

Skull Base Fracture or Abuse Suspicion Based on Intraorbital Lymphangioma Secondary to Periorbital Hematoma

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Intraorbital lymphangiomas are among the orbital tumors that can cause sudden eye protrusion in children. In children with periorbital hematoma (panda eye sign), a skull fracture or abuse is likely first considered as the differential diagnosis. A 7-month-old boy presented to the ophthalmologist with complaints of swelling of the right upper eyelid, subconjunctival hemorrhage on the right ear side, and periorbital subcutaneous hemorrhage, which had appeared since the morning of the day before the visit. The eyeball did not protrude. Based on the interview and clinical findings, right eyeball contusion was suspected. The patient was then followed up for observation. Later, during the physical examination, the abovementioned symptoms were noted. Hence, the patient was admitted for a close examination based on the suspicion of skull base fracture and abuse. Contrast-enhanced magnetic resonance imaging (MRI) after admission revealed a multifocal cystic structure within the right intraorbital muscular cone. Thus, he was diagnosed with right intraorbital lymphangioma. Intraorbital lymphangioma may not show ocular protrusion, and this disease should be considered in cases where abuse is suspected, considering the periorbital subcutaneous hemorrhage.

Key words: Abuse, Children, Intraorbital lymphangioma

INTRODUCTION

In recent years, the annual number of consultations regarding child abuse in Japan has been steadily increasing. Clinicians must suspect physical abuse in cases of periorbital subcutaneous hemorrhage (panda eye symptom) with an unclear mechanism of injury, such as bruising. If abuse is suspected in any case, careful medical care should be provided during the short examination time, with close observation of the relationship between the parents and the child. Alternatively, similar findings may be obtained in intraorbital tumors, such as intraorbital lymphangioma. In this report, we describe a case of intraorbital lymphangioma that was suspected to be caused by basilar hemorrhage and abuse due to the typical panda's eye symptom at the time of initial examination.

CASE

The mother of a 7-month-old male infant noticed sudden swelling of the child's right eyelid (Fig. 1), which led her to visit an ophthalmologist the next day. Subsequently, he first visited our department for an infant health checkup, and upon examination, a periorbital hematoma (panda eye sign) was observed (Fig. 2). Other physical findings, such as skin and neurological symptoms, were normal. However, considering the possibility of a skull base fracture and abuse, the child

was urgently admitted to the hospital on the same day for close examination and treatment. Regarding birth history, he was born at a gestational age of 38 weeks and 4 days, weighing 2846 g, via vaginal delivery and no perinatal abnormality was noted at birth and at 1- and 4-month checkups. Moreover, the administration of vaccines was timely. The mother claimed to have no eventful family history, and the child was living with both his parents.

On admission, the patient was in good health, height 74.2 cm (+1.8 SD), weight 9.64 kg (+1.1 SD), temperature 37.4°C, blood pressure 96/53 mmHg, pulse 130 beats/min, respiratory rate 22 breaths/min, SpO₂ 99% (room air), head and neck; swelling and hematoma in the right eyelid, hemorrhage in the conjunctiva of the eye, no pupil irregularity, No abnormalities of light reflex, no abnormalities of eye movement, no pharyngeal redness, no enlarged tonsils, no cervical lymphadenopathy, chest; clear respiratory sounds, heart sounds; pure, clear.

Abdomen; soft, no increased/weakened intestinal peristalsis, skin; no obvious subcutaneous bleeding except on the face, no burns or trauma findings.

No abnormal findings of note were noted (Table 1).

Whole-body bone radiograph: No fracture or other suspicious findings.

Simple CT scan of the head revealed a wedge-shaped, nodular structure with a heterogeneous, highly



Fig. 1 Findings at the initial ophthalmologic examination (published with written consent from the guardian)



Fig. 2 Findings at the initial visit to the department (with written consent from the parent/guardian)

Table 1 blood test findings

< Blood count >			< Biochemistry >		
WBC	16100	/ μ L	TP	6.9	g/dL
WBCfractionation			Alb	4.9	g/dL
Neu	21	%	CK	245	U/L
Lympho	71	%	AST	50	U/L
Mono	5	%	ALT	32	U/L
Eosino	3	%	LDH	335	U/L
RBC	4.96	10^6 / μ L	Cr	0.3	mg/dL
Hb	12.8	g/dL	BUN	11	mg/dL
Ht	37.5	%	Glu	107	mg/dL
Plt	31.2	10^4 / μ L	Na	142	mEq/L
< Coagulation >			K	5	mEq/L
APTT	33	sec	Cl	108	mEq/L
PT	112	%	Ca	10.8	mg/dL
PT-INR	0.95		CRP	0.02	mg/dL
Fibrinogen	195	mg/dL			

absorptive area in the right orbit above the nasal side (Fig. 3). No other intracranial hemorrhage or occupying lesions, no obvious fracture including the orbital floor.

Contrast-enhanced MRI scan of the head fat-suppressed T2-weighted image shows a high-signal component with hemorrhage within the muscle cone in the right orbit (Fig. 4). There is an irregular multifocal cystic structure. Fluid level formation was observed, with no obvious contrast effect. Although biopsy could not be performed due to intraorbital lesion and his age of infant, the presence of intraorbital lymphangioma was thought to be from typical MRI finding. At the time of

examination, the possibility of abuse was considered, but there was nothing suspicious about the parents' behavior, and physical examination and imaging results were negative for abuse, and the patient was thought to have intra-orbital hemorrhage associated with intra-orbital lymphangioma. After admission, the patient was carefully monitored, and slow resolution of the periorbital subcutaneous hematoma was observed. Since there was no protrusion of the eyeball or obvious ocular motility disorder, and the patient was in good general condition, we referred him to the ophthalmology department of another hospital to determine whether further surgical treatment was appropriate. At

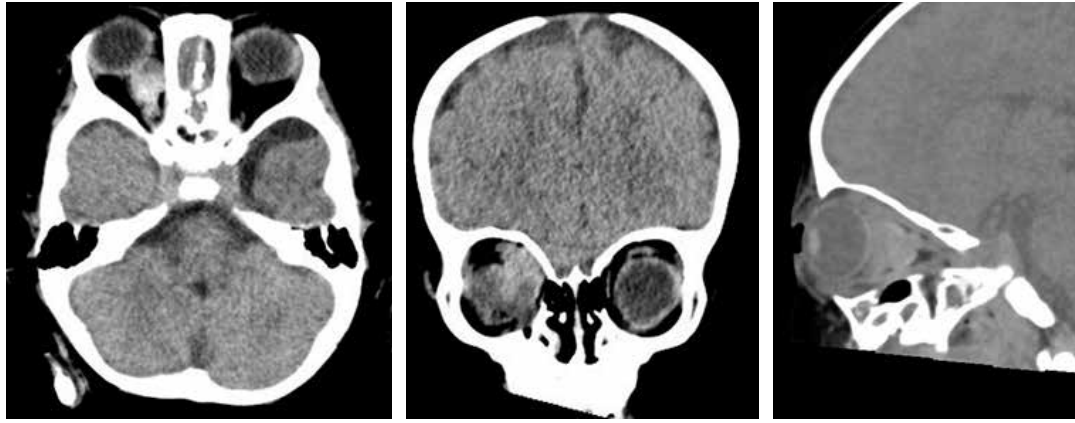


Fig. 3 Simple computed tomography scan of the head
A wedge-shaped, nodular structure with a heterogeneous and highly absorptive area was detected in the right supraorbital nasal region.
No other intracranial occupying lesions and no apparent fracture including the orbital floor were observed.

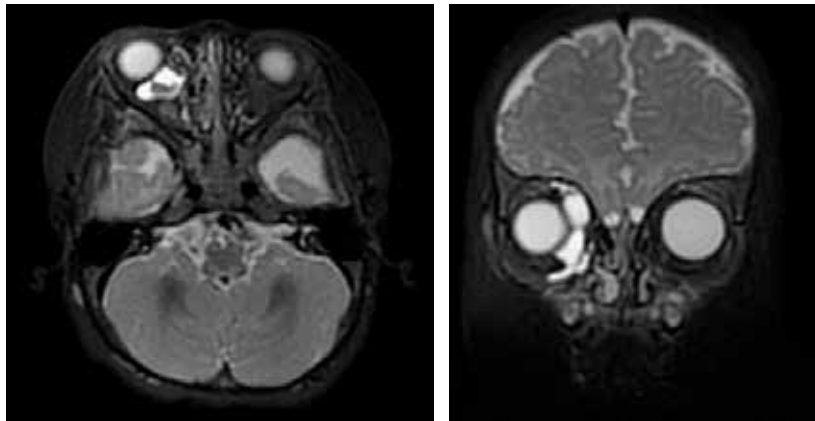


Fig. 4 Contrast-enhanced magnetic resonance imaging scan of the head
Fat-suppressed T2-weighted image shows a high-signal component with hemorrhage within the muscle cone in the right orbit.
An irregular multifocal cystic structure and fluid formation were noted, with no clear contrast effect.

the referral hospital, oral treatment with sirolimus was started, and the tumor was shrinking.

DISCUSSION

Lymphangioma is a histologically benign disease that is mainly found in the neck and axillae. The incidence of lymphangioma is estimated to be 1 in 12,000, and its occurrence is rare in the orbit. The incidence of intraorbital lymphangioma is estimated to be 1.4 per 100,000 people [1], accounting for 1%-8% of all the intraorbital masses. The upper nasal quadrant is predominantly affected [2], and approximately 85% have ocular protrusion, 73% have ptosis, and 46% have ocular motility disorder [3].

On ultrasonography, the cystic mass is seen as a multifocal cystic mass with internal septa. CT scan shows hypo-absorption of the mass and a mono- or multifocal cyst inside. The internal septal structure is often more indistinct than on ultrasound. MRI scans usually do not show contrast effects on the mass, and MRI scans provide more detailed visualization of the mass properties than CT scans. The lesions of lymphangioma are generally comprised of low signal on T1-weighted images and high signal on T2-weighted images [4-6]. In our case, MRI findings were com-

patible with those typical images of lymphangioma. The Japanese Society of Pediatric Surgery defines lymphangioma as “a mass lesion with one or more lymphoid cysts or dilated lymphatic vessels that are clustered or scattered within the lesion. The cyst must meet one of the following criteria: the cyst fluid must contain lymph fluid, the cyst wall must be covered by lymphatic endothelium, or other diseases must be ruled out. As for the lymphatic fluid within the cyst, histopathological examination should be considered; however, if the examination is difficult to perform, the diagnosis is defined as a combination of the aforementioned imaging studies. Wiegand *et al.*, and Woo *et al.*, reported that some cases of lymphangioma are also diagnosed from imaging studies with intraorbital and periorbital lesions [7, 8]. In the present case, the diagnosis of intraorbital lymphangioma was also carried out on the basis of the imaging findings, considering the occurrence of complications associated with biopsy of lymphangioma due to intraorbital lesion and his infant age. No diagnostic criteria have been developed to date that are confined to the orbit, and because of the relatively low prevalence of orbital lymphangioma and the great variety of its manifestations, effective treatment is not currently available [1].

Table 2 Panda's eye sign classified table

· Traumatic	· Infectious
Minor events like the plucking of the eyebrows	Severe pediatric adenovirus infection
Non-accidental injuries	Periorbital leukoderma
Thoracic trauma and crush injuries	Frontal sinus mucocele
Following vigorous sneezing, coughing, or vomiting	· Metabolic
Posttraumatic orbital emphysema	Amyloidosis
· Vascular	Myxedema
Subarachnoid hemorrhage (SAH)	· Genetic
following the rupture of an ophthalmic artery aneurysm	Hemophilia
Venous sinus thrombosis (Including intraorbital lymphangioma)	· Malignancies
Giant cell arteritis	Hematological malignancies
Trigeminal autonomic cephalalgia	Metastatic neuroblastoma
Benign intracranial hypertension	Orbital metastasis of solid malignancies
· Immune-mediated	Kaposi sarcoma
The neonatal lupus erythematosus	Multiple myeloma
Chronic atypical neutrophilic dermatosis with lipodystrophy and elevated temperature (CANDLE) syndrome	
Lichen planus pigmentosus	
Sweet syndrome or acute febrile neutrophilic dermatosis	

source (e.g. quotation) : M Das J, Munakomi S : Raccoon Sign. StatPearls [Internet].2021

The most common treatment and management is to follow-up the patient with orbital lymphangioma showing decreased vision, exposed keratopathy, or severe cosmetic problems [9]. The treatment depends on the size, location, and vascular composition of the cyst, and sclerotherapy may be considered for cysts >2 cm that can be accessed with a needle [10, 11]. Moreover, oral sildenafil, and oral sirolimus are effective treatment options, with sirolimus reported to have better results for those malformations with a venous component [10, 11]. Surgical resection is also considered as an alternative; however, total removal of the cyst is usually difficult because of the indistinct tumor capsule and the indistinct borders due to invasion into the surrounding normal intraorbital structures. Careful consideration is necessary for this as the recurrence rate is reported to be as high as 40%–60% [13]. There is no clear consensus on the prognosis and course of the disease, as treatment options vary widely [1]. In our case, the patient received sirolimus, and the tumor was shrunken. Careful consideration should be needed, and surgical resection should be considered if recurrence occurred.

The patient was initially presented with periorbital subcutaneous hemorrhage, but such a course is rare with intraorbital lymphangioma. Most intraorbital lymphangiomas are found in childhood; however, we found no reports of periorbital subcutaneous hemorrhage or subconjunctival hemorrhage as the initial symptom, as in this case, in any of the previous studies we reviewed through the Central Journal of Medicine and PubMed. Therefore, the apparent mechanism of the hemorrhage was unknown. However, although the nature of lymphangiomas is not clear, some have been reported to be continuous with venous circulation and rarely associated with abnormal arteriovenous morphology [14–18]. Although continuity with veins and abnormal arteriovenous morphology has not been proven in this case, in light of these reports, we assume that the hemorrhage may be associated with enlargement of the lymphangioma or with disruption of some veins due to mild irritation. Moreover, increased lymphangioma, protrusion of the eyeball, eye movement disorder, and ptosis were reported, but these symptoms were not present in this case, and the main

complaints were subcutaneous hemorrhage around the orbit and hemorrhage under the conjunctiva of the eye. Hence, it can be assumed that this may have been an early manifestation of intraorbital lymphangioma.

Abusive head trauma in infants and young children (AHT) is a general term for infant head trauma due to abuse, and was proposed by the American Academy of Pediatrics in 2009 [19]. AHT occurs in 25–30 out of 100,000 infants. About half of AHT cases occur in children under 1 year of age, especially those under 6 months of age, and boys are slightly more frequently affected than girls [20, 21]. In response to this, the prevalence of AHT in children with periorbital skin lesions has increased, and the incidence of AHT is now estimated to be about 25–30 per 100,000 infants. In this context, if a periorbital subcutaneous hemorrhage (panda's eye sign) is observed, the pediatrician should first have in mind a skull base fracture or maltreatment. This sign refers to a clinical presentation [22] consisting of unilateral or bilateral progressive periorbital mottled hemorrhage and edema. These findings are usually not seen on initial evaluation and appear 1–3 days later, but are strongly predictive of skull base fractures, if bilateral [23], and they are present in 50%–60% of cases when skull base fractures are present [24]. However, a small number of differential diseases for this sign have been reported (Table 2). When the panda's eye sign is present, as in the present case, it is important to keep in mind the differentiation of a wide variety of diseases, including intraorbital lymphangioma, as well as abuse.

CONCLUSION

When subcutaneous hemorrhage is observed around the eyelid, as in this case, causes other than trauma or abuse should be considered. Intraorbital lymphangiomas can cause sudden intra-lesion hemorrhage, and the presence of a tendency toward hematoma enlargement or optic neuropathy (visual disturbances, abnormalities of the light reflex, etc.) is highly urgent and should be considered, including surgical treatment.

ACKNOWLEDGMENTS

Code of Ethics

Parental consent was obtained for this report.

In addition, since this report is a case report using unlinkable anonymized personal information, and personal information is not covered by the “Ethical Guidelines for Medical Research Involving Human Subjects,” approval by the Ethics Committee was not required.

Conflict of Interest

There are no disclosures regarding conflicts of interest as defined by The Tokai journal.

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