Title: Multiple arterial macroaneurysms in acquired Coats' disease in an adult - a case report.
Authors: Strueven V, Schalenbourg A, Wolfensberger TJ
Journal: Klinische Monatsblatter fur Augenheilkunde
Year: 2015 Apr
Volume: 232
Issue: 4
Pages: 583-4
DOI: 10.1055/s-0035-1545786

Multiple arterial macroaneurysms in acquired Coats' disease in an adult – a case report

Erworbener Morbus Coats mit multiplen arteriellen Macroaneurysmata im Erwachsenenalter – ein Fallbeispiel

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Financial interest: NONE

Support: NONE

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Background

Coats' disease is an idiopathic, typically unilateral, retinal vasculopathy that causes diffuse telangiectasia with intraretinal and subretinal exudation, aneurysmal formation, capillary non perfusion and lipid deposition [**1**]. Being mainly a disease of childhood, rare cases can be seen in adults. Despite the less severe symptoms and slower progression [**2**], late onset Coats' disease can cause important loss of vision, if left untreated.
In this article, we present a rare case of Coats' disease, diagnosed in adulthood.

History and signs

A 71-year-old male patient was referred to our clinic in 2011 with multiple unilateral macroaneurysms located in the posterior pole of his left, amblyopic eye, causing metamorphopsia since two months. Rings of circinate lipid deposits surrounded all lesions (**Figure 1**) [**3**]. Best-corrected visual acuity (BCVA) at this first examination was 20/40, which corresponded to the vision prior to the onset of symptoms in this eye.

Therapy and Outcome

A recommended laser treatment of the lesions was unfortunately not realized and the patient was lost to follow-up. He presented again in 2013, because of an important loss of vision in his left eye. BCVA was hand movements and biomicroscopy of the affected eye did not reveal signs of inflammation. The intraocular pressure (IOP) was within normal limits. On fundus examination, massive exudation, emanating from one of the aneurysms and extending into the foveal area, could be observed (**Figure 2**). The ophthalmic exam of the right eye was normal.
After detection of capillary dropout zones in the telangiectasia region on fluorescein angiography (**Figure 3**) [**4**], panretinal argon laser photocoagulation was performed. A persistent macular edema required one intravitreal injection of Bevacizumab (Avastin®) four months later. At his last control visit four months after the injection, BCVA had improved to 20/200. A minimal IOP elevation in the left eye was treated successfully with topical medication. Fundus slit lamp and OCT showed no subretinal exudation, compared to OCT imaging taken four months prior to the injection (**Figure 4 A+B**). Repeated peripheral ultra-

wide field photography confirmed a reduction of the circinate lipid deposits and exudation in the macular area (**Figure 5**).

Conclusion

The present case illustrates the importance of considering Coats' disease in adulthood as a differential diagnosis. In contrast to the typical Coats' disease occurring in childhood, adults are often asymptomatic and present with good visual acuity. Circumscribed lipid deposits and hemorrhages from macroaneurysms can appear in the equatorial and peripheral regions as well as centrally. The lesions in our case induced metamorphopsia as a presenting symptom before reducing central vision at a later stage when exudation extended into the juxtamacular region. This observation underlines the important fact that Coats' disease in adulthood needs to be treated as soon as diagnosed to prevent important long-term visual loss. Furthermore, panretinal laser photocoagulation can prevent bleeding from the typical aneurysms and reduce macular edema as well as exudation.

In the more severe cases, adjuvant medical therapy such as intravitreal steroids or other growth factor inhibitors are effective [**5**], as shown in our case where treating the persistent macular edema with Bevazicumab lead to a rapid reduction of the intraretinal liquid.

In conclusion, we present a rare case of Coats' disease diagnosed in adulthood which suffered visual loss because of delayed laser treatment. Despite the milder presentation and slower progression of the adults' compared to the typical children's Coats' disease, these retinal exudative aneurysms potentially associated with ischemia need to be treated as quickly as possible. Moreover, being a chronic disease, it necessitates a long-term follow-up, even if diagnosed at an advanced age [**6,7**].

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Figures

Figure 1 A.

Wide-angle fundus photography of the left eye taken in 2011 showing several macroaneurysms and circinate lipid deposits.

Figure 1 B.

Wide-angle fundus photography taken in January 2014 depicting increased diffuse telangiectasia and a marked increase in lipid deposition.

Figure 1 C.

Fluorescein angiogram of the left eye performed in January 2014 showing areas of capillary non perfusion in the telangiectatic areas.

Figure 1 D.

OCT cross sectional imaging taken in January 2014 showing marked intraretinal exudation and macular edema.

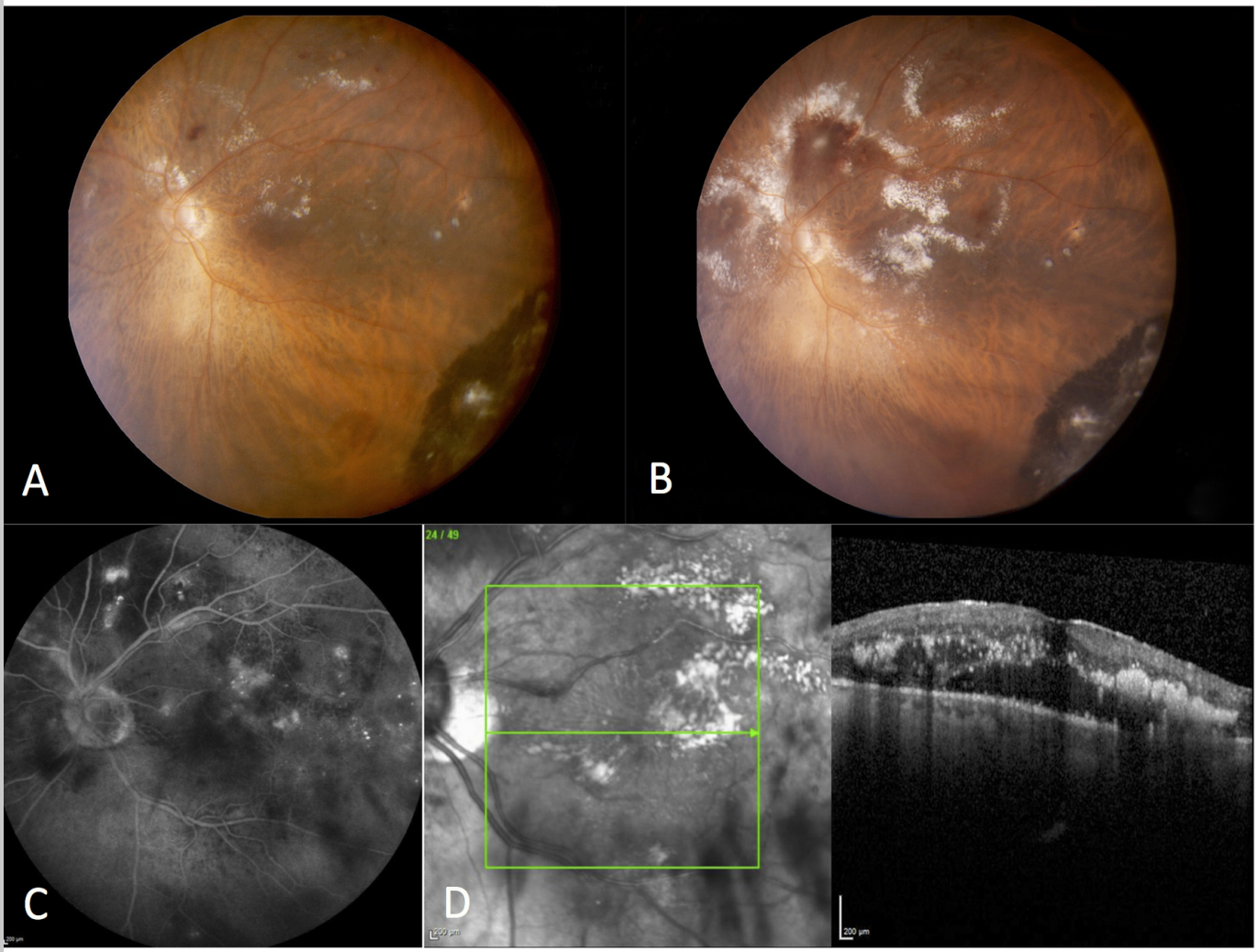
Figure 2 A.

Ultra-wide field fundus photography of the left eye taken in August 2014 showing a reduction of the circinate lipid deposits and exudation both in the macula and in the areas with telangiectasia.

Figure 2 B

OCT cross sectional imaging four months after intravitreal Bevazicumab injection. No intraretinal exudation can be detected.

3. File



4. File