Spontaneous subperiosteal orbital hematoma as a presenting sign of hairy cell leukemia in a patient with a long-standing orbital implant

Subperiosteal orbital hemorrhage most commonly results from blunt trauma. Though rare, nontraumatic occurrences also have been reported from sudden increases in intracranial venous pressure, sinusitis, or bleeding diathesis.[1] There has been only 1 prior reported spontaneous subperiosteal orbital hematoma (SSOH) resulting from a hematologic malignancy: chronic myelogenous leukemia.[2]

Herein we present a case of SSOH as the presenting sign of hairy cell leukemia (HCL) in a patient with a history of orbital fracture repair. The collection and evaluation of patient health information were compliant with the Health Insurance Portability and Accountability Act, and this report adheres to the Declaration of Helsinki.

A 47-year-old male presented with acute left-sided proptosis and diplopia. He had been otherwise well following successful orbitotomy to treat an incompletely reduced left orbital floor fracture with a porous polyethylene–titanium mesh implant (Medpor Titan, Stryker Craniomaxillofacial, Kalamazoo, Mich.) 4 years prior. His examination demonstrated normal visual acuity, normal intraocular pressure, and unremarkable slit-lamp and dilated fundus examinations without subconjunctival or retinal hemorrhages or any other signs of ocular trauma. On the left side there was limitation of extraocular motility, 2 mm of relative proptosis, and upper eyelid ptosis. Review of systems revealed recent weight gain and easy bruising, but he denied fevers, chills, night sweats, and recent trauma. Neuroimaging showed a 2.8 cm ovoid subperiosteal elevation of intermediate attenuation tracking along the rostral aspect of the orbital floor implant without adjacent orbital fat stranding, consistent with a hematic cyst (Fig. 1). A trial of 60 mg of prednisone failed to produce significant clinical improvement, and surgical drainage was recommended.

However, before surgery could be performed, a preoperative systemic evaluation revealed pancytopenia: white blood cells, 760/μL; hemoglobin, 9.20.76 $\times$ 10^6/μL; and platelets, 21,000/μL. A review of the 5 prior years' medical records showed mild thrombocytopenia (95,000/μL) of unknown etiology. The patient was admitted for further work-up and was found to have splenomegaly. Subsequent bone marrow core biopsy was notable for hypercellular marrow extensively involved by small to intermediate-sized lymphoid cells with round to irregular nuclei and abundant pale-pink cytoplasm. The bone marrow aspirate smear showed an increased number of atypical lymphocytes, including some with "hairy projections," as well as markedly diminished trilineage hematopoiesis and background stromal fibrosis, consistent with a diagnosis of HCL (**Supplementary Fig. 1**, available online). Immunohistochemical and special stains were performed and positive for CD20, CD79a, PAX5 (approximately 70%–80% of marrow cellularity), Annexin A1, CD103, and BRAF V600E, consistent with classic HCL. Flow cytometry identified an aberrant monotypic B-cell population that was lambda light chain restricted and positive for CD19, CD20, CD79a, CD10, CD103, CD43, CD11c, FMC7, CD25, HLADR, s/cIgM, and sigD. Cytogenetics demonstrated a normal karyotype. Molecular diagnostics confirmed the presence of BRAF V600E mutation.

Four months after treatment with cladribine monotherapy, a purine nucleoside analogue, proptosis and diplopia had resolved without any surgical intervention and the patient's platelet count had increased to 122,000/μL. Posttreatment neuroimaging showed near-complete resolution of the subperiosteal collection (**Supplementary Fig. 2**, available online). Posttreatment bone marrow biopsy confirmed a complete clinical remission, with 0.04% residual hairy cells, without recurrence 2 years later.

HCL is a rare, indolent B-cell leukemia in which characteristic mature B cells with abundant cytoplasm and "hairy" projections accumulate within the bone marrow, splenic red pulp, and peripheral blood. Although cytopenias, easy bruising, and splenomegaly are common, ocular disturbances in HCL are rare and include retinopathy, panuveitis, and corneal infiltration.[3] To our knowledge, orbital pathology such as SSOH has not been reported previously in HCL.

SSOH has been reported as a delayed phenomenon after orbital surgery, including orbital wall reconstruction with alloplastic implants, bony decompression for thyroid eye disease, and enucleation. Orbital implants may induce a surrounding fibrous capsule (nonporous implants) or allow for fibrous ingrowth (porous implants), and hemorrhages can arise on rupture of capillaries within the capsule or perforating veins within the implant. SSOH has presented symptomatically as late as 16 years after repair; such delayed hemorrhage is potentially attributed to subclinical trauma, implant migration, or chronic irritation.[4] Histopathologic evaluation of explanted porous polyethylene orbital implants has demonstrated granulation tissue and inflammatory debris of the implant and surrounding tissues.

We hypothesize that the hemorrhage in this case resulted from chronic low-grade inflammation produced by the implant and leading to the formation of granulation tissue and subsequent hemorrhage potentially from capillary rupture. The bleeding was likely exacerbated in the setting of thrombocytopenia due to HCL. In the previously reported case of spontaneous hemorrhage as a presenting sign of chronic myelogenous leukemia, the diagnosis was made with histologic evaluation of the evacuated hematoma. Hemorrhage was attributed to leukostasis—obstruction of the microvasculature by blast cells leading to hypoxia, cytokine release, and endothelial damage.[2]

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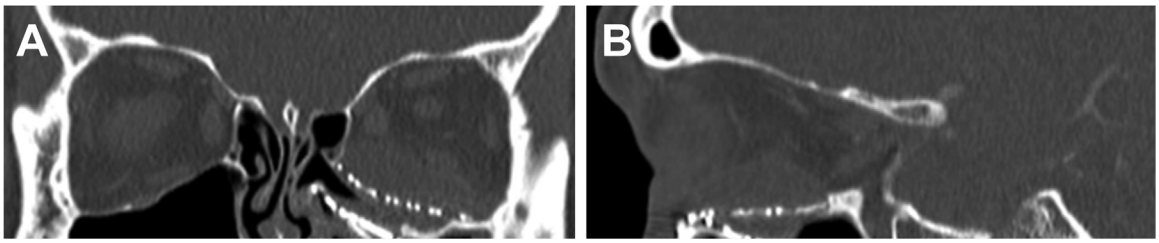

Fig. 1—Orbital imaging at presentation: (A) coronal and (B) sagittal computed tomography scan showing a 2.8 cm ovoid subperiosteal elevation of intermediate attenuation tracking along the rostral aspect of the orbital floor implant without adjacent orbital fat stranding, consistent with a hematic cyst.

Orbital leukemic tumours are rare but occur more commonly with acute myeloid leukemia than with other subtypes of leukemia, often in the first decade of life.[5] Orbital hematomas and leukemic orbital masses can be difficult to resolve by neuroimaging. Orbital hematomas, however, tend to contain greater heterogeneity depending on chronicity and are commonly located along the orbital roof, whereas leukemic orbital masses tend to be homogeneous with a predilection for the lateral orbital wall.[5] Without a diagnostic orbitotomy, leukemic orbital infiltrate cannot be excluded in the present case. However, hematoma was favoured given the contemporaneous acute symptomatology and worsened thrombocytopenia, the location of the subperiosteal infiltrate, and the relative rarity of orbital leukemic infiltration.

SSOH is rare and can result from a variety of postoperative and medical conditions. We describe the first case of HCL presenting with SSOH in a patient with a history of orbital floor reconstruction. Hematologic malignancy should be carefully considered in patients presenting with spontaneous orbital hemorrhages, especially because timely diagnosis can increase survival.

Supplementary materials

Supplementary material associated with this article can be found in the online version at doi:10.1016/j.jcjo.2023.02.002.

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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this correspondence.