Coats Disease: A Case Report

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Case Report

A six-year-old female patient was well till the 11th of June 2002 when she was noticed to have deviation of the eyes of two weeks duration. There was no history of trauma. She is a result of full term normal vaginal delivery. She has 4 siblings who are healthy.

Her development has been normal and she has not had any illnesses of note. She has no similar conditions in her family. She had chickenpox when she was six months old. She is fully vaccinated. She has no known allergies. She has a normal diet and on no medications. Her performance is good at school. Ocular examination: the visual acuity of the right eye was 6/ 6 and of the left was 5/ 60. She had a full range of eye movements and the Lt. eye was slightly divergent. Cycloplegic retinoscopy showed that she was mildly hypermetropic (+1.00 right eye, +1.00 left eye). Slit-lamp biomicroscopy: anterior chambers were normal and there was no afferent pupillary defect. Intraocular pressure of both eyes measured by applanation tonometry was normal. Examination of her fundi by indirect ophthalmoscopy through dilated pupils showed mild subretinal exudation at superior and inferior temporal vascular arcades of the right eye (Fig. 1) and a large subfoveolar yellowish scar surrounded by extensive macular exudation with subretinal exudation mainly at posterior pole of the left eye (Fig. 2).

Figure 1: Mild subretinal exudation of the right eye.

Figure 2: Subfovealar scar surrounded by extensive macular exudation ***of the left eye.***

Pediatric consultation was requested. General physical exam revealed no abnormalities. Full systemic work-up had been carried out. Full blood count showed: Hb 12.2mg/dl, HCT 35.5%, MCV 82.6 fl, MCH 28.4 pg, MCHC 34.4 g/dl, WBC 7.7 x103, and platelets 280×103/microl. Blood film was normocytic normochromic. Sickling test was negative. Blood glucose level was 90 mg/dl. KFT, LFT, ESR, Calcium, phosphorus were normal. CRP was negative. Brain and orbit MRI were normal. Full auto-antibody screen including ANA, ENA and Ds DNA were negative. IgG and

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IgM levels were normal. Infectious screen including CMV-IgM, Toxoplasma-IgM and -IgG, EBV-IgM, rubella- IgM, and Viral capsid antigen-IgM titers were negative. We asked her parents to bring a recent photograph for their child. The photograph was taken about one month before the presentation. We found that she had leukocoria of the left eye (Fig. 3).

Figure 3 Left leukocoria

Fluorescein angiography highlights the underlying vascular malformations. It showed small hyperfluorescent area of telangiectasia that is superotemporal to the macula of the Rt. eye and leaky localized foci of retinal telangiectasia, increased tortuosity, and aneurysmal dilatation in the retinal vessels with capillary dropout in the temporal quadrant of the left eye (Fig. 4). Also, there was well-defined hyperfluorescent area at the left fovea representing submacular Choroidal Neovascular membrane (CNV) (Fig. 5). Ocular Ultrasonography showed no retinal detachment or calcifications. CT scanning showed no calcification, retinal masses or retinal detachment. Based on clinical ground, investigations and Fluorescein angiography findings, the diagnosis of Coats disease was highly suspected.

Four vitreoretinal consultants and one pediatric ophthalmologist confirmed the diagnosis of Coats disease. All advised us to treat retinal changes with laser photocoagulation. So we did diode laser photocoagulation, two sessions for each eye on the 4th of Sept. and 30th of April 2004. Laser settings for each session under general anaesthesia were: power = 230 mV, duration = 100ms, and number of burns for the right eye = 45 and the left eye = 40. Clinically, the retinal exudation decreased slightly and visual acuities remain the same. Fluorescein angiography showed that there are still leaky abnormal vessels, particularly at temporal quadrant of the right eye (Fig. 6).

So we decided to give the child one session of retinocryoptherapy on 28/12/2002 for both eyes in hope to obliterate the leaky vessels. After that, the visual acuity of the left eye slightly improved to 6/60, and of the right eye remained 6/6. The retinal exudation decreased significantly but new dilated abnormal vessels appeared in the upper part of the left retina and the abnormal vessels in the temporal quadrant of the right retina were still present. We examined the child again on May 2003, and we gave her cryotherapy for both eyes on June 2003, to the leaky abnormal vessels at superior part of left retina and temporal quadrant of right retina. Three weeks later, indirect ophthalmology and Fluorescein angiography showed no leakage, exudation or new abnormal lesion in both eyes (Fig. 7) apart from the macular region of the left eye which showed leakage (hyperfluorescent area at left macula).

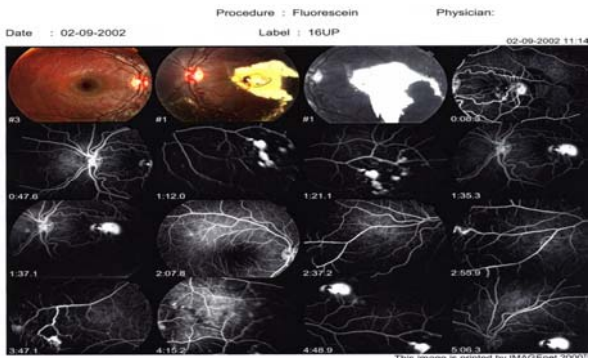

Figure 4: hyperfluorescent area of telangiectasia of the right eye and aneurismal dilatation of retinal blood vessels with capillary drop-out at temporal quadrantof the left eye.

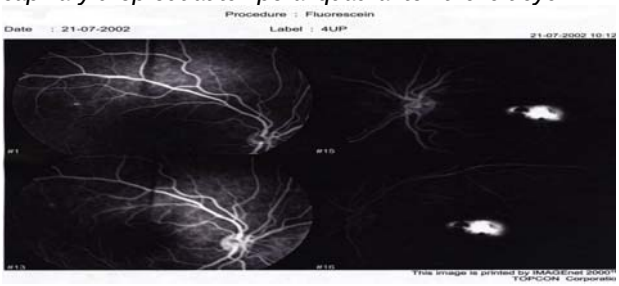

Figure 5.Hyperfluorescent area due to submacular choriodal neovascular membrane of the left eye.

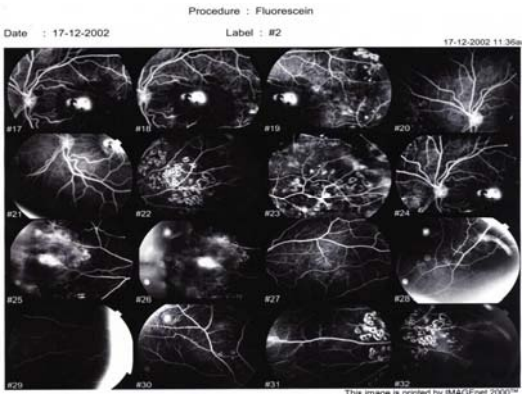

Figure 6: Leaky abnormal vessels mainly at temporal quadrant of the right eye.

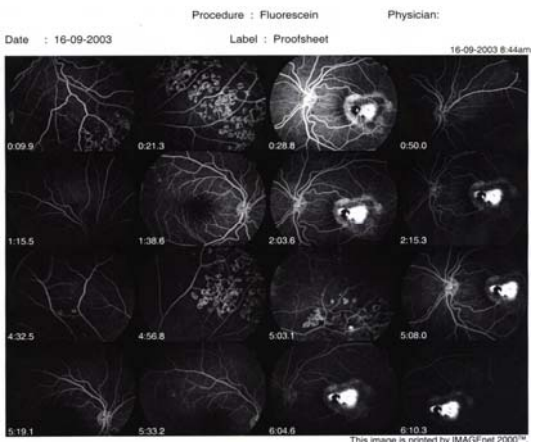

Figure 7: Both eyes showed no leakage, exudation or new abnormal lesions.

Discussion

Coats disease is a rare congenital, but non-hereditary (non familial). It usually occurs unilaterally in young males, and is characterized by idiopathic retinal telangiectasia with intraretinal and / or subretinal exudation, exudative retinal detachment without appreciable retinal or vitreous traction.[1] Coats disease is a predominantly unilateral developmental vasculopathy seen in two forms, the earlier form (<20 years) and the adult form (> 20 years). The disease is observed mostly in males who are younger than 20 years.

The condition is unilateral in 90% of cases and appears to affect males 10 times more than females in the earlier form.[2] The most frequent age at the diagnosis ranges from 3 to 9 years and leukocoria and strabismus are the most common signs.[3] As regards the diagnosis we have made, the presentation of our child with strabismus and later on with leukocoria and the appearance of beaded retinal vessels with subretinal exudates are most likely to be Coats disease, even though she is a girl, and it is bilateral. Coats disease is isolated in the majority of cases, although associations with Focioscapulohumoral muscular dystrophy have been reported.[1] However, this disease entity is not likely in view of her normal muscle strength, and the absence of family history. Other disease entities including retinitis pigmentosa, senior-lokin syndrome, mental retardation, Turner syndrome and the ichthyosis hystrix variant of epidermal naevus syndrome has been also reported.[1, 2, 3]

Pediatrician found no systemic abnormalities in our child. The principal aim and challenge of the differential diagnosis of Coats disease consist in excluding advanced retinoblastoma which is characterized by the triad of retinal detachment, dilated retinal vessels, and appearance of a subretinal mass, requiring enucleation.[4] In one study, one-half of the cases of childhood leukocoria are caused by retinoblastoma and only 16% of leukocoria are caused by Coats disease.[5] Also, strabismus is presenting symptom or sign in about 23% of Coats disease cases,[3, 6] whereas it presents in 20% of retinoblastoma cases.[7] Coats disease is one of the three most common pseudoretinoblastomas.[4] In the vast majority of retinoblastoma cases it has become clinically apparent before the age of 3 years and it has no sexual predilection.[6,7] Birth, medical, and family history would help exclude retinoblastoma.[8] In cases of total retinal detachment, exophytic retinoblastoma is associated with a gray-white material in subretinal space, whereas the retina in Coats disease displays telangiectasia and yellow subretinal fluid. Indirect ophthalmoscopy, fundus photography, and Fluorescein angiography are very helpful in differentiating Coats disease from retinoblastoma.[1] Ocular Ultrasonography and CT scan enable the detection of intraocular masses and/or calcification, which are characteristic features of retinoblastoma.[1]

Ocular ultrasonography and CT scan demonstrated that she didn't have retinal masses, calcification or retinal detachment. Over 90% of advanced retinoblastoms show evidence of calcification on CT scan,[9] whereas it is rarely seen in advanced Coats disease.[10] MR imaging studies can be very helpful in Coats disease. The presence of proteinacceous subretinal fluid in Coats disease leads to production by MR imaging of typical T1- and T2 weighted sequences,[11] whereas retinoblastoma displays hyperdensity on T1 – and proton – weighted MRI and hypodensity on T2- weighted MRI.[2] These MRIs findings were not seen in our patient. Staging of Coats disease can be helpful to select the patients for treatment and expecting the visual outcomes.[1, 12] Stage 1 disease (telangiectasia only): could be treated by periodic observation or laser photocoagulation. This stage is uncommon and the visual prognosis is usually favorable. Stage 2 disease (telangiectasia and exudates): generally managed by laser photocoagulation or cryotherapy. It is also sub- classified into stage 2A and 2B.We classified our patient as stage 2 disease. The Rt. eye showed telangiectasia and exudation without foveal involvement (stage 2A disease). The visual prognosis of stage 2A (Rt. Eye) is generally good and this is evident in our patient because she maintained 6/6 vision through long term follow up. The Lt. eye can be classified into stage 2B disease because she has advanced foveal exudation and submacular CNV. The visual prognosis of the Lt. eye (stage 2B) is poor (6/60). It should be noted that our patient received combined modalities of treatment (laser photocoagulation and cryotherapy) for both eyes. Stage 3A disease (subtotal retinal detachment): can be managed by laser photocoagulation or cryotherapy. It should be noted that laser photocoagulation is less effective than cryotherapy in presence of retinal detachment because of subretinal fluid. Stage 3B disease (total retinal detachment): In presence of shallow retinal detachment, this stage can be managed by cryotherapy. However, if retinal detachment is advanced and immediately posterior to lens, may require an attempt at surgical reattachment. Stage 4 (total retinal detachment with glaucoma): Because of severe eye pain, this stage often requires enucleation. Our patient had normal intraocular pressure. Stage 5 disease: The patients generally have blind comfortable eye and require no aggressive treatment.

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التهاب الجلد المثي: تأثيره على العين والجلد

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ملخص :-

الهدف: دراسة تأثير التهاب الجلد المثي على العين والجلد، و بيان مدى ارتباط شدة تأثر العين بموقع الجلد المصاب.

الطرق: اجريت هذه الدراسة الأستباقية في مدينة الحسين الطبية في الخدمات الطبية الملكية في الفترة ما بين تشرين الثاني من عام 2003 وتشرين الأول من عام 2004. تم تحويل اربع وستين مريضا تم تم تشخيصهم بالأصابة بمرض التهاب الجلد المثي الى عيادة العيون حيث تم اخذ السيرة المرضية من كل مريض متضمنة مدة الاصابة والعلاجات السابقة. اشتمل فحص العين على قياس حدة الأبصار، فحص الحجرة الأمامية بواسطة مصباح شقي، بيان وجود اعتلال القرنية المنقط بعد الفحص بصبغة حيوية، ودراسة السائل الدمعي. تم تسجيل موقع الجلد المصاب و بيان مدى ارتباط شدة تأثر العين به.

النتائج: وجدت اصابات العين لدى 90.6٪ من المرضى حيث كان التهاب الجفن اكثرها شيوعا (84.4٪). تمت اصابة فروة الرأس لدى 71.9٪ من المرضى. اشد اصابات العين وجدت عند اصابة حاجبها بالتهاب الجلد المثي.

الخلاصة: انه لمن المهم فحص العين لدى اي مريض يعاني من التهاب الجلد المثي حيث ان الأغلبية منهم تعاني من اصابة في العين.

الكلمات الدالة: المث، التهاب الجلد، واصابة العين.