UNIVERSITY CHILDREN'S HOSPITAL ZURICH

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Tonic pupil – what's behind?

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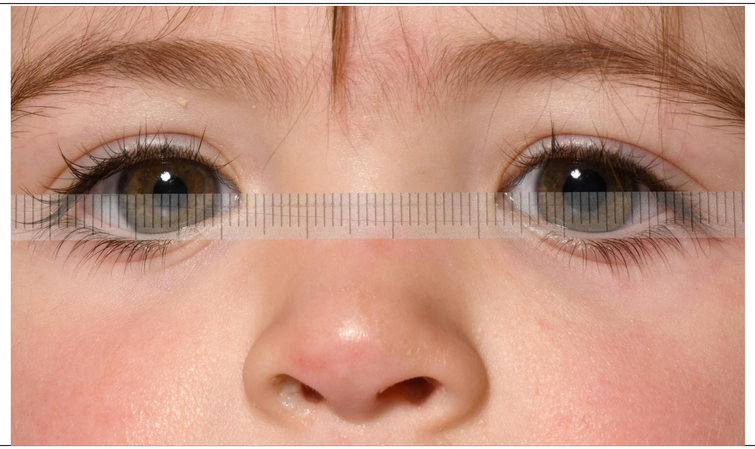
Introduction

Tonic pupils are characterized by

1. Poor response to light
2. Slightly better response to near stimuli
3. Slow redilation phase
4. A dysfunction of the ciliary ganglion

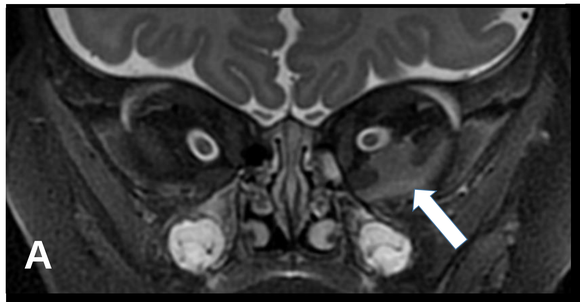
- Tonic pupils in infants are extremely rare.
- In two infants an orbital mass as the cause of the unilateral tonic pupil is documented in literature and a biopsy in both cases revealed a glial-neural hamartoma
- (Lambert S.R. et al. American J of Ophthalmology 2000. Goldstein S.M. et al. Journal of AAPOS 2002)

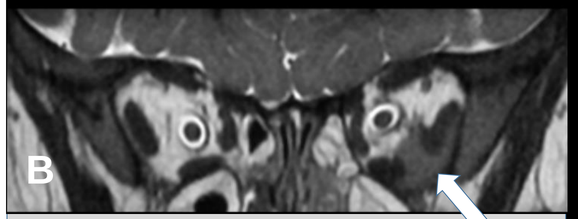


Case report

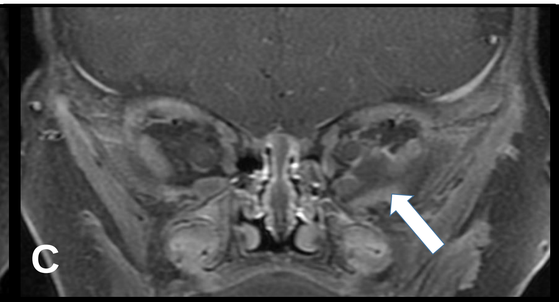
- 2 week-old female infant with anisocoria
- Left mydriasis with greater asymmetry in a brightly lit room
- Imperceptible reaction to light in the left eye, brisk constriction in the right eye
- Normal ocular motility and neurological examination
- 30 minutes after application of 0.1% pilocarpine in both eyes, her left constricted while the right did not, thus, confirming the diagnosis of a congenital tonic pupil on the left side
- Magnetic resonance imaging (MRI) revealed a soft–tissue mass in the inferolateral portion of the left orbit, interposed between the lateral and inferior rectus muscles.
- Repeated ultrasound examinations and MRI scans of the orbit, ophthalmological and neurological follow-up examinations have shown no changes over more than two years.

Imaging

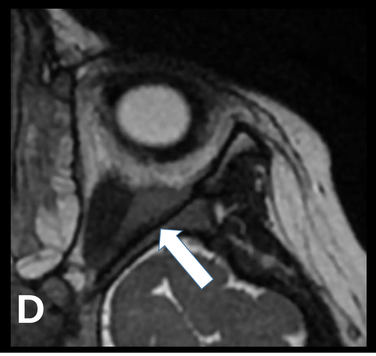




A (T2 fs cor, 2019) and **B** (T2 cor, 2022): left orbit with hyperintense soft tissue mass between the inferior and lateral rectus muscle

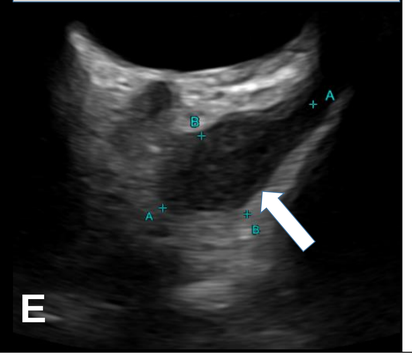


C (T1 fs cor after contrast): lack of contrast-enhancement



D (T2 axial) left orbit: mass lateral to inferior rectus muscle

E (Ultrasound of the left orbit): relatively homogeneous hypoechogenic lesion with sharp smooth demarcation



Conclusion

The clinical findings in this infant in combination with the inferolateral orbital mass without contrast-enhancement is suggestive for a glial-neural hamartoma. Hamartomas are abnormal collections of nonneoplastic cells. They are believed to arise as a result of abnormal migration and differentiation of cells during embryonic development. Several controls over more than two years showed stable findings and absence of growth in our patient.

Clinical examination without a biopsy might be appropriate in infants presenting with congenital unilateral tonic pupil and typical imaging findings consistent with glial-neural hamartoma. In addition, *ultrasound of the orbit is particularly suitable* as a non-invasive examination method, that does not require repeated sedations. Differential diagnoses as malignant rhabomyosarcoma of the orbit must be ruled out.