Abstract
Fetal rhabdomyoma is a rare neoplasm composed of immature striated muscle cells that resemble rhabdomesenchymal cells during weeks 6-10 of embryogenesis. This tumor, though well-described in many anatomic locations, is most commonly found in the head and neck region. We report a case which highlights the difficulty in diagnosing a fetal rhabdomyoma in the lacrimal gland of a 56 year old woman. This case report gives an overview of this rare neoplasm, detailing the presentation, diagnosis, and management.

Key words: rhabdomyoma, neoplasm, lacrimal gland

Introduction
Fetal rhabdomyomas are benign neoplasms derived from rhabdomyoblasts. They are characterized by immature striated muscle cells, resembling rhabdomesenchymal cells during weeks 6-10 of embryogenesis. Although the cardiac form is the most common presentation, on rare occasion rhabdomyomas can be found in the head and neck. These neoplasms present in all age groups; however, more than 50% affect patients under 15 years of age.2 We report a unique case outlining the challenges in diagnosing fetal rhabdomyoma within the lacrimal gland and provide an overview of the clinical presentation, histopathology, diagnosis, prognosis and treatment of this rare entity.

Case Report
A 56 year-old healthy female presented to the clinic with an 8-week history of intermittent right upper lid swelling and conjunctival injection with intermittent pain. Clinical examination revealed mild medial upper lid swelling and right 1mm exophthalmos with 3mm hypoglobus on Naugle exophthalmometry readings (Figure 1a). There were neither sensory nor ocular motor deficits. There was bilateral reduced tear production with Schirmer’s test with signs of ocular surface desiccation in both corneas, with an otherwise normal slit lamp dilated fundus examination. The orbital computed tomography showed a well-defined right homogeneous lesion in the supero-temporal extraconal space, corresponding to an enlarged lacrimal gland in close contact with the globe, enhanced with contrast, particularly in its periphery. There was no evidence of bone erosion or calcification (Figure 1b). An excisional biopsy was planned with the consideration that malignancy was suggested by the short, progressive history, unilateral involvement and age of the patient.

For surgical management, a lid crease was made, and the supero-temporal orbit was exposed. Using four 2-0 silk traction sutures and a Freer periosteal elevator, the soft tissues were carefully dissected, avoiding the levator palpebrae superioris and superior tarsal muscles until the periosteum of the right superior orbital rim was identified. Once incised and elevated, an enlarged lacrimal gland was encountered, and three quarters of the anterior orbital lobe were removed. Frozen sections revealed chronic dacryoadenitis with no evidence of malignancy; therefore, the remaining posterior portion of the orbital lobe and the entire palpebral lobe of the lacrimal gland were left intact.

Interestingly, histopathology revealed maturing skeletal muscle intermixed with smooth muscle with a population of lymphoplasmocytic cells. On staining, the cells expressed smooth muscle actin, muscle

Figure 1. Orbital computed tomography showing a well-defined right homogeneous lesion corresponding to an enlarged lacrimal gland (a,b). Pre-surgical photograph showing mild medial upper lid swelling with 3mm hypoglobus of right orbit (c).
Rhabdomyomas are rare benign neoplasms affecting the pediatric population.1,3,4,5,6 However, the specimen was also found to stain positive for smooth muscle with caldesmin which was interpreted as superior tarsal muscle. Based on the inflammatory findings, Sjogren’s syndrome was systemically investigated and ruled out. Four weeks post-operatively, the patient’s globe malposition returned to normal, her dry eye symptoms resolved, and her ocular muscle function remained intact. At her six month follow-up she showed no signs of recurrence. She will continue to be followed clinically on a yearly basis.

**Discussion**

Rhabdomyomas are rare benign neoplasms of striated muscle affecting mainly the pediatric population.1 Outside the heart, they are extremely rare accounting for only 2% of all striated muscle-derived neoplasms.3 Up to 90% of these extra cardiac rhabdomyomas have been described in the head and neck and are classically differentiated into adult, fetal and genital types based on location and histologic differentiation.2-4 Fetal rhabdomyomas, the least common of extra cardiac rhabdomyomas were first described by Dehner et al in 1972.1 Initially, fetal rhabdomyomas were described affecting boys less than 3 years old in the head and neck; however, they have since been described throughout the body in both children and adults.4 Although this neoplasm has been well-documented in over 12 anatomic locations within the head and neck, to the best of our knowledge, there have been no reports of a fetal rhabdomyoma in the orbit.

Currently, there are only two accepted classifications: fetal and adult.1-5 Histologically, fetal rhabdomyomas are described as oval or spindled cells, in maturation arrest that are irregularly arranged in bundles with or without cross striations.1,5 This is in contrast with the adult-form rhabdomyoma, which is a more mature-appearing fetal cell line with an increase in cellularity.4 Fetal rhabdomyomas stain positive for myoglobin, desmin, muscle-specific actin.1,3,4,5,6 Can also reveal positivity for S-100 and/or glial fibrillary acidic protein and vimentin and smooth muscle actin.1,3,4,5 The histology seen in this case reveals a maturing spindle cell population with positivity for myoglobin, smooth muscle actin, and muscle specific actin, consistent with a fetal rhabdomyoma according to the World Health Organization for Soft Tissue Tumors Classification.1,3,4,5,6 However, since the location in the lacrimal gland has never been described, one could argue that the presence of smooth muscle intermixed with striate muscle is a normal anatomical feature in the supero-lateral anterior orbit. Interestingly, in this particular case, the dissection included not only the anterior aspect of the orbital component of the right lacrimal gland but also a substantial part of the deeper aspect of this lobe, where muscle fibers are supposedly not to be encountered.

When evaluating a supero-temporal mass, it is crucial to identify the duration of symptoms, presence of pain, and any associated motor or sensorial deficits in order to create the differential diagnosis. The differential includes inflammatory, infiltrative, neoplastic and structural processes. Inflammatory conditions classically include dacryoadenitis, which can present in a chronic, sub-acute or acute manner. Localized non-specific orbital inflammatory disease or autoimmune systemic conditions like Sjogren’s or infections should be considered on the inflammatory differential while cysts and dacryops as potential structural lesions. It is essential to keep in mind that these diagnoses often require differentiation through histopathology, since clinically they can look and present similarities. As for lacrimal gland neoplasms, they can be either epithelial or non-epithelial. Non-epithelial tumors, such as lymphoma, atypical lymphoid hyperplasia, and benign lymphoid hyperplasia have a better prognosis in general than epithelial tumors. Amongst the epithelial tumors of the lacrimal gland, pleomorphic adenoma is common and presents as a heterogeneous lesion on imaging. Since it has the potential for malignant transformation, complete excision of the gland is required when this diagnosis is suspected. More aggressive malignant forms are adenoid cystic and mucoepidermoid carcinomas with relatively poor prognoses. More rarely, rhabdomyosarcoma and extra-renal rhabdoid tumors can be encountered, which require aggressive treatment regiments.

Extra-renal rhabdoid tumors, though found in the lacrimal gland, are epithelial in origin, thus are easily differentiated from rhabdomyomas on histologic staining. On the other hand, rhabdomyosarcomas, can be differentiated by their prominent areas of necrosis, mitoses, and local infiltration.4,5

Following a histologic diagnosis of fetal rhabdomyoma, the clinician can be reassured that recurrence will not occur if the lesion is totally removed.4 With incomplete excision, recurrence is possible, but malignant transformation has never been described.4 In the presented case, we left in place only the most posterior aspect of the orbital lobe of the lacrimal gland when the frozen sections described a chronic inflammatory process. Weeks after, when the diagnosis of fetal rhabdomyoma was proposed and knowing the low risk of recurrence of these lesions, we opted to continue follow the patient annually and proceed with additional surgery only if there are any changes in the clinical and/or radiological examination. We believe that diagnosing a fetal rhabdomyoma in the lacrimal gland is a clinical challenge and that differentiation from other smooth muscle tumors, lacrimal gland malignancies, and inflammatory states is crucial for the adequate management and accurate prognosis.

**REFERENCES**