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Diagnosis and treatment of bilateral Coats disease in a 5-year-old girl

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A 5-year-old girl presented with decreased vision and outward deviation of her right eye. Fundus examination revealed multiple hard exudates in the macula in the right eye and nasal to the disk in the left eye. The patient was lost to follow-up in the near term but presented 9 months later with reduced vision and an increase in exudates in both eyes. RetCam fluorescein angiography confirmed the diagnosis of bilateral Coats disease.

Coats disease is an isolated nonhereditary retinal vascular anomaly characterized by vascular telangiectasias and abnormally dilated aneurysmal vessels that cause subretinal and intraretinal exudation and exudative retinal detachment.¹ Coats disease is primarily a disease of young male patients with a predominant unilateral presentation.¹⁻⁴ In two large case series from the United States and India, bilaterality was reported in 5% and 10% patients, respectively. Female sex with Coats disease was reported in 24% and 17.2% patients, respectively.^{1,3} A combination of these 2 scenarios is relatively rare. We report a case of bilateral presentation of Coats disease in a girl diagnosed and managed with the aid of RetCam-based fluorescein angiography (FA; Natus Medical Incorporated, Pleasanton, CA).

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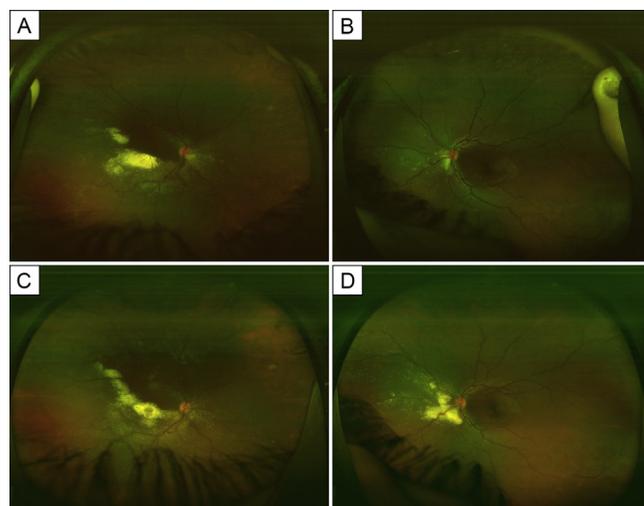


FIG 1. Pseudo-color fundus images of a 5-year-old girl with bilateral Coats disease (Optos 200Tx, Dunfermline, Scotland, UK). A, Right eye fundus at presentation showing deposition of hard exudates at the posterior pole. B, Left eye fundus at presentation showing speckled deposition of hard exudates nasal to the disk. C, Right eye after 9 months showing increase in the hard exudates at the posterior pole with fibrotic focus at the fovea. D, Left eye after 9 months showing increase in the hard exudates nasal to the disk but with sparing of the foveal center.

Case Report

A 5-year-old girl was brought by her parents for evaluation of decreased vision in her right eye of 4 months' duration. There was no associated pain, redness, or photophobia. Past history was significant for outward deviation of her right eye since 2 years of age, but no treatment was sought. The child was born at term with an unremarkable perinatal history. On general physical examination, the child was healthy and had attained appropriate developmental milestones.

At presentation, best-corrected visual acuity was 6/36 in the right eye and 6/6 in the left eye. An exotropia of 60^A was noted in the right eye. Pupillary reactions and anterior segment examination were unremarkable. Posterior segment examination of the right eye showed multiple yellowish hard exudates in the posterior pole, confined to the inferotemporal area of the macula (Figure 1A). The left eye revealed scanty hard exudates nasal to the optic disk (Figure 1B). Widefield FA was planned on the Optos imaging system (Optos 200Tx, Dunfermline, Scotland, UK). The child was uncooperative for imaging, and the parents were counseled on the need for examination under anesthesia; however, they did not consent.

The child was lost to follow-up for 9 months, at which time she presented again with complaints of further deterioration of vision in the right eye. Best-corrected visual acuity was 3/60 in the right eye and 6/9 in the left eye. Fundus examination of the right eye revealed increase in the exudation at the macula and a subfoveal fibrotic scar (Figure 1C). The left eye also showed increase in the exudation nasal to the optic disk (Figure 1D). The

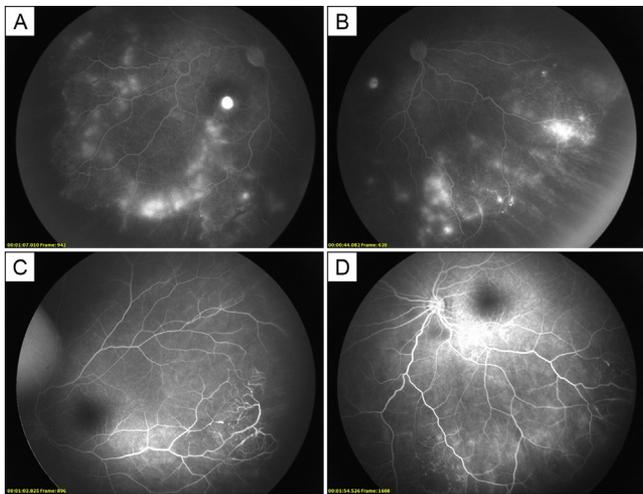


FIG 2. Late-phase fluorescein angiography (RetCam 3; Natus Medical Incorporated, Pleasanton, CA) of a 5-year-old girl with bilateral Coats disease. A-B, In the right eye, there is extensive peripheral capillary nonperfusion areas with retinal telangiectasia in all the quadrants; peripheral leakage is seen in the late phase along with staining of the fibrotic focus at the foveal center. C-D, In the left eye there are areas of retinal vessel telangiectasia and peripheral nonperfusion in the inferotemporal and inferonasal areas.

parents consented to examination under anesthesia along with FA.

Under general anesthesia, sodium fluorescein 20% was injected intravenously and images were captured using the RetCam3. FA of the right eye showed telangiectatic retinal vessels in all four quadrants, with peripheral capillary nonperfusion areas. Late-phase FA images revealed leakage of dye from anomalous vessels and staining of the fibrotic scar at the foveal center (Figure 2A-B). FA images of the left eye showed vascular telangiectasias with capillary nonperfusion areas, limited to the inferotemporal and inferonasal quadrants (Figure 2C-D). A diagnosis of bilateral Coats disease was made: stage 2B in the right eye and stage 2A in the left eye, according to the classification system by Shields and colleagues.⁵ In view of the extensive and progressive nature of the disease, laser photocoagulation was planned. 532 nm laser photocoagulation, using a laser indirect ophthalmoscope delivery system, was performed to the telangiectatic vessels and peripheral capillary nonperfusion areas in both eyes. Systemic evaluation for any signs of cerebral microangiopathy or muscular weakness was negative. At 6 months' follow-up, the visual acuity was stable in both eyes, without any increase in exudation in either eye.

Discussion

This case demonstrates the utility of RetCam-based FA in the diagnosis and management of bilateral Coats disease. Although the diagnosis of Coats disease is mainly clinical, unusual presentations and lack of cooperation in children

can pose challenges. In our case, both bilaterality and female sex were odd for Coats disease, and the differential diagnosis included familial exudative vitreoretinopathy (FEVR) and idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN). FEVR presents with more exuberant retinal exudation, fibrovascular proliferation, and predominant involvement of the temporal retinal periphery. Fundus examination of family members reveals clinical/subclinical features of FEVR. Examination of parents of the girl was normal. IRVAN is characterized by peripapillary retinal arteriolar aneurysmal dilatations with peripheral capillary nonperfusion areas and neuroretinitis. These features were absent on FA. Bilateral Coats-like response can be seen in girls with associated systemic disorders, such as Coats' plus syndrome and facio-scapulo-humeral dystrophy.⁶ However, workup for these was negative in our case.

Coats' disease has been investigated using ultra-widefield FA (UWF-FA) in children. Fluorescein can be administered intravenously or orally in these patients. Tsui and colleagues⁷ demonstrated the utility of UWF-FA after oral fluorescein in children. Blair and colleagues⁸ imaged contralateral eyes of 14 unilateral cases of Coats disease with RetCam FA and found retinal telangiectasias and microaneurysms in 5 cases. Rabiolo and colleagues⁹ demonstrated peripheral retinal lesions in 77.8% of asymptomatic fellow eyes of unilateral cases of Coats disease, on UWF-FA. They proposed that Coats disease might be considered an asymmetric bilateral disease, advancing a two-hit hypothesis to explain the asymmetry.

Young patients with Coats disease who have macular exudates or large areas of retinal detachment tend to have a poor visual prognosis. Therapy is targeted toward preventing total detachment or neovascular glaucoma.⁵ Early stages of disease benefit from FA-guided laser photocoagulation of telangiectasias, as performed in our patient. This case highlights the fact that Coats disease can present atypically in girls, with bilateral ocular involvement. Pediatric patients with retinal exudative pathologies should be examined under anesthesia and imaged with RetCam FA to diagnose and treat these diseases as early as possible.

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Chlamydia trachomatis presenting as preseptal cellulitis in a 3-year-old girl

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Inclusion conjunctivitis usually presents with lid swelling, red eye, foreign body sensation, and a mucopurulent discharge in association with a follicular reaction involving the palpebral and bulbar conjunctiva and semilunar fold. Similar to epidemic keratoconjunctivitis, it may present with preauricular lymphadenopathy, superficial punctate keratitis, and subepithelial corneal infiltrates, which tend to be more peripheral. We present the case of preseptal cellulitis in a 3-year-old child, caused by nonconsensual sexual contact with chlamydia trachomatis.

Case Report

A healthy 3-year-old girl presented emergently at Bronx-Care Health System with worsening left periorbital swelling, redness, and mucopurulent discharge, which had been worsening for 3 days. Review of systems was negative. The child lived at home with her mother, father, and 2 siblings and attended daycare. Visual acuity was not

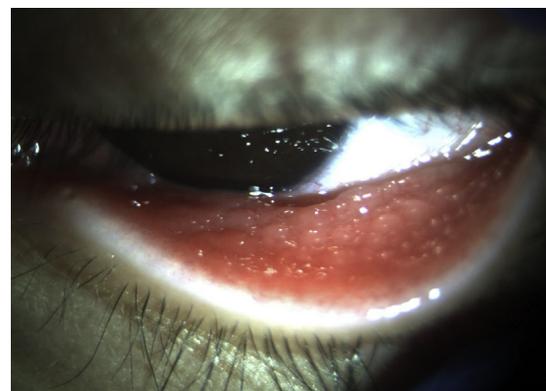


FIG 1. External photograph of the left lower palpebral conjunctiva showing follicular inflammation.

obtainable because of poor cooperation. External ophthalmologic examination revealed a normal right eye; the left eye had marked edema and erythema of the eyelids and a follicular reaction on the inferior tarsal conjunctiva. Because complete ophthalmologic examination was precluded by poor patient cooperation, computed tomography of the face and orbits was obtained to rule out orbital involvement. It showed preseptal swelling on the left, without orbital or sinus inflammation.

A diagnosis of bacterial preseptal cellulitis in association with epidemic keratoconjunctivitis (EKC) was made, and the patient was treated with systemic clindamycin. The following day, the patient was more cooperative, allowing for more complete examination. Uncorrected visual acuity measured 20/20 in each eye, and the eyelid and conjunctival appearance remained unchanged. The rest of the eye examination was unremarkable. Three days later, the child was discharged on a 10-day course of oral cephalexin and topical erythromycin. Two weeks later, the follicular conjunctivitis (Figure 1), lid edema, and erythema were still present, but two midperipheral subepithelial corneal infiltrates were newly noted. As a result, swabs of the conjunctiva were taken and were positive for *Chlamydia trachomatis*, serotype E, as identified by immunofluorescence. Urine *C. trachomatis* ribonucleic acid transcription-mediated amplification testing was also positive. Testing was negative for *Neisseria gonorrhoeae*. Child protection services was notified, and the child was readmitted to the hospital for protection and treatment with oral azithromycin. Her ocular symptoms completely resolved within 1 week.

Discussion

Preseptal cellulitis is common in the pediatric population, and sinus disease has been found to be the most common predisposing factor.¹ *Staphylococcus aureus* and *Streptococcus sp.* are the most common bacterial pathogens causing preseptal cellulitis.^{1,2} Additionally, periorbital changes have been noted to occur with EKC³ and may be misdiagnosed as preseptal cellulitis, which we believe also occurred in our patient with chlamydial conjunctivitis. To our knowledge,

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