New imaging techniques to diagnose orbital cysts in utero

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Congenital dacrocystocoele is an uncommon developmental malformation of the nasolacrimal system usually presenting at birth with periocular

INTRODUCTION

We present the case of a 33 year old G2 P1 Caucasian woman who was referred to the tertiary Foetal Medicine Unit at St. Michael's Hospital, Bristol with an abnormality detected on ultrasound scan adjacent to the foetus right eye. This was detected at a routine growth scan at 32 weeks. Antenatal booking clinic blood tests including an infection screen were normal. Combined first trimester screening revealed a low risk for Down's, Edwards and Patau syndrome and the previous 19 week fetal anomaly scan had not demonstrated any abnormality.

The 32 week ultrasound scan showed an avascular fluid-filled area measuring $8.6 \times 9.8 \times 10.5$ mm within the right orbit, inferomedial to the globe. There was no calcification and blood flow could not be detected within the lesion (Figure 1). Further imaging at 35 weeks gestation included an MRI scan (Figure 1c) and suggested the presence of bilateral medial orbital cysts.

CASE PRESENTATION

There was a vague family history that some form of congenital orbital cyst had been present in the father but no further details were available and there were no known visual consequences. The pregnancy had been uneventful to date but the parents were very anxious to rule out serious pathology. Discussions regarding early induction of labour with the neonatal team took place. An ophthalmology opinion was sought to determine the likelihood of this being a malignant lesion or pathology that could affect development of the globe. After reviewing the dynamic ultrasound scan and pre-natal MRI, it was felt that the presence of bilateral cystic lesions with normal ocular development pointed to a provisional diagnosis of dacrocystocoele. No early intervention was advised and the pregnancy continued with a much lower level of anxiety.

A baby boy was delivered by normal vaginal delivery at 38 weeks with good APGAR scores. At birth he had bilateral swellings inferomedial to the globes consistent with a diagnosis of dacrocystocoeles (Figure 1d). Ophthalmic examination was otherwise normal and the swellings resolved with systemic antibiotic treatment and later gentle lacrimal sac massage.

DISCUSSION AND CONCLUSION

A congenital dacrocystocoele (amniontocoele) is an uncommon variant of nasolacrimal duct obstruction, which occurs when there is both proximal and distal obstruction of the lacrimal system during foetal development. The development of the lacrimal system is first observed at day 32 of embryonic development, when a groove forms between the maxillary and frontonasal prominences and develops into a cord of epithelium. By week 10 the formation of a lumen in the lacrimal cord has taken place which

swelling or neonatal dacrocystitis. This case report demonstrates how new imaging techniques can aid pre-natal diagnosis and help to reduce diagnostic uncertainty, parental anxiety and the potential need for early delivery of the baby.

Key Words: Dacrocystocoele; Pre-natal; Imaging

coincides with cavitation of the interior meatal lamina. Canalisation of the cord begins at 4 months gestation and eventually results in communication with the inferior meatus. Valve-like folds within the cords cover the puncta and nasolacrimal outlet [1]. This is the last portion to become patent, usually becoming patent at term. If this remains imperforate, then a cystic dilatation may result, this accumulates amniotic fluid and mucus [2]. The most common areas of obstruction are thought to be due to a persistent membrane at the valve of Hasner and a functional obstruction at the common canaliculus or valve of Rosemuller [3], resulting in a congenital dacryocystocele. It usually presents as a bluish swelling inferomedial to the medial canthus in the neonate [4] but with improved resolution of ultrasound scans and other imaging techniques, prenatal diagnosis is becoming increasingly prevalent. The differential diagnosis includes haemangioma, encephalocele, glioma, dermoid cysts and malignant processes [5]. Bilateral dacrocystocoeles are found to be present in 25% of cases

Dacryocystoceles can be identified prenatally by ultrasound scanning. They are seen as hypoechogenic masses located inferomedially to the orbit. Young-Hwa [4] found mean gestational age at first detection on ultrasonography is 31.2 weeks with the majority being unilateral [6]. Clinically those with lacrimal sac diameters less than 5 mm are considered normal, and those over 5 mm considered pathological [7]. These findings can be distinguished from haemangiomas and dermoids by Doppler scanning and the presence of calcification [8], but prenatal magnetic resonance imaging (MRI) can delineate structures more clearly than conventional ultrasound scanning. Given dacryocystoceles can be isolated findings or part of other orbital or syndromic anomalies, MRI can be useful for identifying related features and to aid with prognostic and treatment planning [9].

MRI is a non-invasive form of imaging where tissues are exposed to a short electromagnetic pulse which causes rearrangement of the hydrogen nuclei within them. As the pulse subsides, the hydrogen nuclei return to their normal resting state, and in doing so, re-emit some of the radiation they have absorbed. Receivers then detect this change in signal, and depending on proton density, tissue components and their magnetic properties a 3-dimensional image is created (Figure 1b). There are no known risks of in utero MRI but it has been stated that the specific absorption ratio (measure of rate at which energy is absorbed by tissue when exposed to electromagnetic field) and the number of sequences obtained should be kept as low as possible [10].

Although congenital dacryocystoceles are benign lesions, their presence can be associated with significant complications such as neonatal nasal obstruction. This can result in respiratory distress if a large or bilateral dacrocystocoele is present with intranasal extension [6].

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The importance of recognising the diagnosis of congenital dacrocystocoeles through high quality imaging lies in the ability to reassure the parents of the benign nature of the cysts, to avoid unnecessary

early induction of labour and to enable commencement of appropriate antibiotic treatment without delay postnatally.



Figure 1) *GW* (gestational weeks); (a) 2-dimensional USS of foetal head at GW 32 showing unilateral (b) 3-dimensional USS at GW 32 showing swelling inferior to right orbit (c) Pre-natal MRI at GW 35 showing bilateral medial orbital cysts (d) Photo of 1 day old infant showing area of swelling inferomedial to right globe

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