Sarcoidosis With Orbital Tumor Outside the Lacrimal Gland

Initial Manifestation in 2 Elderly White Women

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Two elderly white women (aged 72 and 87 years) were first seen with painless, unilateral orbital swelling. Orbital scanning revealed masses infiltrating the soft tissue around the eye. Biopsy results showed nodular, noncaseating granulomas consistent with sarcoidosis. One patient’s workup revealed systemic manifestations of sarcoidosis at the time of examination with hilar lymphadenopathy noted on gallium scan; the other refused a complete systemic workup. The orbital tumors resolved with systemic prednisone therapy. To our knowledge, our 87-year-old patient is the oldest to be seen with orbital sarcoidosis. These 2 patients demonstrate that this diagnosis must be considered with orbital tumors in the elderly and in unusual locations, such as these which occurred outside the lacrimal gland.

REPORT OF CASES

CASE 1

A 72-year-old white woman was first seen with a 1-month history of acute, painless, right upper eyelid swelling (Figure 1). External examination revealed 2-mm right-sided exophthalmos and a firm, nontender, palpable mass over the right superior orbital rim. Visual acuity was 20/40 OD and 20/30 OS. Findings from slitlamp examination revealed a right-sided posterior subcapsular cataract. A moderate limitation of elevation and adduction in the right eye was also present.

Magnetic resonance imaging revealed an infiltrating mass in the right upper portion of the orbit, encompassing the globe superotemporally and tracking into the periorbital tissues and parotid gland (Figure 2). Findings from biopsy were consistent with sarcoidosis. The results of a gallium scan that was part of a systemic workup showed evidence of hilar adenopathy.

The patient’s symptoms essentially resolved after a 4-month course of oral prednisone in a tapering dose. There was a small amount of residual right upper eyelid ptosis. The palpable orbital mass was no longer present.

CASE 2

An 87-year-old white woman was first seen with a 1-month history of decreased visual acuity in both eyes and a 3-week history of left upper eyelid ptosis (Figure 3). On examination, her visual acuity was 20/25+/-3 OD and 20/30+2 OS. External examination revealed a slightly tender, palpable mass at the medial aspect of the left
superior orbital rim accompanied by left upper eyelid ptosis.

Results of blood tests, chest radiography, mammography, and head computed tomography showed no evidence of sarcoidosis. An orbital computed tomographic scan showed a soft tissue prominence in the postseptal space of the left orbit (Figure 4). Biopsy findings were consistent with sarcoidosis.

The patient's visual acuity improved following 2 months of treatment with tapering oral prednisone. The superior orbital mass resolved markedly. She refused any further workup of systemic sarcoidosis.

RESULTS

The first patient's biopsy results showed a white, fibrous mass with a soft consistency, measuring $7 \times 4 \times 3$ mm. The second patient's biopsy specimen showed 3 masses of firm pink to yellow-tan orbital tissue, the largest measuring $9 \times 5 \times 3$ mm. Both microscopic examinations revealed multiple discrete nodules of granulomatous inflammation comprised of epithelioid cells and Langhans'-type multinucleated giant cells (Figure 5 and Figure 6). Lymphocytes and occasional plasma cells surrounded the nodules. No sign of caseation or malignancy was found. Findings from acid-fast and Gomori methenamine silver stains were negative. Both specimens were consistent with sarcoidosis.

COMMENT

Uveitis, conjunctivitis, central nervous system involvement, lacrimal gland involvement, and skin manifestations of the eyelid are common ocular findings. Approximately half of all ocular sarcoid involvement occurs in patients who were not previously given the diagnosis and leads to findings diagnostic of systemic sarcoidosis.³ Orbital sarcoidosis in particular may have a strong correlation with systemic sarcoidosis. Collison et al² reported 15 cases that were seen initially with orbital sarcoidosis, of which 14 were subsequently demonstrated to have systemic sarcoidosis.

Orbital sarcoidosis presenting as a tumor outside the lacrimal gland is rare in all age populations. In a series of 2000 cases of sarcoidosis, only 2 had orbital involvement outside the lacrimal gland.³ Of 532 cases of sarcoidosis, Obendauf et al¹ noted only 2 have entailed an orbital mass outside the lacrimal gland producing exophthalmos.

Sarcoidosis usually is seen in the second and third decades of life and most commonly in blacks. In 23 cases of orbital sarcoidosis, Henderson¹ described the mean age of the patients to be 57 years. To the best of our knowledge, there has only been 1 case reported in the English-language literature in which orbital sarcoidosis was seen as an initial manifestation of systemic sarcoidosis in a person older than 70 years. Orbital sarcoid occurs predominantly in women older than age 50.
Faller et al described a 73-year-old woman with biopsy-proven orbital sarcoidosis and a systemic workup indicative of this condition. Benedict reported 1 case of isolated orbital sarcoidosis in a 72-year-old woman, but results of a search for systemic sarcoidosis were normal.

To the best of our knowledge, our 87-year-old patient is the oldest in the literature to be seen with orbital sarcoidosis. These 2 elderly patients emphasize that sarcoidosis must be considered in the differential diagnosis of orbital masses in all age groups, even when it occurs outside the lacrimal gland. A search for systemic findings should be undertaken if the orbital mass proves to have characteristics consistent with sarcoidosis, and appropriate therapy should be instituted.

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REFERENCES

A look at the past...

Trachoma

Hirschberg presented statistics upon the occurrence of the epidemic granule disease in Europe, America, and Australia. At 1 or 2 per cent, a locality may be considered free from trachoma, at 10 to 15 per cent moderately, and at 30 per cent, severely affected by the disease. Switzerland seems most fortunate. England and France have only rare cases, Belgium and some parts of Germany have very many. No part of Russia seems entirely free from the disease. Race-predisposition does not appear to exist, but it is a fact that trachoma is a disease of the poor. Terrestrial conditions, particularly swampy lands, are of decided effect upon the occurrence and spread of the disease.