Author's response to reviews

Title: Isolated enophthalmos: an uncommon gateway to orbital tumors in pediatrics. 9 month-old female presenting with isolated enophthalmos as the unique sign of a metastatic orbital tumor: a case report.

Authors:

Sara Touhami (saratouhami@gmail.com)
Emmanuel Bui-Quoc (emmanuel.bui-quoc@rdb.aphp.fr)

Version: 5
Date: 26 August 2014

Author's response to reviews: see over
Isolated enophthalmos: an uncommon gateway to orbital tumors in pediatrics.

9 month-old female presenting with isolated enophthalmos as the unique sign of a metastatic orbital tumor: a case report.

Sara Touhami MD*, Emmanuel Bui Quoc MD-PhD1.

1Pediatric Ophthalmology Department, Robert Debré Hospital, 48 Boulevard Serurier, 75019 Paris, France.

Key words: Enophthalmos, Exophthalmos, Proptosis, Infantile orbital tumors, Neuroblastoma, Pediatric tumors.

Authorship contribution:
ST conceived the study, and participated in its design. ST Drafted and revised the manuscript for content including medical writing for content, analysis and interpretation of data.
EB conceived the study, participated in its design and revised the manuscript.
All authors read and approved the final manuscript.

Financial support: None of the authors have any financial interests to disclose.

Conflict of interest: No conflicting relationship exists for any author.

Consent: Informed consent of the parents was obtained for the publication of this case report.
Corresponding author:

Dr. Sara Touhami
Pediatric Ophthalmology Department, Robert Debré Hospital, 48 Boulevard Serurier 75019, Paris
Tel: +33140035763
Fax: +33140032432
Email: saratouhami@gmail.com

Article type: Case report
Criterion: Unexpected or unusual presentations of a disease
Malignant orbital tumors are a rare and hazardous entity in both adult and paediatric populations. Histological origins are various and they are different in adults and children. Knowing the prognosis of malignancy at such location (near the central nervous system and important vascular features), clinicians are expected to prioritize their early diagnosis. Exophthalmos is the most commonly known symptom of such tumors especially in children where the neoplastic tissue tends to grow outwards given the narrowness of their bony structures. However, clinicians should not neglect that other signs can reveal these neoplasms and should remain cautious before the least symptom. As such, enophthalmos has already been described as a rare symptom of orbital tumors in adults in a few case reports. However it was generally the satellite of other preponderant accompanying signs that one usually cannot miss (mass, erythema, edema…) and more importantly it has never been reported as the unique indicator of an orbital neoplasm in paediatrics. The importance of early diagnosing these tumors is justified by their potentially disastrous prognosis and argues for the need of a perfect knowledge of their clinics, including the scarcest signs. Among these, enophthalmos is probably one of the least known. The case of this little girl alarmed us and made us choose to publish her story because the obvious lack of knowledge of this unusual association was brought to our attention at her expense. Her case deserves being reported because the diagnosis of her tumor was delayed by at least 3 months, which probably modified the prognosis, because of a lack of knowledge of this unusual association.

She in fact had been referred to 4 different paediatricians and pediatric
ophthalmologists at different teaching hospitals; however, all of them stated that her enophthalmos was nothing but a constitutional feature. The most important part of the story was that even the pediatric radiologists declined performing a CT scan because they believed that there was no valid justification for seeking an orbital tumor before enophthalmos alone. We report the case of a paediatric metastatic neuroblastoma revealed by enophthalmos alone and remind the importance of this sign as a revealing symptom of orbital tumors in children. The specificity of this case shows that enophthalmos can be the unique indicative sign of such hazard, which has not been clearly described elsewhere. The association between enophthalmos and orbital tumors is extremely rare in children and unfortunately not known in common practice of pediatrics. The authors believe it is extremely important to remind this statement to all practitioners because there can be no tolerance for ignoring this sign and delaying a potentially lethal diagnosis.
Revisions:

The manuscript was reviewed and edited by a native-English speaker with scientific expertise as suggested.

Consent was obtained from the parents/legal guardians of the patient.

All modifications were included in the manuscript file as suggested.