7]. Outside of the CNS, corticosteroids have also been suggested to complicate the diagnosis of splenic lymphomas [14]. This effect is likely due to the selective destruction of the lymphoma cells within the biopsy specimen along with surrounding inflammatory changes caused by the tumor [14].

Multiple mechanisms have been suggested for the glucocorticoid-induced regression of lymphomas. In the CNS, these include the direct lympholytic effect of steroids, alterations in the permeability of the blood-brain barrier, and steroid-induced vasoconstriction compromising the blood supply to the tumor [4]. Due to the rapid tumor regression over 2 weeks, we favored steroid-induced apoptosis in our patient. Steroids are known to have a strong lympholytic effect and form an essential part of the CHOP therapy for most lymphomas.

The recurrence of these tumors after finishing glucocorticoids has also been observed in the CNS. Both clonal evolution and subclone selection have been theorized to be the cause. Studies investigating IgG rearrangement and DNA sequence analysis have favored clonal evolution [2].

In summary, a lymphoma should be considered in a subcutaneous mass that clinically regresses with steroids. Such vanishing lymphomas have been well documented in the CNS, yet rarely observed outside of it. If a lymphoma is suspected, patient care may be optimized and correct diagnosis may be achieved by delaying biopsy for at least 1 week after steroids are discontinued.

Disclosure Statement

The authors have no conflicts of interest to declare.

References


