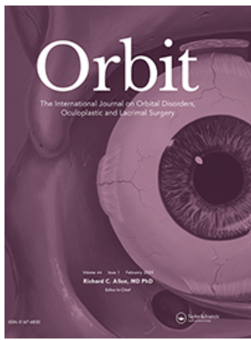

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CASE REPORT

Sarcoid-like reaction of the orbit in diffuse large B-cell lymphoma

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ABSTRACT
Sarcoid-like reaction (SLR) has been reported in patients with solid tumor malignancies, lymphomas, and patients receiving immunotherapy. SLR is often incidentally found during positron emission tomography/computed tomography scans as hilar and/or mediastinal lymphadenopathy. SLR has also been found in the lung, spleen, bone marrow, and skin. Biopsy of these lesions shows noncaseating granulomas. When systemic criteria are not met for sarcoidosis, these noncaseating granulomas are termed SLR. We present the first case in the literature of a case of orbital SLR in a patient with concomitant diffuse large B-cell lymphoma and inverted papilloma of the maxillary sinus. This case highlights the importance of including malignancy in the differential for the presence of a noncaseating granuloma in the orbit.

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Sarcoid; lymphoma; paraneoplastic; proptosis; orbital cellulitis

Sarcoid-like reaction (SLR) is defined as the development of noncaseating granulomas in patients who do not meet systemic criteria for sarcoidosis. SLR has been found to occur with immunotherapy and malignancy.[1–3] We present a case of SLR in the orbit in the setting of concomitant diffuse large B-cell lymphoma (DLBCL) and inverted papilloma.

Case presentation

A 52-year-old male with a history of end-stage renal disease on hemodialysis, heart failure, and previously treated follicular lymphoma presented as a transfer from an outside institution for "orbital cellulitis."

Seven years prior to presentation, the patient underwent treatment for a grade 3A follicular lymphoma. The patient was treated with 5 cycles of bendamustine and rituximab. However, he failed to complete the sixth and final cycle due to multiple hospitalizations for infections. Due to a lapse in insurance coverage, he was then also lost to follow-up for 2 years, after which he presented with increasing abdominal pain. Computed tomography (CT) scan of the abdomen demonstrated increased lymph node size. Positron emission tomography-computed tomography (PET-CT) scan found a right maxillary sinus soft tissue mass extending into the right nasal passage, enlarged retroperitoneal and mesenteric lymph nodes, and a large hypoattenuating mass within the spleen. He underwent biopsy of a retroperitoneal node with pathology concerning for necrotic B-cell lymphoma. After discussion with his oncologist, the patient elected to forgo treatment for his lymphoma in the setting of his multiple comorbidities.

He was asymptomatic for 4 years until he presented to an outside institution with progressive right orbital swelling and pain for 2 months. The CT scan from the outside institution showed proptosis of the right globe with stranding and thickening of the right periorbital region with no identifiable abscess. Also notable on the CT scan was paranasal sinus disease with complete opacification of the right maxillary sinus with soft tissue density extending from the right maxillary sinus to the right nasal passage. He was empirically treated with intravenous vancomycin, cefepime, amphotericin B, and oral levofloxacin. However, his chemosis and extraocular movement restriction did not improve. Given the lack of treatment response, the patient was transferred to our institution for escalation of care.

On presentation to our institution, the patient had a near visual acuity without correction of 20/70 in the right eye and 20/40 in the left eye without a relative afferent pupillary defect. Intraocular pressures were normal. The patient was found to have lid edema, severe chemosis, and right eye gaze restriction in all directions of gaze (Figure 1). The left eye exam was unremarkable. Given a continued concern for an infectious etiology, antibiotics were continued pending additional workup.

Magnetic resonance imaging (MRI) orbits with and without contrast showed edema and enhancement of

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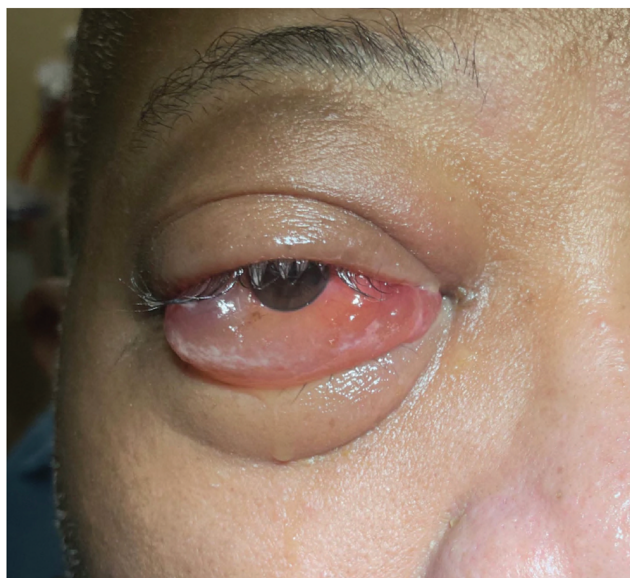

Figure 1. External photograph of the right eye on presentation displaying lid edema with severe inferior conjunctival chemosis and erythema.

the right intraorbital and periorbital tissue as well as a mass of the right maxillary sinus extending into the right nasal cavity (Figure 2). Given the MRI findings, the patient underwent functional endoscopic sinus surgery (FESS) with biopsy, septoplasty, removal of tumor, and maxillary antrostomy with the otolaryngology team. Intraoperatively, there was no purulence or necrosis noted. Following FESS, the patient had improvement in his pain, extraocular motility, chemosis, and visual acuity to 20/25 without correction in the right eye. Pathology of the mass was consistent with inverted papilloma. Given no evidence of infection intraoperatively, clinical improvement after surgery, and no systemic symptoms while admitted (fevers/chills/white count elevation), the patient's antibiotics were discontinued. He received 3 days of intravenous solumedrol 1 g and had continued improvement in his pain, extraocular motility, and chemosis. He was discharged home with close outpatient follow-up.

Four days after discharge, the patient presented to our emergency department for worsening vision and pressure around the right eye. His visual acuity in the right eye had decreased to 20/100 without correction without improvement on pinhole, but without an afferent pupillary defect. On exam, he had further decreased extraocular movements and worsened chemosis on the right side.

Repeat MRI orbits with and without contrast showed post-surgical changes of the right maxillary sinus and redemonstration of edema and enhancement of the right orbital fat with increased stretching of the optic nerve. The case was discussed with otorhinolaryngology, who reported that the inverted papilloma was removed in its entirety. Given the worsening visual acuity, chemosis, extraocular movements, and orbital inflammation on imaging, the patient was started on 60 mg of prednisone by mouth and arranged close outpatient follow-up.

At his clinic follow-up visit, the patient continued to have edema and decreased vision (20/200 without correction that did not improve with pinhole) of the right eye, albeit without an afferent pupillary defect. Given the continued swelling, the patient was scheduled for urgent right anterior orbitotomy.

During the orbitotomy, a firm palpable mass was dissected out of the inferior orbit via a transconjunctival approach. The mass was noted to be nonencapsulated and infiltrated into the surrounding tissues. Pathology demonstrated noncaseating granulomas suggestive of sarcoidosis (Figure 3).

The patient was then referred to the uveitis service for evaluation of possible sarcoidosis. Given his extensive oncological history, the patient was suspected to have a paraneoplastic SLR associated with his previously established malignancy.

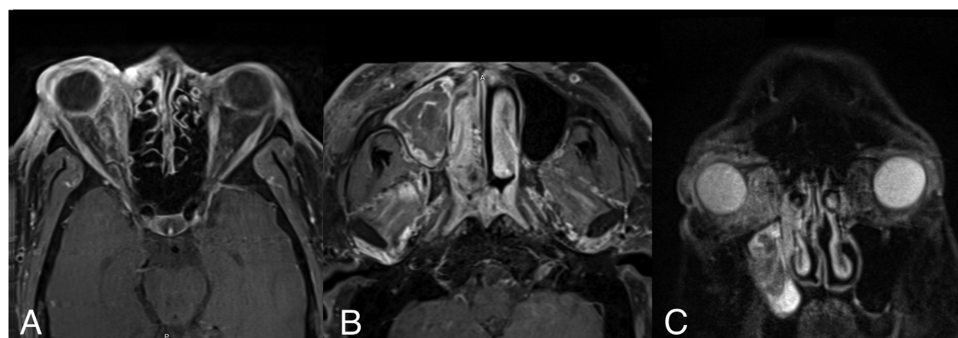

Figure 2. MRI brain and orbits with contrast at presentation. A) T1 Dixon post-contrast water axial cut showing edema and enhancement of the right intraorbital and periorbital tissue. B) T1 Dixon post-contrast water axial cut showing a mass filling the right maxillary sinus and extending to the nasal cavity. C) T2 STIR post-contrast coronal cut showing both edema and enhancement of the intraorbital tissue as well as mass in the right maxillary sinus.

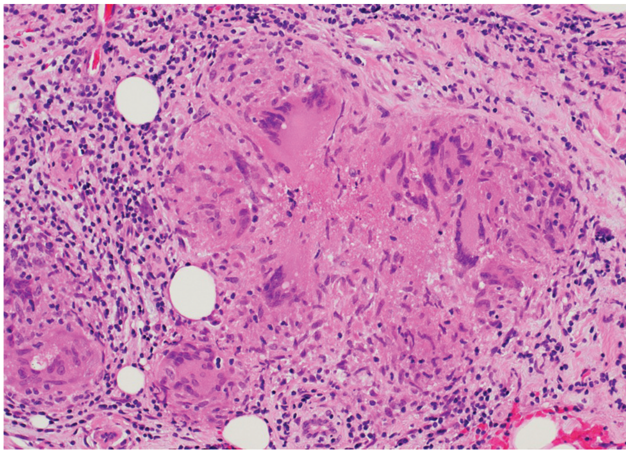

Figure 3. Histology from biopsy stained with hematoxylin and eosin showing a large noncaseating granuloma with numerous giant cells in a background of chronic inflammatory cells.

A repeat PET-CT found new increased uptake in the right orbital region medially, a right supraclavicular lymph node, mediastinal and hilar lymph nodes, and the spleen. The patient underwent biopsy of the splenic mass. Pathology from the biopsy was consistent with a high-grade transformation to DLBCL (CD19+, CD20+, CD22+, CD 45+) with an IGH/BCL2 translocation in 100% of the cells. Due to his vision-threatening orbital disease, despite his comorbidities, the patient was initiated on chemotherapy with rituximab, cyclophosphamide, vincristine, and doxorubicin. After 5 cycles, he has had improvement in his chemosis, pain, and extraocular motility (Figure 4).

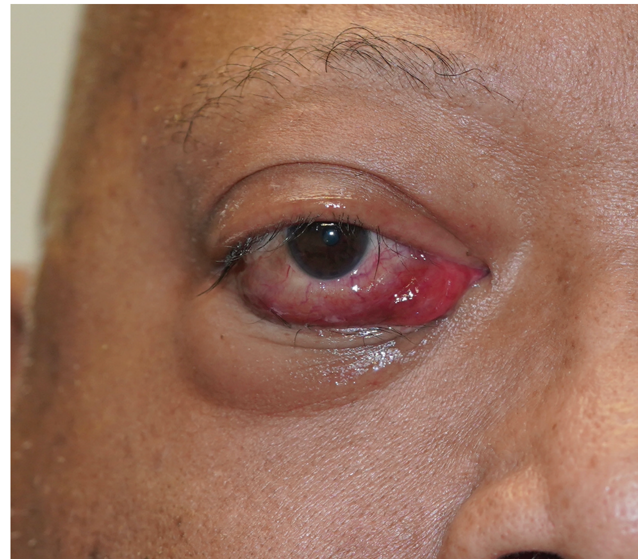

Figure 4. External photograph of the right eye after five rounds of chemotherapy displaying improved lid edema with improved inferior conjunctival chemosis and erythema.

Discussion

SLR has been reported in the setting of various malignancies including colon, breast, and non-squamous lung cancer.[2,3] SLR is also known to occur in the setting of lymphomas, with rates up to 13.8% of patients with Hodgkin's lymphoma and 7.3% of patients with non-Hodgkin's lymphomas.[4–6] SLR has also been reportedly induced by treatment with checkpoint inhibitors.[1]

The exact mechanism of SLR is poorly understood. It is currently theorized that it represents an activation of macrophages against humoral or T-cell mediated factors.[7] SLR typically manifests as mediastinal and hilar lymphadenopathy, incidentally found during a restaging PET/CT scan.[2,3,8] Biopsy of these lymph nodes shows noncaseating granulomas. In addition to mediastinal and hilar lymphadenopathy, SLR has also been described in the lung, spleen, bone marrow, and skin.[9] There is no consensus on the treatment of SLR, with some lesions found to be self-resolving, some responsive to steroids, and some responsive to removal of the insult.[1,2]

We present the first case in the literature of SLR in the orbit in the setting of underlying DLBCL and inverted papilloma. This case highlights the importance of reevaluating the diagnosis of orbital cellulitis when there is a lack of response to antibiotics. This case also emphasizes that an oncologic process should be considered when orbital biopsy shows noncaseating granuloma.

Disclosure statement

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