# Orbital Aspiration as Treatment of Microphthalmos with Orbital Cyst : A Case Report

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**Abstract**

A 6-month-old girl came to the hospital with swelling of the right lower eyelid, exophthalmos, chemosis and upward deviation of the eyeball-all of which had been present since birth. Iris, optic disc, and chorioretinal coloboma were also apparent.

Magnetic resonance imaging revealed a small globe with a large cystic lesion in the orbit of the right eye. Pre- and post-operative photographs and magnetic resonance imaging indicated a safe, simple single orbital aspiration as an alternative treatment for mild microphthalmos with an orbital cyst.

**Key word** : Orbital Aspiration, Treatment, Microphthalmos, Orbital Cyst

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Microphthalmos with a colobomatous orbital cyst is a rare congenital anomaly of the orbital contents due to defects in the closure of the embryonic fissure and invagination of the optic vesicle during embryogenesis. which results in varied clinical presentations[1,2]. In the past, most of these patients were treated by enucleation and cyst excision. This is a case report of microphthalmos with an orbital cyst treated satisfactorily with a single orbital aspiration.

## CASE REPORT

A 6-month-old girl came to the hospital with a history of swelling of her right lower lid since birth. She was a full-term, 3,290 g baby, delivered by normal labor to a 35-year-old $G_3P_3$ mother. The child was normal during the prenatal period and had normal development. Her parents and older siblings were healthy. Ocular examination revealed chemosis, ectropion of the lower eyelid, and upward deviation of her right eye (Fig. 1). In Fig. 1, exophthalmos

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could not be appreciated. However, this condition could be detected in the MRI study in Fig. 2. The iris and chorioretinal colobomas, involving the optic nerve head, were revealed by slit-lamp biomicroscopy and indirect ophthalmoscopy, respectively. The child usually resisted when her left eye was occluded. Non-ocular examination was normal.

Magnetic resonance imaging of the orbits revealed a small right globe with a large cystic lesion located posteroinferiorly, approximately 4 cm in diameter with a low signal in the T1-weighted and a high signal in the T2-weighted images, near the insertion of the optic nerve (Fig. 2). Her chromosome analysis showed 46,XX.

Orbital aspiration was performed under general anesthesia. Fifteen milliliters of clear yellowish fluid was aspirated by passing a 25-guage needle through the right lower eyelid into the orbit in the same fashion as performing the retrobulbar block procedure. Post-operatively, exophthalmos and chemosis were remedied, but the right eye developed esotropia (Fig. 3). Moreover, the child could not fix with her right eye and usually resisted when her left eye was occluded. Serial post-operative magnetic resonance imaging revealed that the orbital cyst had decreased 2 cm in diameter at 1 month and 0.5 cm at 2 years (Fig. 4). A relatively small-sized and mildly distorted right globe was also noted. Repeated aspiration has not been required.

## DISCUSSION

Microphthalmos with a cyst is a rare congenital anomaly but should be suspected in patients with a small or unrecognizable eye and an orbital cystic lesion detected during palpation or visualization[3]. The clinical presentations of the orbital cyst include swelling, with or without bluish discoloration, in the lower lid and an upwardly deviated microphthalmic eye, or a purulent discharge from the sinus associated with an absent or a small-sized eyeball. Most patients had very poor vision in the involved eye. The microphthalmic eye showed a spectrum of posterior segment abnormalities such as retinal disorganization, gliosis, choroidal, optic nerve and scleral coloboma[2,4,5]. This patient could not fix with her right eye and usually resisted when the left eye was occluded due to the markedly upward deviation and the posterior segment lesions of her right eye. Postoperatively, she still resisted the occlusion despite having better alignment.

Microphthalmos with an orbital cyst has been associated with balanced reciprocal translocation between the long arm of chromosome 3 and 5 [4], trisomy 13[6], Aicardi's syndrome[7], optic pit [8], and persistent hyperplastic primary vitreous[9]. However, most of these patients were sporadic[10-11]. Histological studies of the excised microphthalmic eye and the orbital lesion have shown markedly disorganized ocular tissue forming a tumor-like mass,

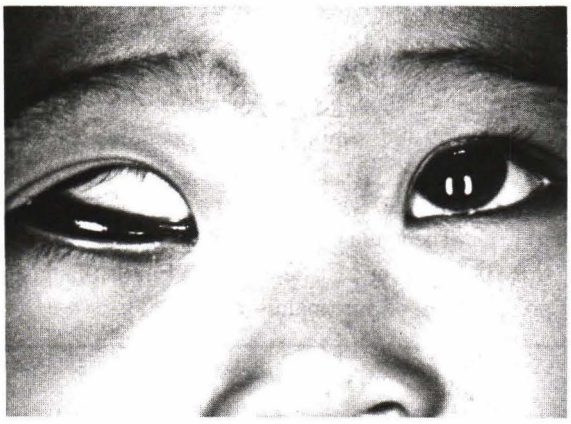

**Fig. 1. Chemosis, ectropion of lower eyelid, and upward deviation of the right eye.**

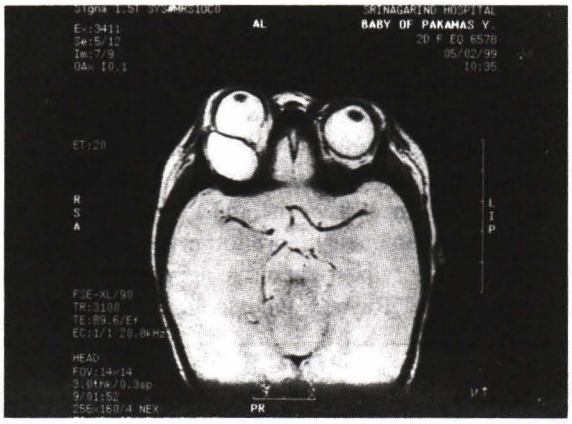


**Fig. 2. T2-weighted magnetic resonance imaging of the orbits showed a small right globe with a large cystic lesion located posteroinferiorly.**

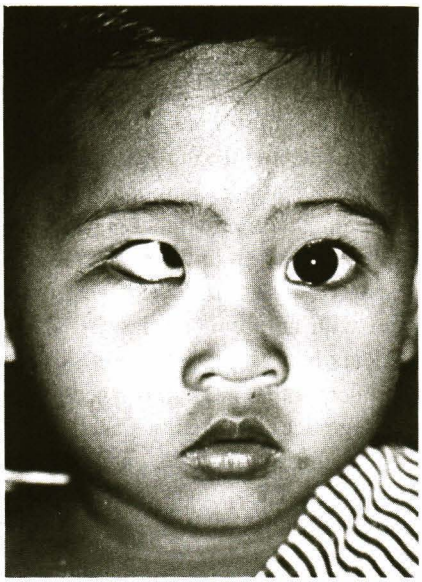

Fig. 3. Two years post-operatively, exophthalmos and chemosis were remedied but the right eye developed esotropia.

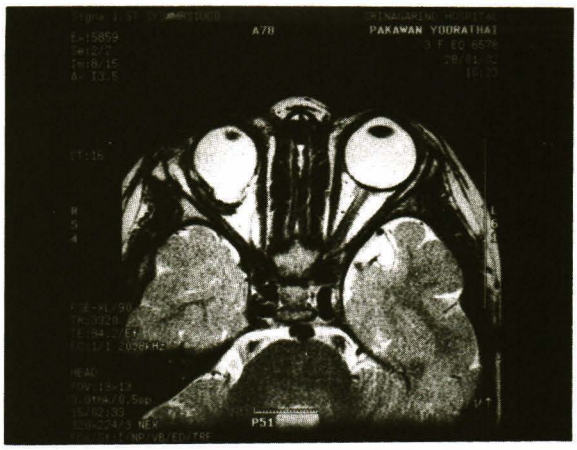

Fig. 4. T2-weighted magnetic resonance imaging of the orbits showed a small-sized orbital cyst at 2 years after orbital aspiration.

an area of discontinuation of the sclera that was suspected to be the defective closure of the embryonic cleft and a cyst lined by primitive, immature retinal tissue, which contained neuroglial elements and scattered dysplastic rosettes[4,12,13]. Lorenz et al[7] suggested that the cysts formed from an abnormal migration of neuroretinal tissue through the border of the optic disc coloboma that was also present.

The usual treatment for extreme cases is excision of the cyst alone or of the microphthalmic eye with the cyst and the fitting of a prosthesis[2]. Polito and Leccisotti[13] treated a case of colobomatous ocular cyst associated with a mild microphthalmos, by a cyst excision and pedicle ligature, *via* a transconjunctival orbitotomy, which resulted in a satisfactory post-operative appearance. Kodama et al[14] and Raynor and Hodgkins[15] proposed repeated aspiration with preservation of the eye as the new treatment technique.

This patient represented a good candidate for a safe, simple single orbital aspiration as the alternative treatment for mild microphthalmos with an orbital cyst. The advantage of an orbital aspiration over excision of the cyst alone or enucleation is reduction of both operative time and tissue destruction. Moreover, orbital aspiration maintains normal orbital and facial growth, which results in better appearance and simplifies post-operative care.

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# การเจาะผ่านเบ้าตาในการรักษาผู้ป่วย Microphthalmos ที่มีถุงน้ำในเบ้าตา : รายงานผู้ป่วย

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รายงานผู้ป่วยเด็กหญิงอายุ 6 เดือนที่มีหนังตาล่างข้างขวาบวม, ตาโปนและตาดำลอยขึ้นบนตั้งแต่กำเนิด ตรวจพบ coloboma ของม่านตา, ขั้วประสาทตา, มอรอยด์และจอประสาทตาร่วมด้วย ภาพถ่ายทางรังสีพบลูกตามีขนาดเล็กและถุงน้ำขนาดใหญ่ในเบ้าตา ผู้ป่วยได้รับการรักษาโดยการเจาะดูดน้ำออกจากถุงน้ำผ่านทางเบ้าตา ผลภาพถ่ายผู้ป่วยและภาพถ่ายทางรังสีหลังผ่าตัดสนับสนุนการเจาะผ่านเบ้าตาเป็นการรักษาที่ดีอีกวิธีหนึ่งในผู้ป่วย microphthalmos ที่มีถุงน้ำในเบ้าตา

**คำสำคัญ** : การเจาะผ่านเบ้าตา, การรักษา, microphthalmos, ถุงน้ำในเบ้าตา

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