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Orbital Pseudorheumatoid Nodule

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Pseudorheumatoid nodules are necrobiotic granulomas that are not associated with any systemic disease but that histopathologically mimic the granulomas of rheumatic fever, rheumatoid arthritis, and systemic lupus erythematosus [1–4]. Pseudorheumatoid nodules usually involve the skin of the extremities or scalp, but may occasionally involve the periorcular area [5–7]. We report an unusual case of pseudorheumatoid nodule demonstrated by computed tomography (CT) to be confined to the orbit, where its appearance mimicked that of intraorbital neoplasms.

Case Report

A 13-year-old boy was struck in the left eye with a friend's elbow while playing baseball 7 months before admission. At that time, he had acute swelling and ecchymosis of the eye. The swelling resolved spontaneously over several days, although he later said that his eye remained slightly swollen. Several days before admission, the patient awoke with acute swelling and pain of the left eye. He was taken to a local clinic and treated with antibiotics for 2 weeks for what was believed to be orbital cellulitis. Although the pain had diminished, the orbital swelling had not decreased in this interval. The patient was referred to the Illinois Eye and Ear Infirmary for further evaluation and treatment.

Physical examination revealed periorbital swelling and tenderness on the left and hyperemic left conjunctiva. Vital signs and the rest of the physical examination were normal. Complete blood cell count with differential cell counts, urinalysis, and chest film were normal. The serologic tests that were performed were all negative: fluorescent antinuclear antibody, lupus erythematosus preparation, antistreptolysin O titer, rheumatoid factor, and C3 and C4 complement titers.

Initial CT of the orbits without and with intravenous contrast material showed a large, slightly enhancing soft-tissue mass containing several hypodense areas in the region of the left lateral rectus muscle (fig. 1A). The radiologic differential diagnosis on the basis of the CT findings included rhabdomyosarcoma, pseudotumor, hemangioma, lymphangioma, and dacryoadenitis with retroorbital infection. Subperiosteal hematoma was also included because of the history of trauma. Because hemangioma was a possibility, a dynamic CT flow study was performed using rapid sequential scans after a bolus injection of contrast material; it did not show sufficient contrast enhancement of the mass to suggest hemangioma.

A diagnostic biopsy after the initial CT study suggested pseudorheumatoid nodule. The prominent microscopic feature was necrotizing granuloma formation, with a marked zonal pattern, lying in the connective tissue (fig. 1B). Immediately surrounding the large central areas of collagen necrosis was a zone of fibroblasts and macrophages (fig. 1C), some of which showed epithelial characteristics with occasional multinucleate giant-cell formation. A cellular infiltrate, consisting predominantly of lymphocytes and macrophages, surrounded the central zone. Within the inflammatory infiltrate, there was capillary endothelial cell proliferation.

On the basis of the diagnosis of pseudorheumatoid nodule, no specific medical treatment was administered. The patient was followed by periodic CT studies. The latest examination, performed 11 months after the initial CT scan, showed almost complete resolution of the mass (fig. 1D).

Discussion

Pseudorheumatoid nodule was introduced by Mesara et al. [1] in 1966 to describe the occurrence of subcutaneous connective tissue granulomas in otherwise healthy children. They described 12 children aged 3–8 years, none of whom showed evidence of rheumatic fever or rheumatoid arthritis during follow-ups of 1–17 years. Many synonyms have been used for pseudorheumatoid nodules, including rheumaticlike nodules, rheumatoidlike nodules, and granuloma annulare. The nodules are seen most often in children but can also occur in adults. They are typically located on the extensor surfaces of the extremities, the dorsa of the hands or feet, and the scalp. Involvement of the ocular and periorcular tissues is much less common.

In 1978, Rao and Font [5] reported pseudorheumatoid nodules of the ocular adnexa in 21 patients. In their study, the mean patient age was 19, and there was no predilection for gender or race. Most patients (16 of 21) had solitary lesions, and the other five had multiple nodules. The most common locations of the nodules in this series were the lateral upper eyelid (eight of 21 patients) and the lateral canthus (eight of 21). Clinical follow-up was available in 15 patients, three of whom had one or more recurrences.

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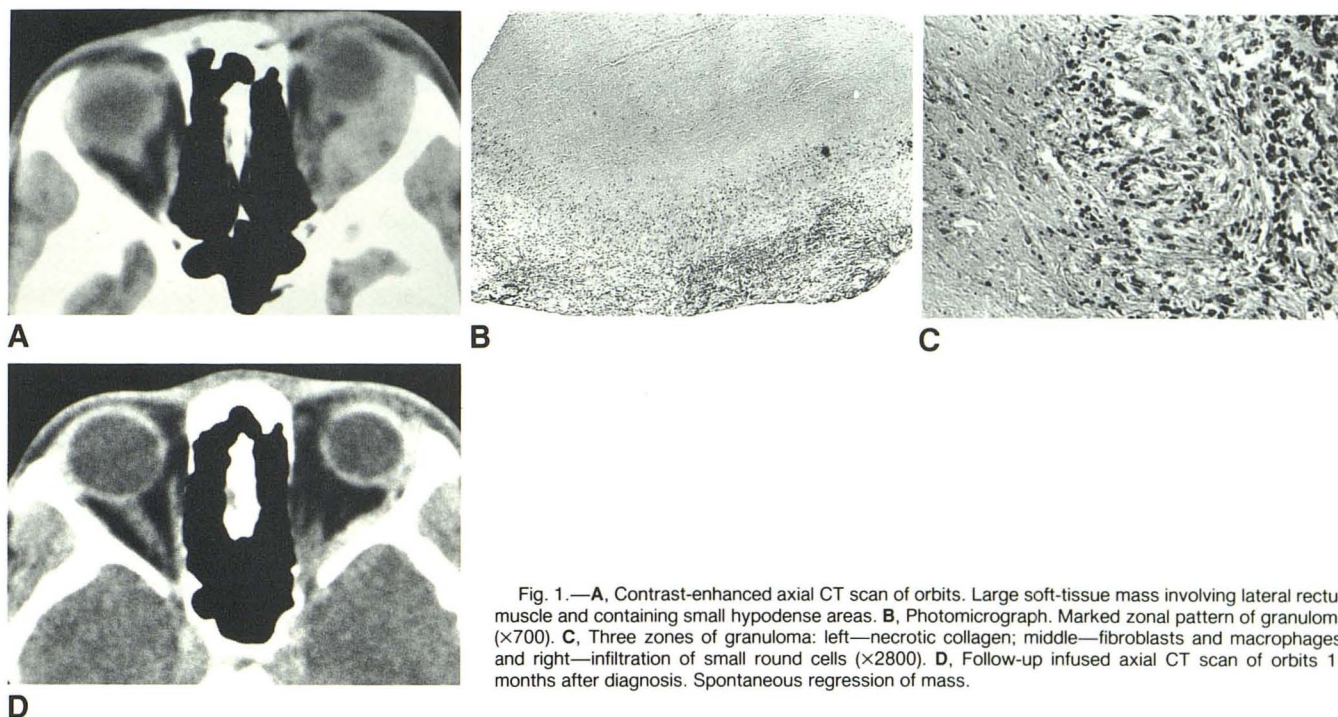


Fig. 1.—A, Contrast-enhanced axial CT scan of orbits. Large soft-tissue mass involving lateral rectus muscle and containing small hypodense areas. B, Photomicrograph. Marked zonal pattern of granuloma ($\times 700$). C, Three zones of granuloma: left—necrotic collagen; middle—fibroblasts and macrophages; and right—infiltration of small round cells ($\times 2800$). D, Follow-up infused axial CT scan of orbits 11 months after diagnosis. Spontaneous regression of mass.

Ross et al. [7] recently reported a 29-year-old woman with pseudorheumatoid nodule involving, concomitantly, the eyelid, eyebrow, episclera, and orbit. This was the first report of a surgically-proven case of pseudorheumatoid nodule involving an extraocular muscle. We believe ours to be the second reported case of proven pseudorheumatoid nodule involving the extraocular muscles. To the best of our knowledge, our case also represents the first report of pseudorheumatoid nodule confined to the orbit and involving an extraocular muscle alone, without involvement of the periorbital tissues and eyelids. Based on our case and that of Ross et al., CT appears to be very useful for determining the presence or absence of orbital involvement with pseudorheumatoid nodule.

The cause of pseudorheumatoid nodule remains unknown. Although Beatty [1] postulated that the lesions could represent an unusual reaction to trauma, other workers have been unable to substantiate this theory [2–4]. The correct histopathologic diagnosis is of paramount importance because of the benign nature of the condition and its spontaneous regression. Patients should be managed with periodic follow-up examinations to monitor nodule regression [5]. CT permits

both localization for biopsy and follow-up monitoring of the intraorbital component of pseudorheumatoid nodule, when present. Pseudorheumatoid nodule should be included in the differential diagnosis when CT demonstrates a soft-tissue mass in the orbit, particularly in children.

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