A CASE REPORT OF ISOLATED INFANTILE HEMANGIOMA WITH MICROPHTHALMOS

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Abstract

Purpose: To report a rare case of infantile facial hemangioma with orbital extension associated with microphthalmos and high intraocular pressure of the left eye.

Case: A full term 1-day-old girl presented with left sided facial hemangiomas since birth.

Observations: Findings include large left-sided facial hemangiomas with orbital extension enveloping the left lateral rectus and optic nerve. Clinical examination showed no involvement of the upper and lower eyelid. Cycloplegic refraction showed anisometropia between the two eyes. The axial length of the left eye was found to be significantly shorter compared to the right eye, and intraocular pressure of the left eye was high.

Conclusions: Infantile facial hemangioma may be present with orbital extension without any eyelid involvement, and associated with microphthalmos of the left eye with no other systemic abnormalities.

Keywords: Infantile Hemangioma, Microphthalmos, Childhood Tumor, Glaucoma

***Key Messages*:**

- Patients with infantile facial hemangioma should undergo complete ophthalmological examination.
- Hemangiomas that cause significant morbidities such as visual impairment, cardiac failure, or bleeding tendencies, require treatment.

Background

Hemangiomas are the most common benign tumor of the orbit in infancy (1, 2). It originates from the proliferation of vascular tissue, involving anastomosis of vascular channels with infiltrative patterns (3). It has a predictable course of rapid enlargement with gradual spontaneous resolution later in life (1). Around 80% of all hemangiomas reach their final size by 3 months of age. The hemangioma may present as small isolated lesions or a large disfiguring mass with overlying ulceration that can cause visual impairment or other systemic manifestations such as airway obstruction or high output cardiac failure. Ocular complications of hemangiomas include amblyopia, refractive error, or optic nerve compression. Oral propranolol is now the first-line treatment, which should be administered early to avoid potential complications. Anophthalmia, microphthalmia and coloboma are likely to be caused by anomalies of pathways that control eye development resulting from primary genetic defects, acquired maternal infection or consequence of drug intake. The association between facial hemangioma and microphthalmos with the presence of other systemic abnormalities, such as posterior fossa anomalies, arterial anomalies, cardiac anomalies, and aortic coarctation, has been well-documented and could provide a clue to the diagnosis of PHACE syndrome (4). However, our case of infantile hemangioma with microphthalmos has no systemic abnormalities.

Case report

A 1-day-old, full-term female infant presented with left facial hemangioma noted since birth. Multiple left-sided facial hemangiomas were observed. Two hemangiomas were located at the lateral side of the left forehead measuring 3 cm x 2.5 cm and 2 cm x 2 cm, respectively. Another large hemangioma extended from the left lower cheek to the temple, measuring 6 cm x 4 cm. The eyelids were not involved (Figure 1a). Her eyes were orthophoric and she had full extraocular motility, bilaterally tested using a light target. No ptosis or proptosis were present, and no relative afferent pupillary defect (RAPD) was detected. Red reflex was good for both eyes. Oral propranolol 3.5 mg was commenced twice daily (at 2 mg/kg/day) starting on day 2 of life. The dosage was increased by 0.5 mg/kg/day every week up to 3 mg/kg/day. The child was under the supervision of the Paediatrics team for follow-ups and did not develop any side effects (such as hypotension, and bradycardia) from oral propranolol administration.

Neurological and cardiac examination were normal. Laboratory investigations for hematological disorders were also normal. Ultrasound of the cranium and abdomen were normal. Contrasted magnetic resonance imaging (MRI) of the brain, done under anaesthesia at 4-months old, showed left facial hemangioma extending to the left frontal, left orbital, left lateral extraconal, intraconal space enveloping the lateral rectus muscle, and left optic nerve. However, there were no orbital cyst detected and the cavernous sinus was not involved. No other brain abnormalities were detected (Figure 2a).

At 8-months-old, the follow-up revealed significant reduction in the size of the hemangioma. Two of the forehead hemangiomas were reduced to 3 cm x 2 cm and 2 cm x 1 cm, respectively, and the cheek hemangioma measured 5 cm x 4 cm (Figure 1b). All three hemangiomas were found to be smaller compared to the initial size. Examination under general anaesthesia revealed the cornea diameter of both eyes measured 10 mm vertically and 11 mm horizontally, with no other abnormality detected in the anterior segments of both eyes. Bilateral fundus examination showed pink and healthy optic disc, with cup-to-disc-ratio of 0.2, and normal retinal vasculature and macula. The intraocular pressure (IOP) in her right eye was 15.7 mmHg, while the left eye had higher IOP of 31.6 mmHg. No proptosis of the left eye was present. Axial length of the globe, measured with A-scan, was 19.65 mm for the right eye and 17.30 mm for the left eye. The cycloplegic refraction was +3.00 DS/-0.75 DC x 180 for the right eye and +6.00 DS for the left eye. Gonioscopic examination showed normal open-angle bilaterally with no abnormal vessels observed in the angle. She was started on topical timolol twice daily for the left eye. Additional serial MRI of the brain and orbit revealed further regression of the lesion (Figure 2b).

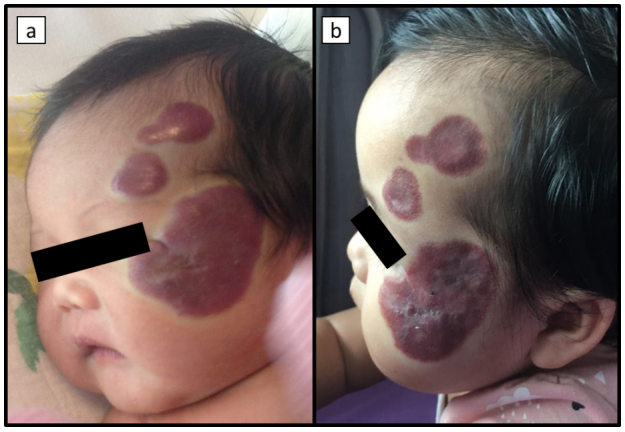

Figure 1: **(a)** Facial hemangioma at birth. **(b)** Facial hemangioma at 8-months old.

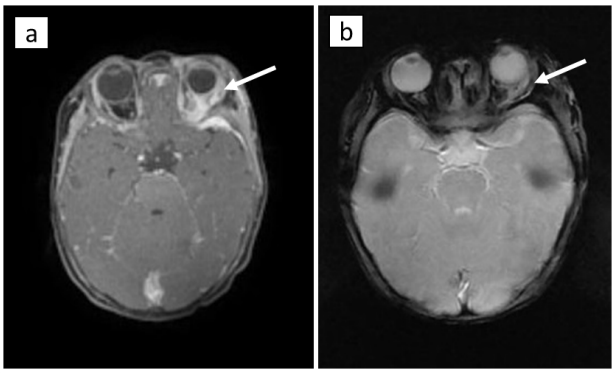

Figure 2: Magnetic Resonance Imaging (MRI) images. **(a)** Contrasted image of lesion extending to the left lateral extraconal and intraconal space enveloping the left lateral rectus muscle and optic nerve. **(b)** Non-contrasted image of the lesion

Subsequent examination under anaesthesia at 16-months old showed further reduction in the size of the left-sided forehead hemangiomas to 3 cm x 1.5 cm and 2 cm x 1 cm, respectively, and cheek facial hemangioma to 5.1 cm x 3.2 cm. The surface had become flatter and shrunken compared to prior presentation. IOP in the left eye showed 50% reduction from baseline to 16 mmHg. Topical timolol was then stopped. Serial MRI of the brain and orbit at 21-months old showed axial length of the globe at 20.10 mm for the right eye, and 17.60 mm for the left eye, which correlated with the axial lengths measured via A-scan (Figure 3).

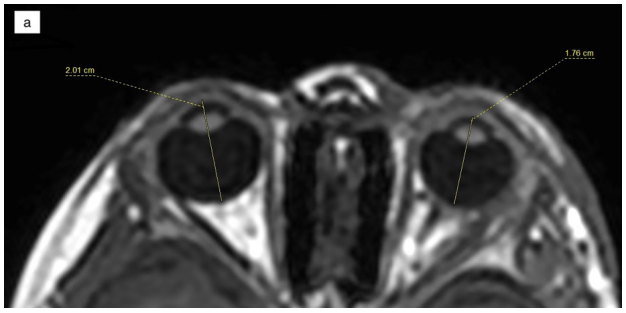

Figure 3: Axial length of orbit visualized via MRI at 21-months old.

Repeated cycloplegic refraction at 1-year follow up was +0.50 DS/-0.50 DC x 180 for the right eye and +5.00 DS for the left eye. Visual acuity assessment was hindered due to the child's lack of cooperation, necessitating the need for an examination under anaesthesia. Oral propranolol was discontinued since then. The diagnosis of left eye microphthalmos was made based on the axial length of the globe and refractive assessment. The child is under regular follow-up to monitor the IOP and refraction for glaucoma and amblyopia.

Discussion

Visual impairment in children with periorbital hemangiomas is commonly due to amblyopia, refractive error, strabismus and optic nerve compression. Isolated infantile facial hemangioma and microphthalmos with no other systemic anomalies in our patient is a rare finding. Large facial hemangiomas are commonly related to PHACE syndrome, which is a rare neurocutaneous disorder that is associated with structural anomalies of the brain, cerebral vasculature, eye, sternum and aorta (5). Clinical manifestation of the disorder includes posterior fossa malformations (P), large, segmental, plaque-type facial hemangiomas (H), arterial anomalies (A), cardiac defects (C) and eye abnormalities (E). The association of congenital glaucoma in patient with PHACE syndrome has been reported once previously (6). Our patient did not have any systemic neurocutaneous disorder related to PHACE syndrome.

Orbital hemangiomas are not rare. A study conducted by Tambe et al. reported that 17% of hemangiomas were orbital in location (7). Orbital hemangioma is frequently present with proptosis or globe displacement. Retro-orbital hemangiomas and proptosis have been reported to occur in patients with PHACE syndrome (8). Proptosis is a serious complication of orbital hemangiomas which can lead to corneal exposure keratopathy and disfigurement (9). Our patient, however, did not develop proptosis and globe displacement due to the smaller eyeball.

Microphthalmos is defined as an abnormally small eye or cornea with an axial length of < 16 mm at birth, and < 19 mm at 12 months of age, with corneal diameter < 10 mm at birth (10). Gordon and Donzis reported the mean axial length of children aged 1-2 years were 20.2 ± 0.3 mm (11). In our case, the axial length of the affected eye measured 17.60mm at 21-months old, with the contralateral eye measuring 20.10 mm (Figure 3).

Willshaw and Deady reported a case of facial hemangioma associated with unilateral microphthalmos (12). Fernandez et al. reported a case with an unusual combination of microphthalmos, facial hemangioma and Dandy-Walker malformation, which is a posterior fossa malformation (13). Similarly, Nishimaki et al. reported a case of Dandy-Walker malformation associated with microphthalmia (14). In contrast, our patient did not have any brain abnormalities detected through MRI imaging.

High intraocular pressure is a rare complication of periocular hemangioma that warrants special attention and management. Persistently high intraocular pressure may lead to the development of glaucoma. High IOP in patients with periocular hemangioma could be due to multiple lesions involving the eyelids, conjunctiva and iris (15). However, the intermittent high IOP recorded in our patient could possibly be due to the involvement of the left intraorbital, left lateral rectus muscle, and left optic nerve. The intraocular pressure was normalised in our case with the use of topical timolol and systemic propranolol, most likely due to the regression of the hemangiomas.

Conclusion

Patients with facial hemangiomas should undergo a complete ophthalmologic examination to exclude any possible ocular involvement. It is crucial to monitor the patient in the first weeks of life. Complete ophthalmological assessment, which includes intraocular pressure measurement, ocular motility assessment, anterior segment, and fundus examination, is mandatory as evidenced in our case. Our patient had no signs of hemangioma on the eyelids or conjunctiva. However, MRI imaging detected the presence of an intraorbital hemangioma. Our patient also developed anisometropia and microphthalmos of the left eye. Patients should undergo serial orbit and brain MRI imaging, as well as tests for other systemic anomalies associated with PHACE syndrome.

Conflict of Interest

The authors declare that there is no conflict of interest.

Informed consent

A written informed consent for patient information and images to be published was provided by the legal guardian of the patient.

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