

## ISOLATED EXTRAOCULAR ORBITAL MASS: A RARE PRESENTATION OF SARCOIDOSIS

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**ABSTRACT.** We report a case of orbital sarcoidosis in a 66 year old male who presented with one month history of right eye swelling and intermittent diplopia. MRI revealed an enhancing infiltrative soft tissue mass in the inferior aspect of the right orbit and biopsy of the mass demonstrated non-necrotizing granulomas. Chest CT scan was normal and PET scan showed no other organ involvement. He was treated with tapering doses of prednisone over six months. Although relapse occurred while tapering prednisone to 20 mg per day, he responded well to the addition of azathioprine with complete resolution of visual difficulties and orbital the mass on repeat MRI. Sarcoidosis, presenting as an isolated orbital mass is rare, can be successfully treated and should be included in differential diagnosis. (*Sarcoidosis Vasc Diffuse Lung Dis* 2016; 33: 302-304)

**KEY WORDS:** orbital sarcoidosis, prednisone, anterior uveitis

### INTRODUCTION

Ocular sarcoidosis is second in frequency to pulmonary involvement. About 25% of the sarcoidosis patients have eye involvement with uveitis, the most common followed by lacrimal gland enlargement (1). Isolated extraocular muscle involvement is rare and most of these patients present with symptoms due to mass effect and inflammation such as pain and diplopia (3).

### CASE REPORT

A 66- year old Caucasian male presented with swelling of the right lower eyelid associated with

tearing, redness discomfort and double vision in the right eye. He had trouble with near vision. His past medical history included hyperlipidemia and prostate cancer. His family history was positive for lung cancer in both his parents. He never smoked. He drank about 1-2 beer per week and did not use illicit drugs. Home medications were acetaminophen and simvastatin. Physical examination revealed erythematous rash on right forehead, palpable swelling under right lower eye lid, tearing and conjunctival injection in the right eye. He had a CT scan followed by MRI of right orbit showing enhancing infiltrative process involving inferior orbital fat of right orbit with involvement of inferior rectus muscle and extension to the medial rectus muscle. Biopsy of the lesion showed non-necrotizing granulomas. Special stains and cultures were negative for fungus and AFB. His PPD was negative. Thyroid function tests were within normal limit and antithyroid antibodies were not detected. Whole body PET scan showed increased uptake only in the right inferior orbital mass (Figure 3). Chest CT scan was normal. Prednisone 60 mg PO daily was prescribed. In four

Received: 13 November 2014

Accepted after revision: 29 September 2015

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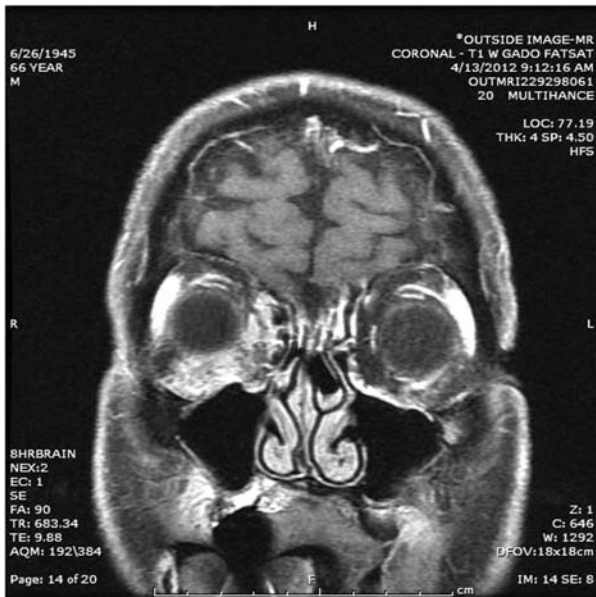


Fig. 1. MRI with contrast (coronal image) showing enhancing infiltrative soft tissue involving rectus muscles in the inferior aspect of the right orbit



Fig. 2. MRI with contrast (coronal image) showing almost complete resolution of enhancing lesion in the inferior aspect of right orbit after treatment

weeks there was resolution of his eyelid swelling, eye redness, discharge and improvement in his vision. While tapering his prednisone dose to 20 mg daily, he again complained of diplopia. He was started on azathioprine 150 daily and prednisone was increased

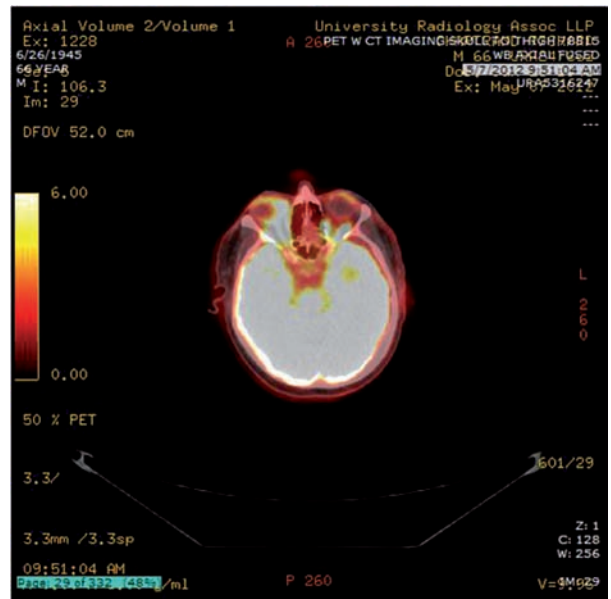


Fig. 3. PET scan showing increased uptake in the mass involving antero-inferior quadrant of right orbit.

to 60 mg daily which again improved his diplopia. Repeat MRI in 3 months showed marked interval decrease in a poorly defined mass in the inferior and medial aspect of the right orbit (figure 2). Prednisone was then slowly tapered to 15 mg daily together with continuation of Azathioprine 150 mg/day. He remained symptom free since then. After 2 years of treatment with Azathioprine and Prednisone, repeat MRI showed stable enhancing residual lesion in right orbit. Repeat PET scan showed no metabolic uptake in that residual orbital lesion suggestive if scar tissue. Prednisone was then slowly tapered off and Azathioprine was continued for another 3 months. Repeat MRI again showed stable residual lesion in right orbit. Azathioprine was then stopped. 6 months after stopping Azathioprine he remained symptoms free, unremarkable ophthalmologic examination. Repeat MRI showed stable residual lesion in right orbit.

## DISCUSSION

Ocular involvement in sarcoidosis is frequent. Although anterior uveitis is the most common manifestation which occurs in up to 85% of patients with ocular involvement, posterior uveitis is the most common cause of visual loss (1). Other manifesta-

tions are less frequently encountered and include lacrimal gland infiltration; optic nerve, chiasmatic, and/or sheath involvement; retrobulbar masses; exophthalmos; proptosis; eyelid swelling and palpable lid masses; optic radiation (meningovascular) infiltration; and bone destruction (2).

Extraocular muscle involvement is rare and reported in very few cases. In one study, orbital and adnexal involvement included the lacrimal gland in 63%, the eyelid in 17%, and the orbit in 13% of the patients (2). In a study by Prabhakaran and his colleague (1), among orbital lesions, the anteroinferior quadrant was the most commonly involved (in 6 out of 7 cases), the same site was involved in our case. In 1986, Collison, Miller and Gren (4) reported 15 patients with biopsy-proven orbital sarcoidosis involving the lacrimal gland, orbital fat and connective tissue, or both. Of the 12 patients, not previously known to have sarcoidosis, upon evaluation, systemic evidence of the disease was found in 11 patients. Only one patient, with isolated unilateral lacrimal gland involvement had no evidence of systemic sarcoidosis. As demonstrated in our patient, sarcoidosis rarely may present only with isolated orbital involvement. There are only three cases of orbital sarcoidosis limited to extraocular muscle described in the literature (5). In a review of orbital masses by Benedict (6), only 2 out of 1000 were due to sarcoidosis. Our patient did not have other systemic manifestation of sarcoidosis and diagnosis was based on biopsy report and exclusion of other granulomatous diseases. In our case the negative PET scan makes other organ involvement unlikely. It is not clear whether PET scan was used in previously reported case series of localized orbital sarcoidosis. In this regard our case report is unique. Systemic steroid is the treatment of choice even for orbital sarcoidosis. Intralesional steroid injection can be considered for localized orbital disease. Agents like azathioprine, methotrexate or infliximab can be added particularly if patients require higher doses of prednisone to keep the inflammation

under control. Initially our patient did respond to prednisone alone but had a relapse while tapering prednisone and required addition of Azathioprine. About 95% of the orbital sarcoidosis patients, who are treated with systemic steroid, show complete or partial response (1, 2). In one case series, during 5 year follow up, systemic sarcoidosis developed in 8% of the patients with only orbital and adnexal sarcoidosis at presentation(2). Our patient has been in remission for the last two years with no evidence of other organs involved.

## CONCLUSION

Orbital sarcoidosis presenting as extraocular muscle involvement is rare. Sarcoidosis should be considered in the differential diagnosis of orbital lesions and new onset eye symptoms. Biopsy is required to confirm the diagnosis and rule out other etiologies. PET scan is useful in ruling out other organ involvement. Although systemic steroid is the treatment of choice, cytotoxic agent may be needed to control the inflammation. Even after the disease has gone into remission with treatment, the patient needs to be followed for long term since active systemic disease can develop months to years later.

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