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Case Report

Visual Impairment Caused by an Intra-Orbital Dermoid Cyst in 9-Month-Old Infant

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Abstract

Introduction: Intra-orbital dermoid cyst usually presents with proptosis or rupture later in life due to the characteristic of slow and painless growth. Thus, visual impairment caused by intra-orbital lesion in infant is an exceptionally rare case.

Case Presentation: A 8-month-old female infant with swelling and ptosis of her left upper eyelid consulted our department after 3 months follow up at a local ophthalmology clinic. Ophthalmology examinations and imaging studies led to initial diagnosis of intra-orbital dermoid cyst. During the follow up, downward deviation of her left eye got worse followed by some findings indicating visual impairment. Soon after the excision of the orbital mass, her left visual function was restored.

Conclusion: Since the symptomatic presentations of intra-orbital dermoid cysts occur mostly in adulthood, there has been no reported case of visual impairment in an infant caused by an intra-orbital dermoid cyst thus far. We suggest that when we encounter a case with symptoms which might be caused by a rapid progressive growth of an intra-orbital space-occupying lesion, imaging studies need to be performed immediately.

Keywords: Amblyopia; Dermoid cyst; Infant; Intra-orbital; Visual impairment

Introduction

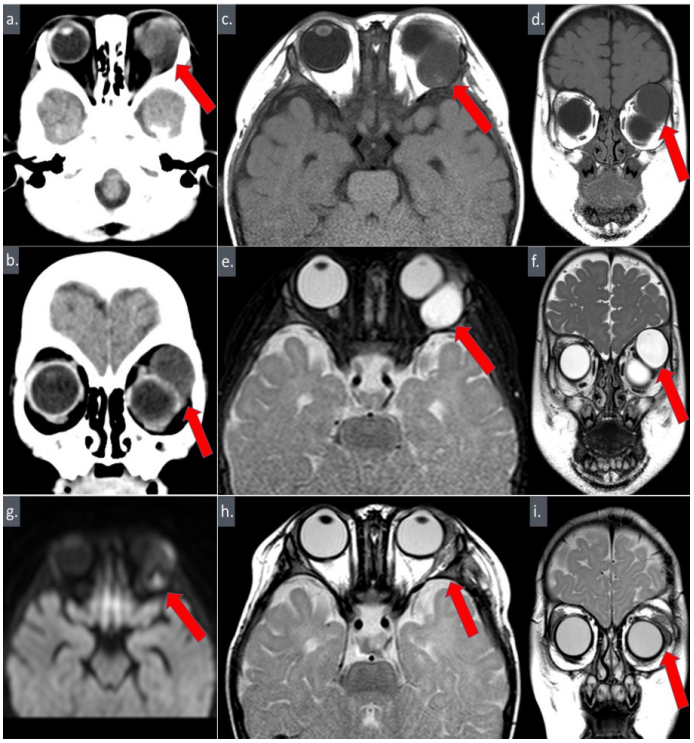
Dermoid cysts are benign cutaneous tumors that originate from aberrant primordial tissues. Many of them are evident in childhood and remain asymptomatic after growing slowly, in general. Although dermoid cysts can occur anywhere on the body, they frequently arise in the head and neck region. Among them, the most common location of the cysts are the orbital region (including periorbital and intra-orbital). Periorbital dermoid cysts are mostly superficial and often noticed early in life, on the other hand intra-orbital lesions are (deep dermoid cysts) usually present with proptosis or rupture later in life due to the characteristics of slow and painless growth. We report the case of an intra-orbital dermoid cyst that occurred in a female infant presenting visual impairment which was managed operatively with a rapid remission.

Case Report

A 4-month-old previously healthy girl presented to a local ophthalmology clinic due to the ptosis of her left eye lasting for about a month. After 3 months of monthly follow up, upper eyelid swelling occurred on her left eye in addition to the ptosis. Progressive enlargement of an intra-orbital tumorous lesion was suspected, and the infant's family consulted the ophthalmology outpatient clinic of Saitama Children Medical Center at the age of 8 months. An initial ophthalmic examination revealed that a soft tumorous lesion was palpable under the left upper eyelid and the left eye was already deviated to the inner lower position (Figure 1c). Also, ocular motility showed restriction of elevation and abduction in the left eye. However, fixation was maintained in both eyes. The cornea, anterior chamber, lens and fundus examination showed normal findings in both eyes.

Figures 1(a-d): a: One month old, b: Six months old, c: Eight months old (at the initial presentation) d: One week after the operation.

The patient was advised to undergo a Computed Tomography (CT) scan and a Magnetic Resonance Imaging (MRI) scan of the orbit. The axial and coronal CT scan of the orbit showed a hypodense, unilocular cystic lesion, including a hyperdense region, measuring 22×20×16mm in the extraconal space of the left superior orbit (Figures 2a,b). The cystic lesion was notably displacing the left eyeball inferiorly. Subsequently, an axial and coronal non-enhanced MRI scan of the orbit revealed a well-defined low intensity cystic lesion with an isointense region on a T1-weighted image and a high intensity cystic lesion with a hypointense cystic region on a T2-weighted image (Figures 2c-f). The cystic lesion was partially hyperintense on the diffusion weighted image (DWI) (Figure 2g). Radiographic evidence was suggestive of an enlarging deep dermoid cyst.

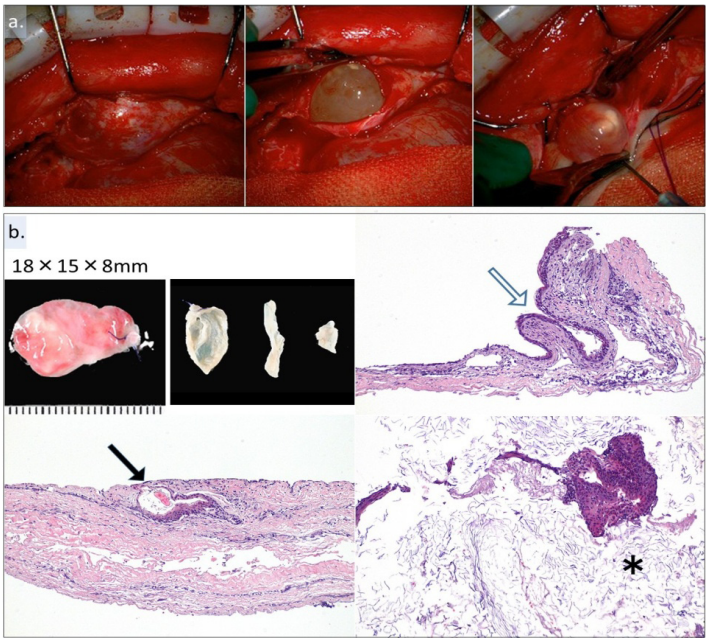


Figures 2(a-i): (a-g: preoperative, h-i: postoperative) a,b: CT scan, c,d: Non-enhanced MRI (T1-weighted image), e,f: Non-enhanced MRI (T2-weighted image), g: Non-enhanced MRI (diffusion weighted image), h,i: Postoperative MRI (T2-weighted image).

While a month had passed from the initial consultation, the condition took a turn for the worse. Fixation in the left eye was no longer present and the occlusion of her right eye showed an aversion reaction. It was diagnosed as having a visual impairment of her left eye, accordingly. Since it was obvious that the visual impairment was due to the displacement caused by the orbital tumorous lesion, the decision was made to have it surgically removed as soon as possible under the direction of the neurosurgical department.

Patching of her right eye (the dominant eye), 5 hours per day, was started immediately since there was approximately a month to wait until the operation.

Excision of the orbital mass was done through the subcranial approach. The operation revealed a cystic lesion covered by a capsule measuring 18×15×8mm containing a yellowish serous fluid (Figure 3a). The cyst was completely removed from the extraconal space of the left orbit with no disruption of the cyst wall. The cyst was sent for histopathological examination. Results showed the presence of hair follicles and keratinized tissues, covered in squamous epithelium, which was consistent with the diagnosis of dermoid cyst (Figure 3b).



Figures 3(a-b): a. Intra-operative photos; a cystic lesion covered by a capsule measuring 18×15×8mm with content of yellowish serous fluid, b. Histopathological examination. (upper left side) macro view of the dermoid cyst, (→) hair follicle, (→) squamous epithelium, (*) keratinized tissues.

Excision of the orbital mass displayed a good postoperative course. Ptosis and upper eyelid swelling of her left eye were no longer visible within a week after the operation. Deviation to the inner lower position evanesced completely as well (Figure 1d). Postoperative MRI revealed a complete removal of the cystic lesion and a properly located eye inside the left orbit (Figure 2h,i). The aversion reaction was no longer seen in her right eye and her left eye regained adequate fixation lasting time. However, elevation of her left eye was still restricted for about 16 months postoperatively.

Discussion

Dermoid cysts are benign developmental choriostomas formed by ectoderm and mesoderm that get trapped within the embryonic fusion lines. They are covered by squamous epithelium and may contain keratin and hair [1]. Approximately 7% of all dermoid cysts arise in the head and neck [2]. Among them, over 50% occur in or adjacent to the orbit [3,4]. However most of them are superficial and intra-orbit ones are rare. Also, Shepherd G. Pryor et al. reported that the pediatric dermoid cysts of the head and neck examined between 1980 and 2002 at Mayo clinic were 100% superficially located, in which all presenting signs were a palpable mass [3]. In contrast, intra-orbital dermoid cysts are clinically discovered later in life due to their slow painless growth ending up mostly in proptosis or rupture [5-7].

In our case, an intra-orbital dermoid cyst showed rapid progressive growth in an infant and presented ptosis, ocular deviation, ocular motility restriction and visual impairment. Since the symptomatic presentation of intra-orbital dermoid cysts occur mostly in adulthood, the presentation of such symptoms in an infant, as in this case, is clearly a rare exception. There has been no reported case of visual impairment in an infant caused by an intra-orbital dermoid cyst thus far. We reported this case as an announcement that intra-orbital dermoid cysts in infants could rarely show rapid growth and might cause visual impairment leading to amblyopia. It is known that complete surgical excision of dermoid cysts with an intact capsule could treat most of the symptoms caused, [3] in addition, our case showed a good therapeutic effect to the visual impairment. Imaging studies, such as CT scans and MRI scans, could reveal the characteristics of the cystic lesion and help in the preoperative diagnosis. Such studies will make it easier to carry out complete surgical excision as well. We must keep in mind to conduct imaging investigations as soon as we encounter a case with symptoms which might be caused by an intra-orbital, space-occupying lesion, not only dermoid cysts. If such cases are difficult to manage at small clinics, a quick decision has to be made to consult a general or university hospital.

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