Subperiosteal deep granuloma annulare of the orbital rim

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OBJECTIVES:
• To review the presentation of deep granuloma annulare
• To discuss prior reports of this relatively rare lesion in the head and neck area

INTRODUCTION
Granuloma annulare is a benign, usually self-limited lesion that most commonly occurs on the extremities. Facial and scalp masses of this type are rare, and usually occur in the superficial skin. Deep granuloma annulare of the head and neck is an exceedingly rare lesion, and to date, has only been reported in women and children. We report the first case of this type of lesion in an adult male.

CASE REPORT
A 52-year-old Caucasian male with rheumatoid arthritis presented with a five-week history of a 1 cm left upper lid central mass. He denied antecedent trauma, insect bites, recent viral illness, or other skin lesions or changes. He had no recent fevers, chills, weight loss, or night sweats. His past medical history was significant only for hypertension and rheumatoid arthritis, with a positive Rheumatoid factor and anti-CCP antibodies. His medications included methotrexate, sulfasalazine, intermittent Prednisone, and folate. On exam the mass was non-tender and appeared fixed to the lateral aspect of the left superior orbital rim. There was an additional flesh-colored plaque involving the skin overlying the medial aspect of the superior lid consistent with a xanthoma. He had no associated diplopia, vision changes, or ocular discharge. There was no change in his lid closure.

The patient underwent magnetic resonance imaging (MRI) for further characterization of the lesion. (See Figure 1B). This demonstrated a 6 mm heterogeneous enhancing nodule within the lateral aspect of the left superior palpebra. The lesion was located between the intrapalpebral component of the orbicularis oculi muscles and the orbital septum, causing bulging of the orbicularis oculi muscles. The lacrimal gland did not appear to be involved.

An excisional biopsy with orbitotomy and resection of the left upper lid mass was performed utilizing an upper lid blepharoplasty incision. An incision was made in the left eyelid along the dominant lid crease superiorly, extending through the skin and orbicularis muscle. Dissection was carried out between the orbitopalpebral component and the orbicularis muscle fibers, where the mass was noted to be adherent to the peristome of the upper orbital rim. The periosteum was sharply incised and a plane was defined between the lesion and healthy orbital bone. The incision was closed with 6-0 fast absorbing plain gut suture. Upon follow up, the patient has been free of recurrence. Histologic evaluation revealed palisading granulomas with central necrobiosis. A colloid iron stain revealed central zones of connective tissue mucin within the granulomatous foci. (See Figure 3A and 3B). The specimen was interpreted by a head and neck pathologist and a dermatopathologist, both of whom suggested that the histology was consistent with deep granuloma annulare or a rheumatoid nodule.

DISCUSSION
Granuloma annulare (GA) is a benign, granulomatous dermatosis that is usually a self-limited disease. There are four well-documented subtypes of GA:
- Localized
- Generalized
- Perforating
- Subcutaneous

The subcutaneous form is the least common type, which is characterized by a deeper location with occasional fixation to underlying bone or soft tissues. Subperiosteal deep is a rare form of the subcutaneous subtype.

The exact etiology of GA remains unknown but multiple etiologies have been suggested, including trauma, healing herpes zoster scars, verruca vulgaris, bites from the giant Caiman crocodilus, tuberculin skin tests, and diabetes. There has even been report of GA occurring after an octopus bite. Because of the tendency of GA to be associated with the extremities, sunlight has also been implicated.

The type of treatment or the need for treatment at all, is also debated. Several treatment modalities exist, including intralesional and topical steroids, local excision, cryotherapy, laser therapy, and photo-therapy.

Prognosis of GA is excellent, with spontaneous remission seen within months to years regardless of therapy. Recurrences are reported at 15-40%, and can usually be treated conservatively.

Interestingly, this patient has rheumatoid arthritis and no evidence of granuloma annulare at other sites. Approximately 20% of patients with RA will develop a rheumatoid nodule; 50% of patients with subcutaneous GA will manifest RA at other sites. The histopathology of rheumatoid nodule is similar to subcutaneous granuloma annulare, and the criteria for distinguishing these two entities remains controversial. While many authors indicate that the presence of abundant connective tissue mucin (hyaluronic acid) favors the diagnosis of subcutaneous GA, there have been reports of mucin in clinically characteristic rheumatoid nodules. Some authors believe the distinction cannot be made histologically and should be based on clinical findings and/or the presence of RA.

Whether this lesion represents a rheumatoid nodule or subcutaneous GA, it is an exceedingly rare occurrence at this site.

REFERENCES