

Prenatal diagnosis of dacryocystocele: a possible marker for syndromes

R. Sharony, J. Raz*, R. Aviram, I. Cohen, Y. Beyth and R. Tepper

Ultrasound Unit, Departments of Obstetrics and Gynecology, and *Ophthalmology, Sapir Medical Center, Kfar-Saba, Sackler Faculty of Medicine, Tel Aviv University, Israel

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ABSTRACT

We present our experience of the sonographic prenatal diagnosis of dacryocystocele and review the literature. This lesion can be distinguished from a facial hemangioma, dermoid or anterior encephalocele by the ultrasound findings and Doppler flow studies. These conditions are separate entities and their associated diagnosis and prognosis are very different in each case. As dacryocystocele may be part of numerous syndromes, its prenatal visualization raises the rare possibility of associated anomalies.

INTRODUCTION

Six cases of dacryocystocele were diagnosed in our unit during 1995–98 from 6000 fetal system surveys of low- and high-risk pregnancies. Dacryocystocele, or lacrimal duct mucocele, originates from a stricture in or obliteration of the lacrimal system, resulting in a fluid-filled, closed sac¹. It is an uncommon and benign condition in neonates. As many cases are mild and may resolve spontaneously antenatally or in the immediate perinatal period, the prevalence of this anomaly is probably higher than reported^{2,3}.

We present our experience and review of the literature concerning this entity and other periorbital masses. As dacryocystocele may be part of several congenital syndromes, these conditions are described. Meticulous

ultrasound examination of specific fetal systems is recommended.

CASE REPORTS

Six fetuses with dacryocystocele have been diagnosed in our ultrasound unit during the past 4 years. An Acuson 128 XP/10 ultrasound machine (Acuson, Mountain View, CA, USA) with a 3.5-MHz abdominal transducer was used. A normal fetal survey at 20 week of gestation was obtained in all patients. In two cases, in which amniocentesis was performed owing to advanced maternal age, a normal karyotype was obtained (Table 1). No accompanying fetal anomalies were detected, although one neonate died of Canavan disease. The dacryocystocele was demonstrated after the 27th week of gestation in all cases. The features were those of a hypoechogenic mass, inferiomedial to the orbit with no blood flow in or around it (Figure 1). The appearances of the face and anterior brain were normal. Blood flow in the middle cerebral arteries was normal. The lesion was unilateral in five cases and bilateral in one case.

The prenatal diagnosis was confirmed after delivery in all cases. The newborns were found to have isolated dacryocystocele with no additional pathology (Figure 2). The postnatal course was variable in the different cases; in

Table 1 Demographic and sonographic data in six cases of prenatal diagnosis of dacryocystocele

Case number	Gestational age at diagnosis (weeks)	Maternal age (years)	Gravida	Para	Lesion	Outcome	Comments
1	27	28	1	0	10 mm, right	died at 6 months	Canavan disease
2	29	40	4	1	4 mm, right	healthy	IVF, missed twin
3	28	27	5	4	6 mm, left	healthy	
4	28	40	6	3	5/3 mm, bilateral	healthy	maternal diabetes
5	36	26	1	0	10/4 mm, bilateral	healthy	left pyelectasis 7 mm
6	29	21	1	0	4 mm, left	healthy	MCKD+ dysplastic kidney

IVF, *in vitro* fertilization; MCKD, multicystic kidney disease

Correspondence: Dr R. Sharony, Department of Obstetrics and Gynecology, Sapir Medical Center, Kfar-Saba, Israel 44281



Figure 1 Prenatal ultrasound scan of nasolacrimal dacryocystocele (NLDC)



Figure 2 Postnatal photograph of dacryocystocele. A bluish swelling was seen over the lacrimal sac area, inferior to the medial canthal ligament (arrow)

two newborns the dacryocystocele resolved spontaneously within a few days after delivery. Conservative management, massage and topical antibiotics cured two further newborns. The dacryocystocele was evacuated following a second probing in one baby. The last case, complicated with dacryocystitis, was treated with topical and intravenous antibiotics and eventually required drainage via a nasal approach.

DISCUSSION

The development of the lacrimal drainage system begins around the 6th week of pregnancy. As the surface ectoderm in the naso-optic fissure thickens, an epithelial cord detaches from it and buries itself between the lateral nasal and maxillary processes. Cephalic and caudal growth of this epithelial cord will give rise to the lacrimal canaliculi, sac and duct⁴. Canalization of the nasolacrimal pathway begins at about 12 weeks of gestation and is complete from as early as 24 weeks. However, the nasal (distal) end may perforate only at birth or even later. Atresia of the Hasner valve, at the distal end of the nasolacrimal duct, is the main

cause of simple congenital dacryostenosis. The estimated incidence of dacryostenosis is 6–73%, but only 2–4% of cases are symptomatic. Symptoms arise at about 10–30 days after birth⁵. Dacryocystocele, a much less common disorder, is already symptomatic at, or shortly after, birth. In this condition, additional functional obstruction at the orbital end of the lacrimal sac is present together with atresia of the distal end of the nasolacrimal duct. The sac fills with amniotic fluid and intrinsic mucoid secretion, and becomes distended, causing the canaliculi to kink. These then act as a one-way valve, permitting fluid only to enter into the sac⁶.

Two earlier studies reported the antenatal diagnosis of dacryocystocele at 30–36 weeks of gestation^{6,7}. We described the visualization of this lesion as early as 27 weeks, just prior to the fetal age at which canalization is completed. It therefore seems logical that the fetal survey performed at 20 weeks would have been normal.

An inferiomedial periorbital mass diagnosed antenatally by ultrasound may be the manifestation of one of the following: anterior encephalocele, teratoma, hemangioma, glioma or rhabdomyosarcoma.

Dacryocystocele is a hypoechogenic mass located inferiomedially to the orbit. It can be imaged in the coronal plane of the face and in a parasagittal plane in which the side of the nose and the medial angle of the eye orbit(s) are included. Hemangioma is cutaneous in origin, located in the head or neck, septated or solid^{8,9}, and with a typical Doppler flow pattern¹⁰. Dermoid cysts arise most commonly in the superiotemporal quadrant (49% of cases) and have been found in the inferiomedial quadrant in only 11% of cases¹¹. A dermoid cyst has a complex appearance on ultrasound with areas of calcification frequently present⁶. Anterior encephalocele is usually a mid-line defect accompanied by a calvarial defect and hydrocephalus⁶.

Dacryocystocele is a structural anomaly that may be seen in numerous syndromes and may serve as their marker. Some of the additional characteristic findings of these syndromes can also be detected by prenatal ultrasound. This fact encourages the search for additional markers of the specific syndromes once a dacryocystocele is demonstrated. The face, limbs, kidneys, eyes and genitalia are the organs most frequently involved in these syndromes.

The genetic counsellor of the couple should discuss the fact that the dacryocystocele is usually an isolated finding. However, rarely, it might be part of a syndrome that may not be identified before delivery.

Early neonatal presentation of other conditions includes neurofibromatosis, lymphangioma and rhabdomyosarcoma, demonstrated ultrasonographically as a solid orbital mass⁶. It should be remembered that these conditions are extremely rare.

In summary, the nature of the sonographic findings, including location, size, time of appearance, echogenicity and Doppler flow characteristics facilitate the antenatal diagnosis of dacryocystocele.

The sonographic appearance of dacryocystocele allows prenatal diagnosis during the late second trimester and

appropriate postnatal management aimed at decreasing the incidence of neonatal dacryocystitis^{12,13} and respiratory distress¹⁴. Prenatal diagnosis of dacryocystocele and additional malformations as part of a genetic syndrome will assist in the diagnosis of a known pattern of anomalies. In cases of syndromal dacryocystocele, the prenatal diagnosis will assist in the decision-making process of the parents and their physician.

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