ECHOGRAPHIC DIAGNOSIS OF ADVANCED COATS' DISEASE

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SUMMARY

Standardised echography was utilised to diagnose two cases of advanced Coats' disease. The characteristic acoustic features included extensive, poorly mobile retinal detachment; dense, dispersed subretinal opacities; looping of peripheral retina and absence of solid mass lesion and calcification. As enucleation of two eyes was necessary, it has proved possible to correlate the echographic and histological findings of this disease. The echographic features of other common causes of leukocoria are also discussed.

Coats' disease constitutes one of the important causes of leukocoria. In young children early stages of the disease may be missed mainly due to the lack of presenting symptoms. When suspected, as in cases of squint, the diagnosis can easily be reached clinically, by observing the distinguishing retinal telangiectases and associated exudates. As the disease progresses, continuous deposition of subretinal lipids, fibrin and phagocytes produces extensive 'exudative' retinal detachment, and the common sequelae of uveitis and rise of intraocular pressure, bringing the disease to the notice of parents and health personnel.

The differential diagnosis of advanced Coats' disease, particularly against retinoblastoma, can be extremely difficult and usually requires the summation of various diagnostic aids, in addition to detailed and thorough clinical examination, before reaching the correct diagnosis and undertaking the appropriate course of action. If detected and treated at an early stage, useful vision may be preserved and secondary complications and eventual loss of the eye may be prevented.²

Ultrasonography (echography) with its safety, sensitivity and unique ability of 'tissue differentiation'³ is ideally suited for the investigation of children with Coats' disease and other 'leukocoric' conditions.

Two cases of children with advanced Coats' disease are presented and the characteristic echographic features are described. As enucleation of the two eyes was necessary, it

has proved possible to correlate the echographic and histological findings. Echographic features of other causes of leukocoria are also discussed.

ECHOGRAPHIC METHODS

The ultrasound examinations were carried out using the instruments and techniques of 'Standardised Echography'.^{3,4}

The equipment used was the Ophthascan-S combined standardised A- and real time contact B-scan, the Kretz-technik 7200 MA standardised A-scan, and the Ultrascan Digital-B 'system 4'. The A-scan examination consisted of screening of eight meridians antero-posteriorly using 'tissue sensitivity' (high) and measuring (low) decibel levels. The B-scan examination was performed using axial, transverse and longitudinal sections also employing high and low decibel levels. By using the topographic, quantitative and kinetic examinations described in this technique, it was possible to assess the extent and mobility of retinal detachment, degree of subretinal exudates, and exclude the presence of solid mass lesion and calcification. In both children, echography was carried out during examinations under general anaesthesia (EUA).

CASE REPORTS

Case One

A one-year-old boy presented with a few weeks' history of 'watery and red left eye'. There was no relevant paediatric or family history. The left anterior chamber was shallow and raised intraocular pressure was suspected on palpation of the globe. A provisional diagnosis of buphthalmos was made and a course of Acetazolamide was given until an EUA and possible goniotomy was performed. Echographic examination was not available at that time or at the time of EUA which showed a marginal rise of left intraocular pressure, corneal diameters of 12 mm right and 14 mm left. A very shallow left anterior chamber was judged to preclude goniotomy. A small pupil due to posterior synechiae, and a cataract were also noted and prevented useful fundoscopy. Examination of the right eye

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Fig 1 (top).

Fig 1 (bottom).

Fig. 1. Case-1. Two axial B-scan sections. Top is a vertical axial, showing a closed funnel retinal detachment. The retina is taut and the inferior leaf shows a large loop extending into the periphery (arrow). Bottom is a horizontal axial section. The funnel is more opened. Dense opacity anterior to retina indicates cyclitic membrane (asterisk).

was normal. It was decided therefore to free the syn- aechiae and perform a Fuch's cataract wash out. Post-operatively a yellow pupillary reflex was noted but fundus details remained obscure. Ultrasound examination at this stage became available and was performed during a second EUA.

ECHOGRAPHIC FINDINGS

B- scan examination demonstrated a total almost closed funnel retinal detachment (Fig 1). The retinal leaves were taut and poorly mobile as they stretched from the optic disc to a highly reflective 'cyclitic' membrane. Additionally the retina showed marked convolution and looping towards the periphery. Dense, dispersed subretinal opacities, seen best on transverse sections (Fig 2), showed constant, slow motion on kinetic echography. Standard- ised-A scan examination demonstrated a retinal detachment and subretinal deposits in all quadrants (Fig 2). Both A- and B-scans showed no evidence of solid mass lesion and low sensitivity settings did not demonstrate any calcium foci. Based on the above findings the echographic diagnosis of Coats' disease was made.

Further investigations and management

An electro-retinogram was performed during the second EUA. This showed no response in the left eye but normal pattern in the right. Because of the constant irritation, poor visual prognosis and the remote possibility of retinoblas- toma it was decided to enucleate the eye. This enabled his- tological examination which confirmed the diagnosis of Coats' disease and correlated well with the echographic findings (Fig 3).

Case Two

A one-and-a-half-year-old boy presented to the eye casualty with a few days history of red right eye with abnormal pupillary reflex, noted by his mother. The child was born two weeks prematurely, one of identical twins. There were no neonatal problems and family history was negative especially for retinoblastoma. Examination showed an injected right eye with shallow anterior cham- ber. The right pupil was irregular and sluggish, with dilated iris vessels and heterochromia. A thick, vascular-

Fig 2 (top).

Fig 2 (bottom).

Fig. 2. Case-1. Top is a transverse B-scan demonstrating the peripheral looping of the retina (arrows). By avoiding scanning through the lens, more resolution is obtained, allowing better visualisation of the subretinal, dispersed (cholesterol) opacities. Bottom is an A-scan. Thick, tall retinal spike (R) is separated from the scleral signal (S) by a group of low reflective spikes indicating subretinal opacities. I = initial spike.

Fig. 3. Case-1. Histological section of the enucleated eye. The retina is extensively detached and showing marked convolution and looping. Large quantities of subretinal exudates are seen, including the characteristic cholesterol clefts.

ised retro-lental opacity was also noted. An EUA including echographic examination and C.T. scanning was performed shortly afterwards, during which fundoscopy of the right eye showed extensive retinal detachment with abnormal, enlarged vessels on the retinal surface. A small, clear vitreous funnel allowed the view of a white exudative response on the deeper portion of the retinal leaves. Detailed ophthalmoscopy of the left eye demonstrated no retinal abnormalities.

Echographic Findings

A total, narrow funnel, retinal detachment was seen. The retinal leaves appeared to extend anteriorly from the optic nerve to the posterior lens surface, before looping extensively into the periphery (Fig 4). The retina was thickened and showed reduced mobility. The subretinal space was completely filled with small, dispersed opacities exhibiting continuous slow motion. There was no evidence of solid mass lesion or calcium deposition. An echographic diagnosis of Coats' disease was made.

Further Investigations and Management

C.T. scan examination showed the right globe to be almost entirely occupied by dense non homogenous tissue. There was no evidence of intraocular calcification; other details were not seen (Fig 5). The findings were compatible with the diagnosis of advanced Coats' disease. Family screening, involving examination of the twin brother under EUA, and of both parents revealed no ocular abnormalities. In spite of the poor visual prognosis, it was felt appropriate to preserve the eye to promote the development of the bony orbit and achieve good cosmetic result if eventual enucleation proved to be necessary. The child was treated with topical steroids and cycloplegics, but nine months later the eye became increasingly painful with further shallowing of the anterior chamber and rise of intraocular pressure. Compared to his twin brother he also

appeared to lag behind in growth. It was therefore decided to enucleate the eye. Echography performed immediately before surgery demonstrated significant increase in

Fig 4a.

Fig 4b.

Fig 4c.

Fig. 4. Case-2. 4a, transverse B-scan echogram of the upper globe shows a total retinal detachment with peripheral looping (arrows). Subretinal opacities can be seen. 4b is a longitudinal section demonstrating a funnel retinal detachment attached to optic nerve (ON). Peripheral convolution of the retina is shown (arrow). 4c presents another longitudinal section demonstrating taut retinal leaves stretching from the optic nerve to the posterior surface of the lens (arrow). Kinetic echography showed the retina to be poorly mobile.

Fig. 5. Case-2. C.T. scan demonstrating increased density in the right globe. No calcification is noted. A 'less dense' central area was interpreted as clear vitreous cavity but no further ocular details are seen.

density of the subretinal exudates and further retinal immobility (Figure 6). Enucleation allowed histological examination of the eye. This confirmed the diagnosis of advanced Coats' disease and correlated closely with the echographic findings (Fig 7).

Fig 6 (top).

Fig 6 (bottom).

Fig. 6. Case-2. Echograms taken immediately before enucleation. Top is the same section as in Fig 4a and bottom is the same as in Fig 4c. Massive increase in subretinal exudation has almost masked other details.

DIAGNOSIS

Although reported earlier by a number of authors⁵ Coats' disease was first described in detail by George Coats in

Fig 7a.

Fig 7b.

Fig 7c.

Fig. 7. Case-2 Histology. 7a is the macroscopic appearance of the eye. Massive subretinal exudates are seen containing shiny cholesterol and dark pigment particles. In 7b thickened, gliotic retina appears to adhere to the posterior lens surface and is seen in 7c, turning back on itself. Subretinal cholesterol clefts are also demonstrated in this figure.

Fig 8a.

Fig 8b.

Fig. 8. Retinoblastoma. 8a is a B-scan showing a mushroom shaped, calcified tumour with shadowing effect (arrow). 8b is a 'tissue sensitivity' A-scan. I = initial spike, T = tumour surface spike, S = scleral spike. The tall spikes between T and S represent the highly reflective, calcified tumour tissue.

1908.¹ Assuming an inflammatory origin; he labelled it 'external haemorrhagic retinitis'. Some authors also attempted to include other disease entities under this

Fig. 9. ROP Total funnel retinal detachment with peripheral 'cyst' formation (Arrow). Unlike Coats', the subretinal space is clear and the vitreous cavity is opacified.

label.⁵ It is now accepted however, that Coats' disease should only be applied to the specific disorder of progressive (developmental) retinal telangiectases associated with variable degrees of retinal and subretinal exudates and retinal detachment.^{6,7} Recent microscopic (including E.M.) studies have also demonstrated the abnormal permeability⁸ and structure⁹ of retinal vessels characteristic of this disease.

Early and progressive exudation of lipids leading to retinal detachment is a constant feature of Coats' disease. Exudates contain numerous quantities of lipid and pigment-laden macrophages (foam cells), cholesterol crystals, seen as empty clefts on microscopic sections, pigment epithelial cells and free floating pigments. Infiltration is heaviest in the outer retinal layer and subretinal space, with the choroid, vitreous and anterior segment relatively spared.

The echographic findings in Coats' disease seem to correspond with the histo-pathological process described above, and in our two cases correlated well with the histological examinations of the two enucleated eyes.

In the early stages of the disease, echography is unspecific showing areas of retinal detachment with or without subretinal opacities.¹⁰ The absence of solid mass and calcification usually rules out retinoblastoma. Additionally the ophthalmoscopic appearance of early Coats' disease will reveal the characteristic telangiectases and retinal exudates. In its advanced form visualisation of the fundus is often hampered by the high, closed funnel retinal detachment and other factors such as corneal oedema, posterior synaechiae and retrolental (cyclitic) membrane. The echographic appearance however becomes more diagnostic as the disease advances and consists of:

- (1) narrow or closed funnel retinal detachment with looping of the peripheral retina
- (2) poor retinal mobility
- (3) dense, dispersed, low to medium reflective subretinal (cholesterol) opacities exhibiting constant slow convection movement
- (4) absence of solid mass lesion and
- (5) no evidence of calcium foci.

It is of interest to note that looping or 'cyst formation' of

Fig. 10. P.H.P.V., a longitudinal section showing vitreous bands originating from the optic nerve head.

the peripheral retina may also be seen in cases of advanced retinopathy of prematurity (ROP).¹¹ Calcification of the detached retina, and other ocular tissues, may also be encountered in long standing Coats' disease. In such cases calcium deposition is seen along expected anatomical structures and is clearly distinguished from that seen in retinoblastoma.

The reliable differentiation of advanced Coats' disease depends on an adequate knowledge of the echographic features of other common causes of leukocoria.

Retinoblastoma produces a solid mass lesion. Its most significant echographic finding is the presence of calcification. This is a diagnostic feature in the majority of cases.¹² Calcification is easily seen on echography as highly reflective areas with shadowing effect (Fig 8). Calcification however may be absent in a small percentage of retinoblastoma cases,¹³ particularly in tumours presenting as diffuse lesions along retinal surfaces. These may be difficult to diagnose but further echographic findings such as tumour vascularity (best seen on standardised-A scan as fast vertical movements of tumour spikes), irregular internal structure and increasing growth on serial examinations will help reach the correct diagnosis.

Retinopathy of Prematurity (ROP) produces a retinal detachment with a prominent retro-lental membrane. In its advanced form a thick, highly reflective opacity appears to obliterate the vitreous cavity anterior to a narrow or closed funnel retinal detachment. Looping or cystic formation of the peripheral retina is common (Fig 9). The subretinal space, unlike Coats' disease, is relatively clear or contains small, thinly dispersed opacities. No solid mass lesion is seen and calcification is absent unless the disease is long standing.

Persistent Hyperplastic Primary Vitreous (PHPV) is easily detected on echography by the presence of a single or multiple 'bands' extending from the optic disc to the lens and peripheral retina (Fig 10). The axial length of the affected eye is usually shorter than that of the fellow eye.

Other imaging techniques such as computerised tomographic (CT) scanning and magnetic resonance imaging (MRI) have been successfully used to diagnose cases of leukocoria.^{14,15,16} In addition to exposure to ionising radiation and high cost, they are inferior to echography in delineating the kinetic properties of lesions.^{15,17} They are however more applicable in detecting extension of retinoblastoma into the optic nerve and intra-cranially.

In conclusion, echography was accurately utilised to diagnose two cases of advanced Coats' disease. Histological examination of two enucleated eyes correlated well

with the echographic findings. The sensitivity, safety and low cost of this technique and the ability to conduct it as a part of EUA makes echography an ideal tool in the initial diagnosis and serial examinations of children with leukocoria.

Key words: Coats' disease, Diagnostic ultrasound, Echography, Leukocoria, Persistent hyperplastic primary vitreous, Retinoblastoma, Retinopathy of prematurity, Standardised echography, Ultrasonography.

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