Orbital Deep Granuloma Annulare in an Adult

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Abstract

We herein describe the second case in the literature of an adult with a deep GA confined to the orbit. We could not identify any potential etiology from the patient’s history, and neither recurrence nor additional lesions have been noted after 2 years of follow-up.

Keywords: Granuloma Annulare; Orbital Neoplasms; Pathology, Surgical

Granuloma annulare (GA) is a benign idiopathic granulomatous skin and soft tissue disorder. The nodular subcutaneous or deep form (deep GA) bears histopathological resemblance to the granulomas of rheumatic fever and rheumatoid arthritis, leading to the term pseudorheumatoid nodule. Characteristic histopathology includes a necrobiotic collagen center with palisading histiocytes surrounding a central core of mucin, best seen on Alcian blue staining.1,2 Deep GA is most commonly encountered in children, and usually affects the lower extremities or scalp. The prognosis is favorable with observation or excision, and spontaneous partial or complete regression may occur.1-10 Evidentiary treatment information, however, is limited due to the rarity of this lesion.2 Despite characteristic histopathology, atypical presentations may pose a challenge for clinical and radiological diagnosis. Although various periocular presentations have been reported, very few reports describe orbital deep GA.4-10 We present a case of an adult male with deep GA confined to the orbit.

A 32-year-old Caucasian male was referred with a one-month history of left upper eyelid swelling that failed to improve with oral and topical antibiotics. The patient reported no significant past medical or surgical history and denied any family history of ocular or autoimmune disease. On examination, optic nerve, eyelid and extraocular muscle functions were normal. There was a 3 mm left proptosis with a 1 mm left upper lid ptosis and mild fullness of the superior sulcus. There was no erythema, injection, or chemosis, and the lesion was not palpable through the skin or identifiable through the superior fornix. Computed tomography (CT) without contrast demonstrated a left anterior orbital mass that appeared to involve the belly of the levator muscle in close relation to the superior rectus. (Figure 1A).

An excisional biopsy was performed via left anterior orbitotomy, and a well-defined hard lesion was encountered adjacent to the antero-lateral aspect of the superior rectus, embedded in thelevator muscle belly. The lesion, measuring 9 x 8 x 4 mm, was completely removed with care to minimize dissection of surrounding muscle (Figure 1B). Histopathology revealed fibromuscular tissue with palisading granulomatous inflammation, central necrobiosis and fibrin (Figures 2A-
B. Periodic acid-Schiff and Fite's stains were negative for fungi and mycobacteria, respectively. Alcian blue demonstrated mucin within the granuloma (Figure 2C), and trichrome highlighted concentric fibrosis around granulomatous foci, yielding the diagnosis of a deep GA. A thorough investigation for systemic inflammatory and autoimmune disease was unremarkable, including: complete blood count with differential, blood urea nitrogen, creatinine, erythrocyte sedimentation rate, C-reactive protein, anti-nuclear antibodies, anti-neutrophil antibodies, neutrophil cytoplasmic antibodies, anti-DNA antibodies, anti-SSA/SSB, rheumatoid factor, RPR, Hepatitis B and C antibodies and antigens, blood cultures for bacteria and fungus, free thyroxine, thyroid stimulating hormone (TSH), and TSH-receptor antibody.

To our knowledge, this is only the second case in the literature of an adult with a deep GA confined to the orbit. We could not identify any potential etiology from the patient's history, and neither recurrence nor additional lesions have been noted after 2 years of follow-up.

Discussion

The term pseudorheumatoid nodule was first used by Mesara et al. in 1966 to describe a series of subcutaneous nodules with necrobiotic collagenous centers in 12 children (mean age 4 years). The locations varied, with most occurring on the lower extremities overlying bony protuberances and several over the occipital skull. No patients had evidence of rheumatic or rheumatoid disease at presentation or during extended follow-up. Treatments included observation or excision, with a high rate of recurrence and development of additional lesions in various locations.

In 1975, Rao et al. published the first report of ocular adnexal pseudorheumatoid nodules in 21 patients with a mean age of 19 years (range 3–37 years) and a mean follow-up of 6 years. The lesions occurred most frequently on the lateral upper eyelids and lateral canthi, although lesions also occurred on the lower lids and medial canthi. All lesions were treated with local excision, with 3 patients noting recurrence at the site of excision and 3 patients noting lesions in new sites including the periorbita, scalp and chin. No significant difference in gender or ethnicity was observed. A recent review by Chiang et al. found 14 cases of pediatric periorbital GA with a mean age of 5 years (range 1–11 years). Most lesions were located on the upper eyelid and no sex predilection was noted, however a majority of these cases occurred in African American individuals. With respect to orbital deep GA, Chiang's review included a 1982 report of an 8-year-old with a lower lid lesion extending into the orbit, and 2006 report of a 2-year-old with a subperiosteal lesion adherent to the superior orbital rim. In addition, Ross et al. described a 29-year-old woman with numerous pseudorheumatoid nodules involving the upper lid,
episclera and orbit, including the medial rectus muscle. After an extensive literature search, we found only two cases of an isolated deep GA confined to the orbit. One lesion involved the lateral rectus muscle of a 13-year-old boy with a history of traumatic injury to that eye 7 months prior. The other lesion involved the medial and inferior rectus muscles of an 86-year-old man with a history of prostate cancer.

As there are many differential diagnoses of subcutaneous nodules including rheumatoid nodules, metastatic lesions, sarcoid granulomas, tuberculous granulomas, tophi and xanthomas, it is important to carefully evaluate patients for systemic disease. Biopsy is required to confirm the diagnosis of deep GA. The prognosis is favorable as these lesions are self-limiting and may shrink or resolve with time. There is little evidentiary literature on the treatment of deep GA, however common treatments include observation and excision with reported local recurrence rates of 20% to 40%. Although some associations have been made with trauma, viral infection or paraneoplastic syndromes, the etiology remains unknown and patients do not typically develop systemic autoimmune disease.

Given the benign course of this lesion and the multitude of more concerning differential diagnoses, we would like to make readers aware of this uncommon variation of deep GA presenting as an orbital mass.

References

Figure 2.—A. Subcutaneous tissue with a large necrobiotic nodule with surrounding fibrosis (Hematoxylin and Eosin, 1x). B. Palisading granuloma surrounding a core of brightly eosinophilic fibrin rich necrobiotic (Hematoxylin and Eosin, 20x). C. Abundant mucin is present in the center of the palisading granuloma (Alcian blue, 20x).