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Original Article

# Histiocytic lesions of the orbit: A study of 9 cases

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## Abstract

*Purpose:* To describe the clinical presentation, treatment, and outcome of patients with histiocytic lesions of the orbit.
*Methods:* Retrospective study of 9 patients treated and followed up between October 2001 and January 2018.
*Results:* Eight patients in our series were males and one patient was female. The mean age at presentation was 16.8 years (range, 1 to 42 years). All patients had unilateral disease. The most common presenting complaint was upper eyelid swelling in 8 of 9. All patients underwent preoperative computed tomography (CT) and magnetic resonance imaging (MRI). Eight of 9 patients demonstrated orbital bone erosion with adjacent soft tissue mass. Destruction of the orbital roof and contrast enhancement of dura were detected in 3 cases. All cases underwent orbitotomy and subtotal tumor excision with additional bone curettage (4 cases) and intraorbital steroid (40 mg triamcinolone acetonide) injection (3 cases). Adjuvant systemic chemotherapy consisting of vinblastine and prednisone was administered in 3 cases with dural involvement. External radiotherapy (1000 cGy) was applied in one case because of widespread disease. Histopathologic diagnoses were eosinophilic granuloma (7 cases), necrotic xanthogranuloma (1 case), and Langerhans cell sarcoma (1 case). The mean follow-up period after diagnosis was 19.7 months (range, 1–96 months). There was no systemic or multifocal bone involvement in eosinophilic granuloma cases at initial presentation and follow-up. None of these patients developed diabetes insipidus or neurologic symptoms. The patient with Langerhans cell sarcoma died from systemic disease 1 month after diagnosis of the orbital tumor. The patient with necrotic xanthogranuloma did not develop any malignancy at 9 months follow-up.
*Conclusions:* Eosinophilic granuloma was the most frequently encountered orbital histiocytic lesion in our series. Eosiophilic granuloma usually responded well to subtotal tumor excision, bone curettage, and intraorbital corticosteroid injections. Systemic chemotherapy was used in cases with full thickness bone destruction and adjacent dural enhancement in an effort to prevent the development of central nervous system disease.

**Keywords**: Eye, Orbit, Langerhans cell histiocytosis, Necrotic xanthogranuloma, Langerhans cell sarcoma, Eosinophilic granuloma, Intralesional steroids, Chemotherapy, External beam irradiation


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## Introduction

Histiocytic disorders are a group of diseases that occur when there is an over-production of white blood cells known as histiocytes that can lead to organ damage and tumor formation. Histiocytic disorders are made up of a wide variety of conditions that can affect both children and adults.[1,2] In 1987, the Histiocyte Society classified these disorders into three groups based on the types of histiocyte cells involved.[3] The first group is called a dendritic cell disorder, and the most common disease in this group is Langerhans cell histiocytosis (LCH). (www.histo.org) Also included in this dendritic cell group are more rare diseases of non-Langerhans cell histiocytosis including juvenile xanthogranuloma (JXG), necrotic xanthogranuloma (NXG), and Erdheim-Chester Disease (ECD). The second group is called a macrophage cell disorder, and includes primarily hemophagocytic lymphohistiocytosis (HLH) and Rosai-Dorfman Disease (RD). The third group is

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called malignant histiocytosis and includes certain kinds of leukemia and malignant tumors such as Langerhans cell sarcoma (LCS). (www.histo.org) In this report, the clinical features and treatment results of 9 orbital histiocytic lesions seen at a tertiary referral center are reported.

## Materials and methods

We retrospectively reviewed the clinical and histopathology records of orbital histiocytic lesions managed on the Ocular Oncology Service from October 2001 to January 2018. Histopathologically confirmed cases of orbital histiocytosis were included. Institutional ethics committee approval was obtained and informed consent was available for all cases.

Medical records were analyzed for age at presentation, gender, laterality, symptoms, duration of symptoms, clinical features, radiological features, treatment methods, histopathological diagnosis, and outcome. Computed tomography (CT) and magnetic resonance (MR) images of the orbit were reviewed. All cases underwent anterior orbitotomy to obtain tissue diagnosis. The tumor was debulked with bone curettage and intralesional steroid triamcinolone acetonide (40 mg/ml) injection as necessary. In cases with full thickness destruction of the upper orbital wall and adjacent dural enhancement on MRI, systemic chemotherapy consisting of vinblastine and prednisone was given to prevent central nervous system (CNS) disease. Cases with extensive disease or those in which repeat orbital imaging failed to disclose any resolution were considered for low-dose (1000 cGy) orbital external beam radiotherapy (EBRT). All cases underwent systemic work-up including, complete blood count, chest radiograph, abdominal ultrasound, ultrasonography, and bone scan at initial diagnosis. Repeat systemic evaluation was done by the pediatric or medical oncologist as necessary during follow-up.

## Results

A total of 9 patients were included. Patient demographics, clinical features, treatment results, and follow-up are depicted in Table. Eight patients were males and one was female. The mean age at presentation was 19.7 months (range, 1–96 months). All patients had unilateral disease with the right orbit being involved in 6 and left orbit in 3 patients. The presenting complaints included swelling in the upper eyelid (n = 8) (Fig. 1a), proptosis (n = 1), and redness of the upper eyelid (n = 1). The mean duration of symptoms was 6 weeks (median, 3 weeks; range 2–20 weeks). There was no history of trauma, systemic illness, or neurological symptoms in any of the cases. At presentation, inferior globe displacement was seen in 3 cases. A palpable mass lesion was documented in the superior orbit in 2 cases. There was no regional lymphadenopathy. Differential diagnosis included dermoid cyst, rhabdomyosarcoma, metastatic neuroblastoma, and lacrimal gland malignant epithelial neoplasm.

Computed tomography (CT) and/or magnetic resonance imaging (MRI) was done in each case and revealed a superiorly located well defined heterogenous mass with bony erosion involving frontal bone in 6 patients, sphenoid in 1 patient, and frontal and ethmoid in 1 patient (Figs. 1b, c, 2a). Destruction of the orbital roof and adjacent dural enhancement on MRI was seen in 3 patients (Figs. 1b, 1c). The patient with NXG presented with an indurated hard nodule on the upper eyelid and CT showed anterior orbital involvement as well. There was no bone involvement in this patient. The patient with LCS had a diffuse orbital tumor with an epicenter in the inferior orbit and there was also no orbital bone involvement in this patient as well.

All patients underwent anterior orbitotomy via skin approach. An extraperiosteal approach was preferred but this was not possible in many cases with bone destruction. Four cases received bone curettage because of the remaining irregular protruding bone fragments found during orbitotomy. Three cases were given intralesional steroid in the same sitting after debulking the tumor mass. No postoperative complications were noted. Complete resolution of symptoms was seen in all cases except for one case with extensive disease which was treated using external beam radiotherapy (1000 cGy). Three patients having destruction of the orbital roof with enhancement of adjacent dura on MRI were given systemic chemotherapy consisting of vinblastine and prednisone. These 3 patients were followed up for a mean of 20.3 (range 3–54) months. After orbitotomy and tumor biopsy, the patient with NXG underwent systemic evaluation but was negative for paraproteinemia and other hematologic abnormalities. The patient did not accept further treatment and was lost to follow-up after 9 months.

Histopathologic diagnoses were eosinophilic granuloma (7 cases), necrotic xanthogranuloma (1 case) and Langerhans cell sarcoma (1 case). Systemic workup was negative in 8 cases except for the case with Langerhans cell sarcoma. There was no evidence for multifocal involvement in LCH cases (Fig. 2b). The patient with Langerhans cell sarcoma died from systemic disease 1 month after ocular diagnosis and almost 1.5 years after initial diagnosis despite intensive systemic chemotherapy. The mean follow-up period after surgery was 19.7 months (range, 1–96 months) (Figs. 1d, 2c). The final mean visual acuity was 0.007 logMAR (0.00–0.05). All patients or their families were contacted by phone at the time of the writing of this manuscript. Their eye and systemic status were updated and they were asked to send in any pertinent medical documents relating to their condition. The mean follow-up from initial diagnosis to the last phone contact was 103.2 (range: 33–194) months. None of the patients had local recurrence nor developed diabetes insipidus or neurologic symptoms during follow-up.

Gross examination of the excised material showed yellowish-white mass with necrotic appearance in all cases. Histopathologic examination revealed Langerhans cells, eosinophils, and histiocytes with multinucleated giant cells composing a polymorphous granulomatous inflammatory reaction. There were also neutrophils and lymphocytes. The Langerhans cells stained positive with CD1a and S-100 protein immunohistochemically while the histiocytes were CD68 positive. In the case with necrotic xanthogranuloma, histopathological examination showed foamy histiocytes palisading around areas of necrobiotic collagen, fibrosis, Touton giant cells, and lymphocytes. The lipid containing cells did not stain with S-100. The case with Langerhans cell sarcoma exhibited additional cytological markers for malignancy including atypia, nuclear grooving, hyperchromatic nuclei, and prominent nucleoli.

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