Histiocytosis is a family of disorders with varied clinical presentation. Orbital involvement is rare and the diagnosis can often be challenging. Orbital histiocytic disorders are classified into Langerhans’ cell histiocytosis (histiocytosis X); sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman syndrome); juvenile xanthogranuloma; Erdheim-Chester disease; necrobiotic xanthogranuloma (NXG). Herein, we present a retrospective analysis of orbital histiocytic lesions, with an emphasis on their clinical, histopathologic and immunohistochemical profile.

MATERIALS AND METHODS
A retrospective review of medical records of patients with histopathologically-proven orbital histiocytic lesions that were diagnosed between 2003-2013 was undertaken. Clinical characteristics such as patient’s age, gender, presenting symptoms, duration of symptoms and the site of involvement were noted. Formalin-fixed paraffin embedded sections of the biopsy specimen were reviewed for histopathologic findings on light microscopy and immunohistochemical stains including CD1a, S100 protein and CD68.

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Source</th>
<th>Dilution</th>
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<tbody>
<tr>
<td>CD1a</td>
<td>Dako</td>
<td>1:100</td>
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<tr>
<td>CD68</td>
<td>Thermoscientific</td>
<td>1:50</td>
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<tr>
<td>S100</td>
<td>Diagnostic Biosystems</td>
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RESULTS
Eleven cases of orbital histiocytosis were identified. The mean age at presentation was 21 years (Range, 2-84 years). Of these, 6 (54.6%) were children. There were 6 females and 5 males. The mean duration of symptoms was 9
months (Range, 1-24 months). Abaxial proptosis was the most common clinical presentation (9/11, 80.8%). The superotemporal orbit was the commonest location in 54.6 % cases (6/11). Bilateral orbital involvement was seen in 4 patients (36.4%). Systemic lymphadenopathy was noted in 4 cases (36.4%).

Histopathologically, 5 cases (45.5%) were diagnosed as Langerhans’ cell histiocytosis (LCH), 5 cases (45.5%) as Rosai-Dorfman disease (RDD), and 1 case (9%) as Erdheim-Chester disease (ECD). All RDD specimens depicted sheets of histiocytes with vesicular nuclei and abundant cytoplasm. Emperipolesis (erythrophagocytosis) was identified in some of these cells. CD-68 immunostains were diffusely positive in 4 out of 5 RDD cases, and focally positive in one case. S-100 was focally positive in all 5 cases.

LCH cases revealed numerous monomorphic Langerhans’ cells with a grooved nuclei and variable number of eosinophils, lymphocytes and neutrophils in the background. Immunohistochemistry showed strong CD1a and S-100 positivity in all LCH cases. In ECD, there was diffuse infiltration by lipid-laden macrophages, inflammatory cells including lymphocytes and macrophages, and multinucleated giant cells of the Touton type which showed CD-68 positivity.

**DISCUSSION**

This study describes the spectrum of orbital histiocytic lesions which were diagnosed at a tertiary care centre over a 10 year period. Langerhans cell histiocytosis (LCH) is an uncommon disorder of the dendritic Langerhans’ cell that lacks histologic evidence of malignancy but behaves in an aggressive manner. Involvement of the orbit by LCH is uncommon and accounts for less than 1% of all orbital tumors. The average age of onset is 1 to 3 years, and the disease occurs more commonly in males. The presenting symptoms may vary from proptosis to dislocation of the globe. A definitive diagnosis requires that lesional cells exhibit positive staining with S-100 and CD1a on immunohistochemistry.

Rosai-Dorfman disease (RDD) is most frequently seen in children and young adults. Painless lymphadenopathy with fever, night sweats and weight loss is the most frequent systemic presenting symptom and involves the cervical region in upto 90% of patients. The presence of emperipolesis, or the engulfment of lymphocytes and erythrocytes by histiocytes that express S-100, is considered diagnostic of RDD. Apart from S-100 antigen positivity, immunohistochemical stains of RDD cells are also positive for CD68, CD163, α1-antichymotrypsin, α1-antitrypsin, fascin and HAM-56, while CD1a is typically negative.

Erdheim-Chester disease (ECD) is a very rare, nonfamilial, disseminated, non-Langerhans cell form of histiocytosis that is characterized by multisystem involvement. The classical ophthalmological manifestations of Erdheim-
Chester disease are bilateral xanthelasma and proptosis. It usually affects middle-aged and elderly patients and rarely affects children. The single case of ECD in our series was an 84 year old male presenting with bilateral intra and extraconal mass lesions. Histopathology showed lipid-laden macrophages, inflammatory cells and giant cells of the Touton type, with only CD-68 positivity.

**Conclusion**

The clinicopathologic spectrum of orbital histiocytic lesions revealed Langerhans cell histiocytosis and Rosai-Dorfman disease to be equally common. For establishing the exact category of orbital histiocytosis, a knowledge of clinical features, characteristic light microscopic findings and immunohistochemical study is the key. A strong clinical suspicion backed by close co-ordination with the pathologist is necessary for accurate diagnosis and appropriate management of orbital histiocytic lesions.

**REFERENCES**