Idiopathic Granulomatous Orbital Inflammation

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Idiopathic granulomatous inflammation is regarded as a histopathologic variant of an orbital pseudotumor and has a similar clinical presentation and treatment. However, the differential diagnosis including sarcoidosis, Wegener's granulomatosis, trauma, and infection must be ruled out. We present a 77-year-old man with a biopsy-proven lacrimal gland granulomatous inflammation of an unknown origin. Management by surgical debulking via an anterior orbitotomy achieved a good result with minimal complications. (Chang Gung Med J 2003;26:847-50)

Key words: idiopathic granulomatous inflammation, orbital pseudotumor, anterior orbitotomy.

When an orbital mass is composed of noncaseating granulomatous inflammation and the diagnostic tests fail to show systemic sarcoidosis or other identifiable local or systemic causes, a diagnosis of idiopathic granulomatous orbital inflammation may be established.

Idiopathic granulomatous orbital inflammation is an uncommon lesion. Recently, it has been regarded as a histopathologic variant of idiopathic orbital inflammation (IOI or pseudotumor) because of similar clinical and therapeutic behaviors. In this study, we present the clinical features, histopathologic analysis, therapy, and outcome of our patient.

CASE REPORT

A 77-year-old man presented with a 2-month history of right upper eyelid swelling. Ocular examination revealed 3 mm of proptosis, S-shape ptosis, and a palpable painless indurative mass in the superolateral orbit. However, no significant ocular inflammation was found (Fig. 1). A complete blood count and the lysozyme level were within normal limits. The antinuclear cytoplasmic antibody (ANCA) test was negative. A chest radiograph was normal. Orbital computed tomography showed a well-defined 2.1×2.5-cm superotemporal lacrimal gland mass with a mold of the globe (Fig. 2). Then, surgical debulking via an anterior orbitotomy was performed. The operative finding revealed that the tumor was rubbery firm, and adhered to the levator

Fig. 1 A 77-year-old man presented with insidious onset of ptosis of the right upper eyelid and slight downward displacement of the globe. An indurative mass in the superolateral orbit was found. Note the absence of significant inflammatory signs.
muscle without no distinct border. The pathology examination demonstrated noncaseating granulomas, infiltration of plasma cells, lymphocytes, and multinucleated giant cells (Figs. 3, 4). Acid-fast stain, PAS stain, and GMS stain of the biopsy tissue were all negative. Postoperatively, the proptosis of the globe and swelling of the right upper eyelid completely resolved. Meanwhile, Schirmer’s test did not show dry eye. Thirteen months after the operation, the granulomatous orbital inflammation had not recurred. However, mild ptosis persisted, and the patient received a corrective operation later (Fig. 5).

**Fig. 2** CT scan demonstrating a right superotemporal lacrimal gland mass with a mold of the globe.

**Fig. 3** Lacrimal gland biopsy revealing a granulomatous inflammation distorting the acini of the gland. Lymphoplasmatic infiltration and a multinucleated giant cell can be noted. (H&E, 200×)

**Fig. 4** Idiopathic granulomatous inflammation of the lacrimal gland. A well-formed noncaseating granuloma was characterized by epithelioid clusters with admixed lymphocytes and scattered multinucleated giant cells. (H&E, 200×)

**Fig. 5** After anterior orbitotomy and ptosis correction. This patient achieved good results.

**DISCUSSION**

Among lacrimal gland tumefactions, pseudotumors (idiopathic orbital inflammation), lymphoid tumors, and epithelial tumors are the 3 most common. However, a preoperative diagnosis is difficult. By the use of clinical history and computed tomography (CT), Jakobiec et al. evaluated 39 patients with primary lacrimal gland fossa lesions. They demonstrated that contour analysis of the soft-tissue mass is a valuable diagnostic method. The inflammatory lesions or lymphoid tumors presented diffuse and
molded enlargement of the lacrimal gland. Meanwhile, the epithelial tumors exhibited rounded or globular soft-tissue outlines. They also found that patients with a lymphoid tumor were much older than those with other lesions, with a mean age of 71 years. In addition, a large survey of orbital tumors in adults older than 60 years found that malignant lymphoma was the most commonly encountered tumor. Therefore, because of our patient’s old age and the absence of significant inflammatory signs and radiographic findings, a lymphoid tumor was highly suspected. A biopsy is always required to verify the diagnosis.

The early literature reported 30 patients with a diagnosis suggestive of granulomatous orbital pseudotumor without systemic involvement. Their clinical presentations did not differ from that of histopathologic classical pseudotumor. Idiopathic granulomatous inflammation may present clinically with an acute, subacute, or chronic onset, characterized by signs of a mass effect or inflammation or both. It is more likely to be located in the lacrimal gland, accounting for 50% of cases. In regard to treatment, the majority were managed successfully with 1 or more courses of systemic corticosteroids, while partial tumor excision, done in a few cases, was not successful. However, in a study of 7 patients, 4 tumors located in the lacrimal gland or anterior orbit were effectively managed with partial or complete surgical excision. As for our patient with a circumscribed mass located in the anterior orbit, the result of surgical debulking via an anterior orbitotomy was satisfactory, and there were very few complications.

However, the differential diagnosis of an orbital mass lesion composed of granulomatous inflammation includes certain systemic and local causes. The prime suspect is sarcoidosis. Sarcoidosis is a multi-system granulomatous disorder that affects the lungs in nearly 90% of patients with a significant prevalence in Africans. Anterior granulomatous uveitis is the most common ocular manifestation, and orbital involvement is typically bilateral. Although the serum ACE test was not done on our patient, his chest radiographs and follow-up showed no relation to sarcoidosis. Furthermore, the possibility of Wegener’s granulomatosis must be considered because it will lead to grave visual or systemic consequences. Wegener’s granulomatosis may present atypically in the orbit, with granulomatous inflammation as the only histopathologic finding, and without vasculitis or tissue necrosis. A determination of ANCA will help to trace this disease. Furthermore, granuloma formation can be an inflammatory response to infection with tuberculosis, syphilis fungus, or parasites. A ruptured dermoid cyst, foreign body, or blunt orbital trauma can elicit a granulomatous inflammatory reaction as well. Therefore, a detailed historical investigation and histopathological study such as a routine hematoxylin-eosin (H&E)-stained section, polarized light microscopy, methenamine silver, and Ziehl-Neelsen stain for fungi and acid-fast bacilli are needed.

In conclusion, when all identifiable causes of orbital granulomatous inflammation have been ruled out, a diagnosis of idiopathic granulomatous inflammation is established. Although idiopathic granulomatous orbital inflammation responds to steroid therapy, but the circumscribed mass located in the anterior orbit, total or subtotal resection of the orbital mass is easily accessible, we prefer surgical intervention.

REFERENCES

原發性眼眶肉芽腫炎症

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原發性眼眶肉芽腫炎症為眼眶間接腫瘤炎症之病理組織學上的變異。但是必須先排除一些會引起肉芽腫之局部或全身疾病，包括類肉瘤病，wegener's肉芽腫症，外傷或感染等，才可以歸類為原發性眼框肉芽腫炎症。而它的臨床表現與治療大體上與眼眶間接腫瘤炎症類似，主要以藥物治療為主。我們在這篇文章報告一位77歲男性表現為一個侷限於淚腺之眼眶肉芽腫炎症，經過我們手術切除後得到很好的效果。(長庚醫訊 2003:26:847-50)

關鍵字：原發性眼眶肉芽腫炎症，眼眶間接腫瘤炎症，淚腺。