CASE REPORT

Bilateral Neuroretinitis as an Ocular Manifestation of Cat Scratch Disease in a 9-year-old Boy

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SUMMARY

Purpose: 1. To highlight a less-known clinical entity neuroretinitis and the need for differentiation of this entity from the other retinal disease that can mimic. 2. To be familiar with ocular finding in Cat scratch disease.
Material and methods: Case report. Authors describe a clinical course of bilateral neuroretinitis in a 9-year-old boy who was referred to our clinic with painless decreased corrected visual aquity in the right eye (6/18) and in the left eye (6/9). Fundus examination disclosed bilateral stellate maculopathy. Patient had a history of close contact with a cat. Serologic tests for infective disease confirmed the presence of IgG antibody against Bartonella henselae (1:64). Specific antibiotic treatment with bacteriostatical activity against Bartonella henselae restored functional and anatomical changes in both of eyes within two month.
Results: Noninfective etiology of bilateral neuroretinitis was essential to exclude in differential diagnosis. Diagnosis of Cat scratch disease was based on positive epidemiological diagnosis, bilateral manifestation of neuroretinitis, high IgG antibody titre against Bartonella henselae and successful treatment of this disease after specific antibiotic therapy.
Conclusion: Neuroretinitis is the most common ocular manifestation of cat scratch disease. Familiarity with differential diagnosis of neuroretinitis is essential for prompt causal treatment initialisation.

Key words: cat scratch disease, bilateral stellate maculopathy, neuroretinitis, boy

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INTRODUCTION

Cat scratch disease is an inflammatory disorder brought about by the gram negative bacteria Bartonella henselae, which as a rule spontaneously subsides in immunocompetent individuals. It concerns a motionless rod, which is spread worldwide, with a higher prevalence in warm and moist climatic regions [4]. In cats the disease has an asymptomatic course for several months, young cats aged less than one year living outside and flea-infested cats are a particular risk.
Worldwide these bacteria are reported in 15-55 % of healthy cats [4]. In the Czech Republic the number of infected cats living wild is 67 %, whereas only 5% of domestic cats are infected [5].

Fig. 3 Stellate maculopathy of right eye one month after treatment
Fig. 4 Stellate maculopathy of left eye one month after treatment
Fig. 5 Almost complete resorption of stellate maculopathy of right eye 2 months after treatment
Fig. 6 Normal finding in macula of left eye 2 months after treatment

An important vector of mutual transmission between cats is the cat flea Ctenocephalides felis. Bartonella henselae is capable of surviving in flea excrement for several days [4, 3]. During the course of grooming the fur, the claws and mouth of the cat become contaminated by flea excrement, which contains Bartonella henselae. Transmission of Bartonella henselae to humans takes place after scratching or biting by a flea-infested animal. Transmission upon leeching of a flea or tick [5] is also considered a possibility, other carriers may be dogs, monkeys or porcupines [4].
Despite the high percentage of infected cats, the incidence of cat scratch disease is only 4-9 cases per 100 000 of the population, with a high occurrence in persons aged between 2-24 years, with a slight predominance of males. The incubation period is 3-10 days, usually one week.
The pathogenesis of infection with Bartonella henselae depends on the condition of the immunity of the host. In immunocompetent individuals this disease has a benign course and spontaneous healing occurs. A necrotising inflammatory response occurs histopathologically in the place of inoculation in immunocompetent patients. By contrast, in immunodeficient patients a general vasoproliferative response develops, with manifestations of peliosis hepatitis, bacillary angiomatosis and sepsis. In immunodeficient patients the disease has a serious to fatal course, affecting the entire organism.
One week after contamination, primary skin lesions appear in the place of inoculation: indolent macula, papula or vesicle, similar to insect bite. Necrosis occurs in the centre of the skin lesion, concentrically surrounded by layers of histiocytes, lymphocytes and Langhans cells, forming a granuloma [1]. 1-6 weeks after the appearance of the skin granuloma regional lym-

Fig. 1 Initial finding of stellate maculopathy of right eye

Fig. 2 Initial finding of stellate maculopathy of left eye

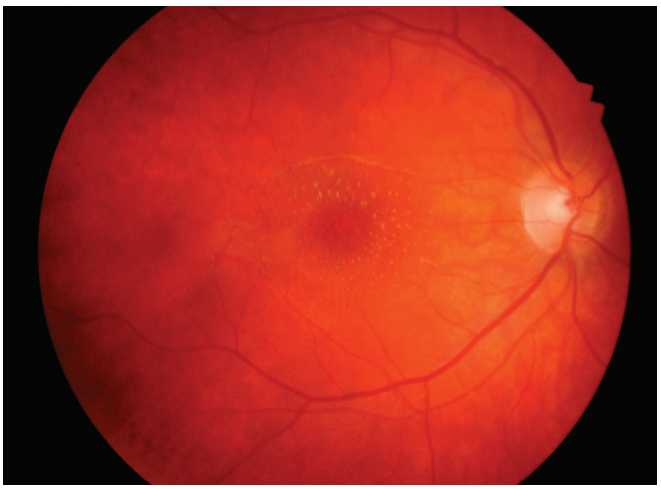

Fig. 3 Initial finding of stellate maculopathy of right eye

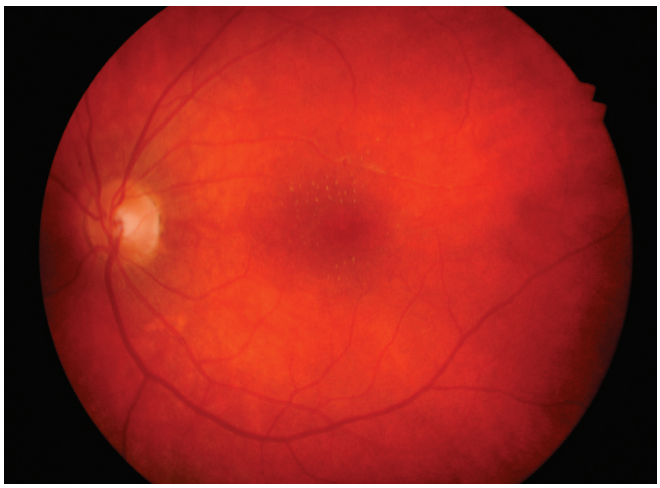

Fig. 4 Initial finding of stellate maculopathy of left eye

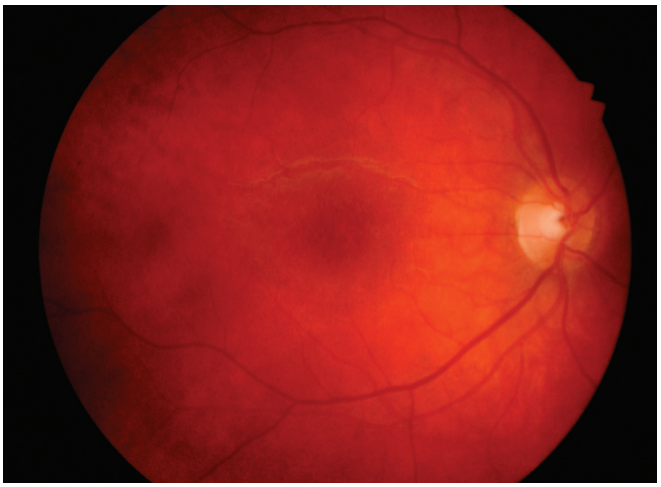

Fig. 5 Initial finding of stellate maculopathy of right eye

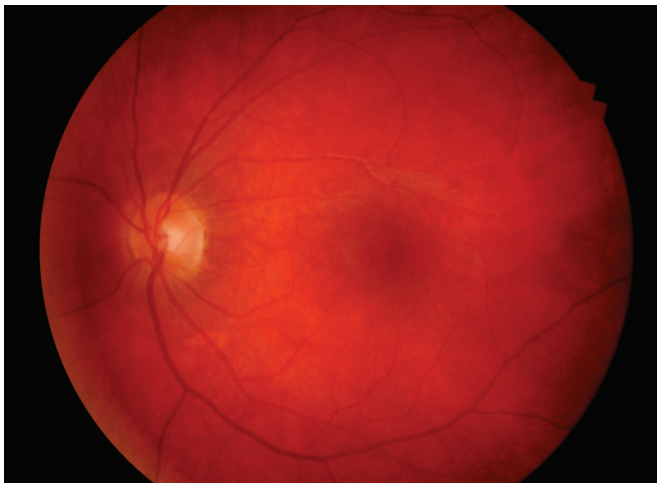

Fig. 6 Initial finding of stellate maculopathy of left eye

phadenitis appears, usually unilateral, affecting one ganglion. Enlarged ganglion, most frequently in the cubital, axillary and cervical area is generally painful, stiff, with surrounding skin erythema, reaching an average size of up to 8 cm. In one half of cases the condition may be accompanied by general symptoms, similar to influenza: fatigue, headache and neck pain, anorexia and subfebrile temperatures. Regional lymphadenitis usually regresses spontaneously within a few months. In rarer cases the ganglion form of cat scratch disease is linked to splenomegaly, pharyngitis and skin complaints – erythema nodosum, erythema multiforme and maculopapular rash on the skin.

The only ocular manifestation of the ganglion form of cat scratch disease is

Parinaud's oculoglandular syndrome. This is unilateral granulomatous conjunctivitis with preauricular lymphadenopathy. The place of entry of the agent is the conjunctiva. Penetration of the infection into the conjunctiva occurs in 5% of symptomatic patients, primarily by means of infected hand or by airborne cat flea excrement [1].

Intraocular affliction occurs in 5 to 10% of patients with cat scratch disease. It develops with hematogenous spread of the infection from the place of inoculation. Bartonella henselae reproduces on the surface of the endothelial cells and entrocytes [1], by which it stimulates local thrombogenic mediators. The most frequent ocular manifestation of this disease is neuroretinitis [1, 7]. Less occurring manifestations include neuritis, retinitis, choroiditis, serous retinal detachment, vasculitis, occlusion of the retinal vein/artery arm, peripapillary angiomatosis and intermediate uveitis [1, 7]. This disseminated form of cat scratch disease may be complicated by:

1. Neurological affliction – encephalopathy, transient paresis of facial nerve, myelitis.
2. Hepatosplenenic form – granolomatous hepatitis, abscesses of the spleen.
3. Haematological disorders – haemolytic anaemia, thrombocytopenic purpura, eosinophilia.
4. Affliction of endocardium.

The case study refers to a rare case of bilateral neuroretinitis in a nine-year-old boy with positive serological examination of antibodies against Bartonella henselae. The aim of the case study is to draw attention to the need to differentiate this disease from other forms of neuroretinitis and other retinal disorders which it may imitate.

CASE STUDY

In April 2012 a nine-year-old boy was sent to us from the local eye department due to suspicion of bilateral intermediate uveitis. The previously healthy boy complained of a progressive, painless deterioration of corrected visual acuity (CVA) in both eyes, persisting for 14 days. Even despite corrected astigmatism he stated blurred vision especially in the right eye. He had last been afflicted by a tick in summer 2011 without any skin reaction in the place of leeching, and during regular visits to his grandmother had played with her two cats. Upon an ophthalmological examination, CVA in the right eye was 6/18 and in the left eye 6/9. Examination of the anterior segment and intraocular pressure were within the range of the norm in both eyes. Indirect ophthalmoscopy of both eyes detected the presence of yellow, hard exudates of a stellate character in the macula, with a more pronounced finding in the right eye (fig. 1, fig. 2). The disc of the optic nerve and the surrounding retina were without pathologies. From the conducted examinations of the serum positivity of IgG antibodies against Bartonella henselae (1:64) and Mycoplasma pneumoniae was determined. IgM antibodies against these bacteria and other immunological, bacteriological, virological and biochemical examinations of serum were negative. Blood pressure was 115/65 torr. A paediatric examination did not confirm the presence of clinical symptoms of cat scratch disease, and the paediatrician did not indicate further examinations due to the normal immunological profile of the patient. General oral treatment was commenced with clarithromycin 500 mg every 12 hours for a period of 14 days. One month after the beginning of treatment CVA in the right eye had improved to 6/12 and CVA in the left eye to 6/6, with pronounced regression of stellate maculopathy bilaterally (fig. 3, fig. 4). One month later CVA in the right eye improved to 6/12 and CVA in the left eye 6/6. The finding of stellate maculopathy in the right eye had virtually disappeared, in the left eye there was now a normal macular finding (fig. 5, fig. 6).

DISCUSSION

Neuroretinitis is an intraocular inflammation, manifested in the form of a painless deterioration of visual acuity, oedema of the disc of the optic nerve, frequently linked to exudative retinal detachment in the peripapillary area and macular lipid exudates forming in the shape of a star. There is also a frequent cellular reaction in the vitreous body together with numerous focal yellowish-white retinal lesions. Despite the fact that there are lots of pathogens which cause neuroretinits (table 1), the clinical course of the disease is surprisingly similar. More than two thirds of patients with neuroretinitis have:

1. Prodromes similar to influenza.
2. Age within the range of 9 to 55 without gender predilection.
3. CVA fluctuates during the period of presentation of the disease from 6/12 to 6/60.
4. Afferent pupillary defect is often present.
5. There is generally centrocecal or central defect of the visual field in the perimeter.

Edema of the disc of the optic nerve occurs approximately one week before the manifestation of stellate maculopathy, and begins to regress 2 weeks after the onset of the complaints, with complete restitution after 8 to 12 weeks. Stellate maculopathy is not always expressed during the period of initial complaints, but may however persist for up to one year [6]. In our nine-year-old patient, probably due to his good immunological condition and due to the longer course of the disease, we recorded only lipid exudates of a stellate shape in the area of the deeper layers of the macula. The diagnosis of cat scratch disease was stipulated on the basis of the following findings.

1. Epidemiological anamnesis confirmed contact with cat.
2. Finding of neuroretinitis is suspect due to infectious disease with neuro-ophthalmological complications.
3. Examination of serum confirmed positivity of IgG antibodies against Bartonella henselae.
4. Anatomical and functional restitution of ophthalmological finding after treatment with antibiotics, bacteriostatic for Bartonella henselae.

After a probable cat scratch during play, in our nine-year-old patient spread of Bartonella henselae very probably occurred via blood, without manifestations of ganglion syndrome.

Table 1 Infection etiology of neuroretinitis

Infection:
1. Bacteria – Cat scratch disease
Syphilis
Lyme borreliosis
Tuberculosis
Actinomycosis
2. Viruses – Influenza (Orthmyxoviridae)
Rubella
Measles
Mumps
Chickenpox
Infectious mononucleosis
Herpes simplex
3. Mycoses – Coccidiomycosis
4. Tapeworms – Toxocara
5. Protozoa – Leptospirosis
Giardiasis (Lambliasis)
6. Chlamydia – Psitacosis

Table 2 Other etiologies of neuroretinitis

Vascular
- Anterior ischemic neuropathy of optic nerve
- Venous occlusion
- Systemic hypertension
- Diabetic retinopathy
- Papillophlebitis

Various
- Behcet's disease
- Sarcoidosis
- Juxtapapillary capillary haemangioma
- Non-specific uveitis
- Papilledema
- Trauma

Serological examinations for Bartonella henselae do not confirm an active disease [8] in up to 70% of cases. The incidence of positive serology for Bartonella henselae in the healthy population is very low, less than 3% [8]. As a result serological positivity is sufficient for stipulation of a diagnosis of cat scratch disease upon positive epidemiological anamnesis and upon positive ocular and systemic symptoms of the disease, also for an ongoing complaint, as in our case study.

The treatment of cat scratch disease remains controversial. It is assumed that this concerns a benign disorder, which completely subsides without treatment. In our case we additionally located the disease in an advanced phase of development, in which only stellate maculopathy was present bilaterally. Despite this fact, we are of the opinion that targeted antibiotic treatment shortens the course of the disease, accelerates restitution of visual functions and prevents the possible recurrence of the illness. The most effective antibiotic therapies against Bartonella henselae are macrolides (erythromycin, clarithromycin, azithromycin), doxycycline, rifampin, trimethoprim sulfamethoxazole [1, 6, 7, 8]. Due to good penetration into the tissue, support for function of phagocytising leucocytes and minimal side effects on the child's organism, we chose 14-day treatment with clarithromycin.

The most frequent infectious cause of neuroretinitis in childhood age is cat scratch disease [6]. We have already described bilateral retinitis in an eight-year-old girl as a manifestation of Lyme borreliosis [2]. Oedema of the disc of the optic nerve, together with oedema of the adjacent neuroretinal tissue and exudation in the macula in the shape of a star may however in childhood age be imitated by non-infectious diseases (Table 2). We consider the possibility of non-infectious etiology in the case of an absence of positive epidemiological anamnesis, the presence of atypical symptoms, in particular the presentation of a bilateral finding, in the absence of a cellular reaction in the vitreous body and if progressive renewal of CVA does not occur.

CONCLUSION

The most frequent ophthalmological manifestation of cat scratch disease is neuroretinitis. Differential diagnosis of neuroretinitis is extensive and arduous. Despite this fact, the majority of etiological units causing or imitating neuroretinitis are treatable. A correct stipulation of the cause of cases of neuroretinitis requires precise taking of epidemiological anamnesis, eye and general examination and performance of serological tests in the diagnosis of the infection etiology.

THANKS

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CASE REPORT

Retinitis Pigmentosa Mimicking Uveitis. A Case Report

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SUMMARY

Purpose: To describe a case report of a 23-year-old patient with retinitis pigmentosa (RP) misdiagnosed as uveitis.
Methods: A comprehensive eye examination including automated visual field assessment, contrast sensitivity, colour vision discrimination, ultrasound examination (US), spectral domain optical coherence tomography (SD-OCT) and full-field electroretinography (ERG) was performed in a patient diagnosed elsewhere as having intermediate uveitis because of the observation of a cellular reaction in the anterior chamber, bilateral cystoid macular oedema and suspected left optic disc swelling.
Results: The patient reported nyctalopia. The best corrected visual acuity in both eyes was 6/12. Concentric visual field constriction was detected bilaterally (less than 25 degrees in the right eye and 15 degrees in the left eye). Fundus examination revealed a few pigment clumps and cystoid macular edema in both eyes confirmed by SD-OCT. Contrast sensitivity was decreased to 1,20 in the right and 0,9 in the left. No colour vision disturbance was present. The B scan ultrasound showed left optic disc drusen. Rod ERG responses were bilaterally not detectable and cone ERGs were abnormally reduced. Based on the examination results, a diagnosis of nonsyndromic RP was made.
Conclusion: Clinicians should be aware of various manifestations of RP, including mild inflammation, to avoid possible confusin with uveitis.

Key words: benign masquerade syndrome, retinitis pigmentosa

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INTRODUCTION

Masquerade syndromes (MS) are non-inflammatory eye disorders which may be clinically manifested as uveitis. MS are divided into malignant, which threaten the sight and even life of the patient, and benign – e.g. capillary anomaly, Schwartz's syndrome, intraocular body, pigment dispersion syndrome, retinal dystrophy and degeneration, ocular ischemic syndrome, juvenile xanthogranuloma, choroidal osteoma, post-medication and post-vaccination uveitis [13, 24]. Nussemblatt et al. also classify chronic postoperative infection as a benign masquerade syndrome [22].

CASE STUDY AND RESULTS

In October 2010 a 23 year old female patient was sent to the Centre for Diagnosis and Treatment of Uveitis of the 1st Medical Faculty of Charles University and the General University Hospital in Prague due to suspected intermediate uveitis most probably of a parasitic type. The reason for the suspicion of this disease was a three--week residence in Thailand approximately one year before the manifestation of the disorder.
From the personal anamnesis we determined that the patient was overall in good health. Apart from cataract and glaucoma in her grandmother, no other ocular disorders occurred in her family. Of more interest is the ocular anamnesis of the patient. She states that since childhood she had not seen as well as her peers (e.g. she had never seen the stars), that she suffered from nyctalopia and perceives a constricted visual field (she likens the conditions to blinkers on a horse). The patient is being observed at the outpatient eye clinic in the locality of her place of residence, from where she was sent to a private centre for a surgical solution due to a finding of a posterior subcapsular cataract. In September 2010 the patient underwent a cataract operation on her left eye (LE) at this workplace, with implantation of a monofocal intraocular lens (IOL). According to the documentation the finding was complicated in the postoperative period by cystoid macular oedema (CME) and oedema of the papilla of the disc of the optic nerve. 2 weeks after the operation betamethasone was applied to the patient by the parabulbar method, Nd-YAG laser capsulotomy of the LE was performed and a full examination by a general practitioner recommended, with consultation at our uveal outpatient clinic.
Upon the first examination at our outpatient clinic, central visual acuity (CVA) of the right eye (RE) was 6/12 with -0.5 cyl ax 180°, J. no. 1, and LE 6/12 with +3.05 Dsf = -2.5 Dcyl ax 180°, J. no. 3 with addition +3.0 Dsf, intraocular pressure by applanation 9 torrs bilaterally. On the passive anterior segment of the RE we recorded a posterior subcapsular cataract (fig. 1a), on the LE there were perceptible cells in the anterior chamber 1+, flare +/-, IOL in situ and fenestrum in the posterior capsule after capsulotomy (fig. 1b). On the fundus of the RE there was a pink papilla with a yellowish halo, most pronounced in the upper quadrant of the disc, macula with oedema, constricted retinal artery and in the periphery isolated pigment clusters (fig. 2a). On the fundus of the LE the finding was similar, only that there was also a present clump on the disc of the optic nerve (fig. 2b), which we elucidated by ultrasound examination (US – fig. 3). We verified oedema in the macula (fig. 4) by optical coheren-

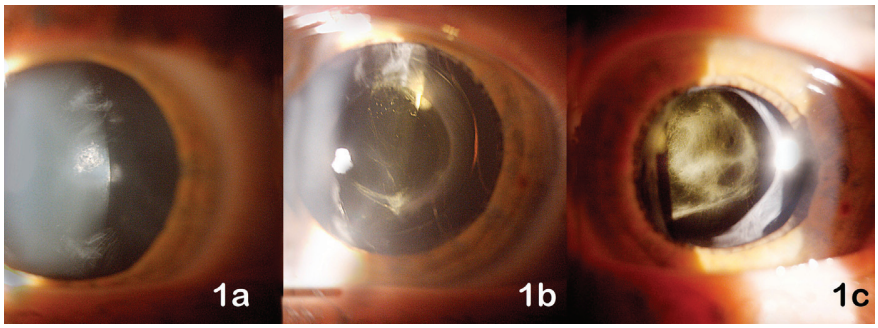


Fig. 1

ce tomography with spectral domain (SD-OCT, Spectral OCT/SLO, OTI Ophthalmic Technologies Inc., Canada). The examination of the visual field (fig. 5) demonstrated bilateral concentric constriction (RE to 25°, in LE to 15°). Contrast sensitivity on a Pelli-Robson table was bilaterally reduced: RE 1.20, LE 0.9. Colour sensitivity (15-Hue test) was within the norm.

We contacted the attending ophthalmologist who sent the patient for the cataract operation. According to her documentation, the constricting of the visual field had been present for a longer time. The patient had undergone a neurological examination due to a suspected finding on the discs in order to eliminate demyelinating central nervous system disorder, the result of which was negative, including electroencephalography.

We recommended an electroreginographic examination (ERG – Ratiscan KL95/Roland konsult, Germany) for the patient, which demonstrated areactive scotopic phenomena and photopic and flicker phenomena on the boundary of reactivity (fig. 6), and confirmed a working diagnosis of retinitis pigmentosa (RP). We applied local and general antiedematous therapy (indometacin gtt. 4xd, dexamethasone gtt. 4xd, escinum alfa tbl. 3x2), and with regard to iatrogenic anisometropia and astigmatism we trained the patient in wearing toric contact lenses. After 3 months there was a deterioration of CVA in RE to 6/18 with +3.5 Dsf = 1.5 Dcyl ax 180°, J. no. 4 with addition +3.0 D (with difficulty), intraocular pressure by applanation 9 torr. On the passive anterior segment there was a perceptible thickened membrane behind the IOL (fig. 1c). According to the finding on the control SD-OCT we did not influence CME LRE (left and right eye) by conservative therapy. As a result Nd-YAG membranotomy was performed on the patient, after which LEV (visus of the left eye) was improved to 6/12 (+2) with correction, J. no. 2 with addition +3.0 (with difficulty). There was no worsening of the finding in the macula of the LE after the laser procedure. With an interval of one year after the operation, vision in both eyes was unchanged, although there was a further constriction of the visual field (fig. 5b – RE to 20°, temporally and downwards to 15°, LE to 10°only nasally to 15°). We supplemented an eye examination of the patient's sister (including perimetric examination and ERG), which was within the norm.

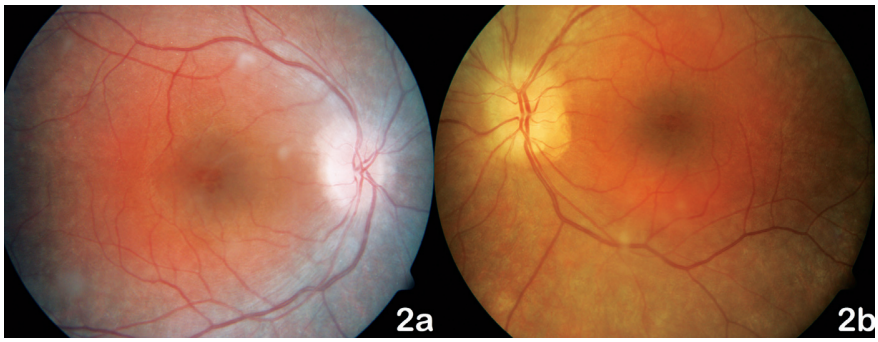


Fig. 2

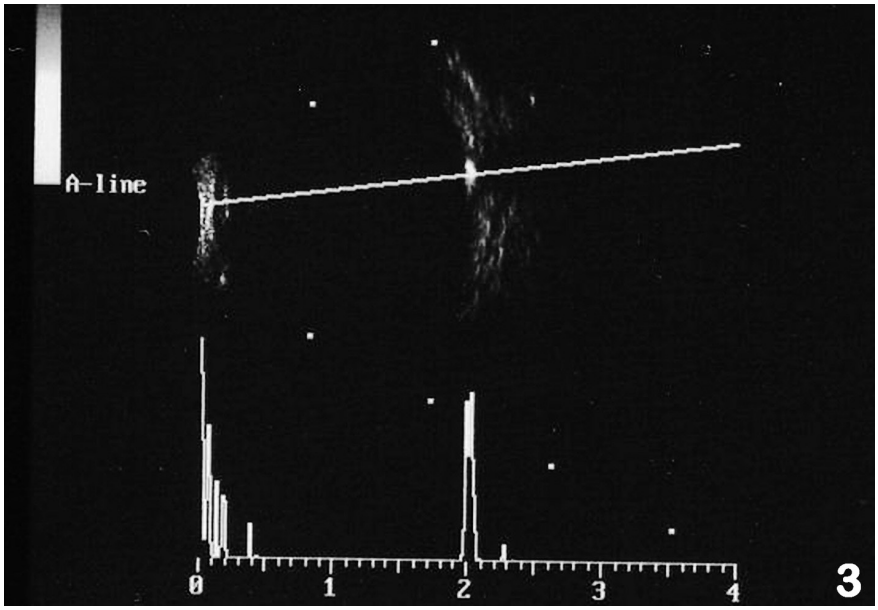


Fig. 3

DISCUSSION

RP is a genetic, heterogeneous group of progressive disorders. In the case of confusion of the clinical manifestations of RP for uveitis, we classify it amongst benign masquerade syndromes.

With regard to uveitis and CME, postoperative complication following extraction of the cataract also came into consideration in the differential diagnostic of our patient. This however does not explain CME in the second, non-operated eye. Pigment paravenous retinochoroidal atrophy could also be considered, though this is an illness acquired following a suffered infection (e.g. after meningoencephalitis, tuberculosis, syphilis or rubella), which the patient negated. A clinical picture similar to the finding of our patient could also be caused by innate pigmentation of the retina with an image of bear traces on the fundus, which is caused by hypertrophy of the retinal pigment epithelium, although neither the fining on the perimeter nor the patient's vision corresponded to this disorder. One of the

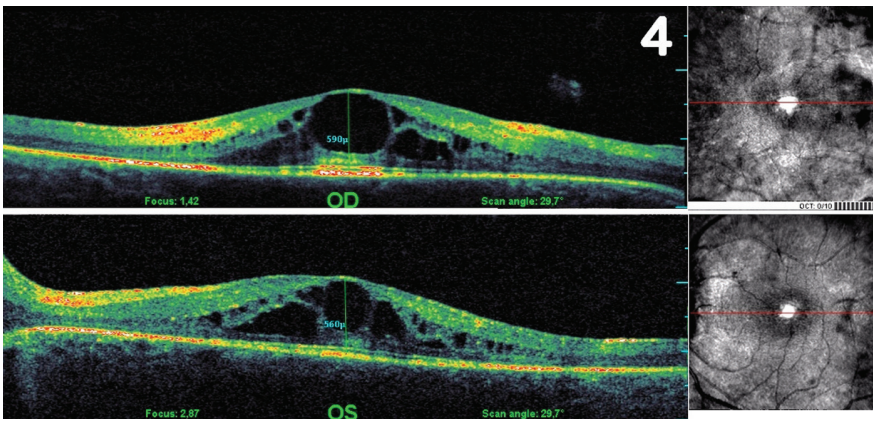


Fig. 4

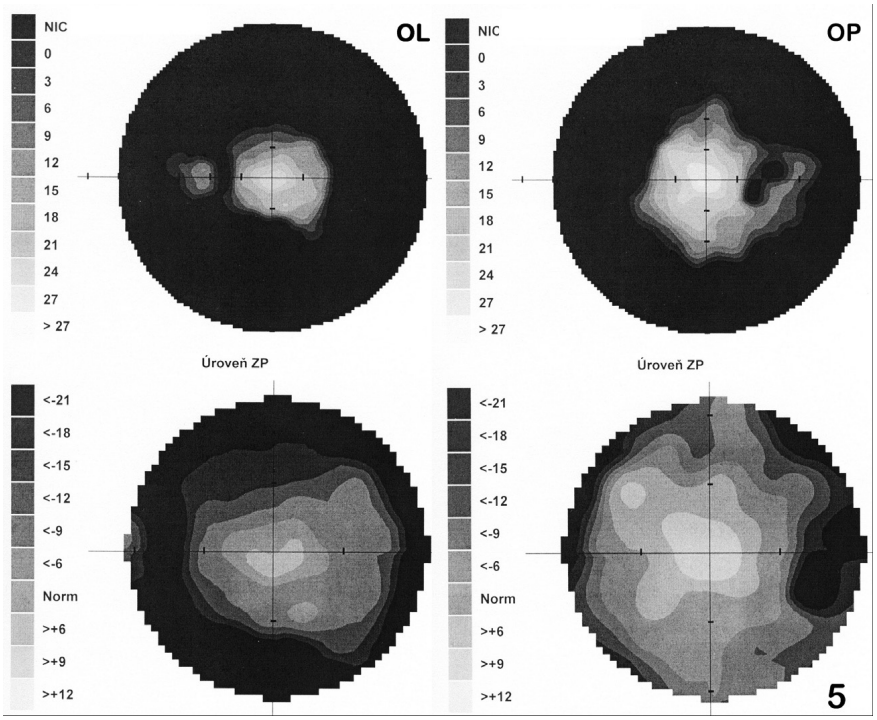


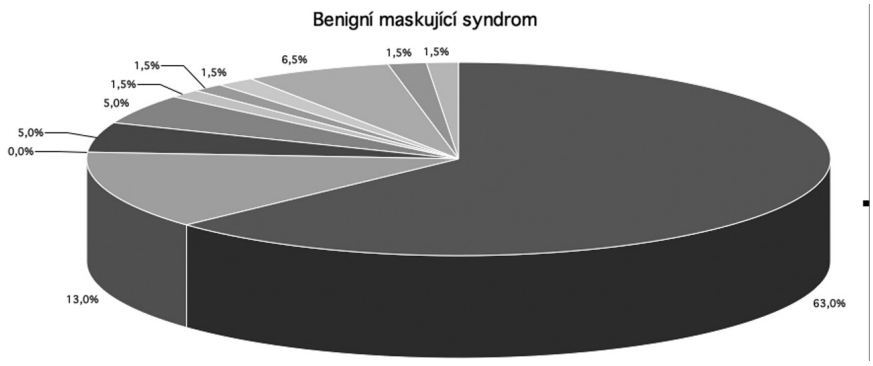
Fig. 5

most frequent causes of pigmentation on the fundus is traumatic retinopathy, in which generalised loss of RPE and migration of melanin to the retinal layers may occur. However, the finding is exclusively unilateral and the patient negated any trauma. Autoimmune paraneoplastic retinopathy as an ocular manifestation of a remote tumour, most frequently small-cell carcinoma of the lungs or carcinoma of the cervix, is also manifested as nyctalopia and constricted visual field, but has a far more rapid progression and occurs in older patients. Toxic retinal degeneration following ingestion of chloroquine, chlorpromazine or phenothiazine was excluded with regard to the negative anamnesis. Similar pigment clusters on the retina as are present in RP may be generated also by congenital rubella (mostly in connection with deafness), measles or hereditary syphilis [14, 12, 23]. All was anamnestically excluded in our patient.

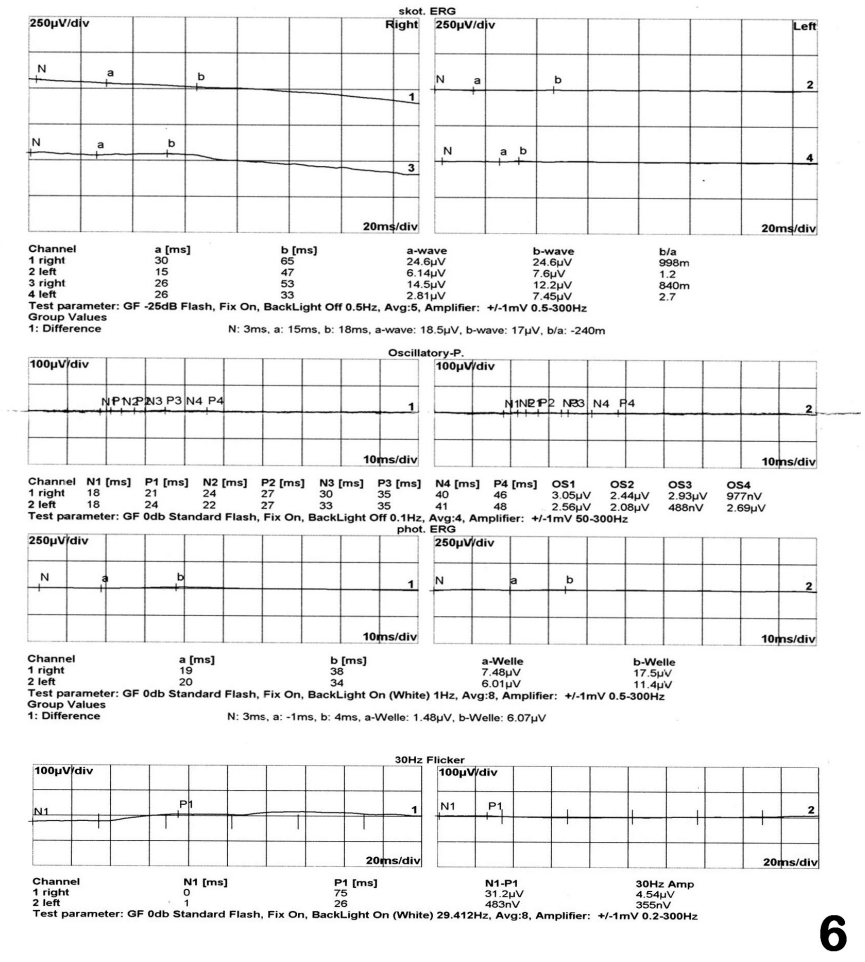
In RP a dominant symptom is degeneration of the rods, which is mostly followed by degeneration of the cones and may lead to total blindness. A typical feature is the presence of "bone cells", which may however be absent in the early stages. The disorder has considerable phenotype variability (unilateral RP, sector RP, RP sine pigmento, retinopathia punctata albescens, cone-rod dystrophy) [10]. RP is divided into syndrome (primarily Usher syndrome) and non-syndrome. The prevalence of the most frequent non-syndrome RP in the regular population is estimated at 1:4000. The disorder is manifested in progressive nyctalopia and constriction of the visual field. In later phases patients complain also of deteriorated CVA. The cause of the deterioration of vision may be, similarly to our patient, the development of a subcapsular cataract (in 35-50%) or CME (in 15 to 23%), as well as the presence of macular atrophy (43%), and less frequently epiretinal membrane [9].

Objectively in patients we see a passive anterior segment, there may be present opacification beneath the posterior lens capsule. The vitreous area is generally clear, the papilla waxy white, in earlier phases it may also be pink, without border, with a yellowish halo in the surrounding area. In 10% of patients there are perceptible clusters of the disc of the optic nerve, as we also identified in our patient [11]. The macula remains intact for a long time, later the changes described above occur. The capillaries are constricted, straightened and from the central periphery pigment clusters (bone cells) are generally perceptible. In rare cases there may be present neovascularisations of the disc of the optic nerve or retina.

We verify the diagnosis using an ERG examination, which may demonstrate a pathological finding even upon an entirely negative finding on the fundus and perimeter. An integral component of the diagnostics is an examination



Graph. 1



6

Fig. 6

of the related proband, which we conducted on the sister of the patient. If circumstances allow, a genetic examination is also appropriate.

Despite the fact that there are a range of clinical trials conducted on patients with degenerative retinal disorders, effective treatment of RP in practice does not yet exist. All we can currently do for patients is to recommend professional rehabilitation services for persons with afflicted vision in a timely manner (thus in the case of sufficiently preserved CVA). Sighted patients more quickly learn everyday tasks which they must perform blind. When the moment of blindness arrives, they are capable of taking care of themselves.

Table 1 Table of patients with benign masquerade syndrome

Diagnosis	Number of patients	%
Morbus coats, capillary anomaly	39	63.0
Schwartz's syndrome	8	13.0
Intraocular foreign body	0	0.0
Pigment dispersion	3	5.0
Retinal degeneration and dystrophy (e.g. RP)	3	5.0
Ocular ischemic syndrome	1	1.5
Juvenile xanthogranuloma	1	1.5
Choroidal osteoma	1	1.5
Post-medication, post-vaccination reaction	4	6.5
Chronic Postoperation Endoftalmitis	1	1,5
Contusion (masquerade toxoplasmosis)	1	1.5
Total	62	100

ves.

The National Eye Institute of the USA recommends vitamin A for slowing the progression. However, because the evaluation of patients in this therapy did not produce an unequivocal result from the perspective of certain other independent experts, this recommendation is not universally accepted. Partial clinical trials have further been conducted on the effectiveness of docosahexaenoic acid, omega-3 fatty acids and lutein, but have not reached entirely clear conclusions [4, 5, 6, 7, 18, 21].

There is therefore no causal treatment of RP in practice. Great hopes have been placed in gene therapy [19], but for successful treatment it is necessary to preserve at least a residue of the patient's visual functions [25]. A pilot study was conducted at the university in Tübingen with implantation of retinal microchips containing photosensitive diodes (up to 1500), which are capable stimulating the inner part of the retina (ganglion cells) and transmitting a signal to the visual cortex [8, 19]. At John Hopkins University in Baltimore, 21 patients with RP took part in a study with the epiretinal prosthesis Arcus II (Arcus II, Second Sight Medical Products, Sylmar, California). None of the patients had better central visual acuity than light sensitivity. The study demonstrated an improvement in the fine motor skills of the arm under visual control [3]. The use of stem cells is also in the stage of laboratory research. Unlike gene therapy, this provides hope for already blind patients [9, 19]. Another new feature is neurotropic factors (CNF), which slow down the degeneration of rods and thereby help the preservation of vision [9, 16, 19]. These are being applied intravitreally or subretinally so far only experimentally on mice.

Nagpal et al. [20] analysed the percentage representation of individual eye diseases which were primarily diagnosed as posterior uveitis. From a total number of 32 patients with benign masquerade syndrome, RP was found in 3 patients (9.3%). First of all healed chorioretinitis was diagnosed in these 3 patients (due to finding of pigment scars on the retina). The age was within the range of 7 and 55 years, and central visual acuity was 6/12 to counting fingers in front of the eye. All suffered nyctalopia, with positive ERG finding. In one patient there was present pigmented paravenous atrophy, which may imitate pigmented

healed scars following chorioretinitis. In our group of patients with uveitis we diagnosed benign masquerade syndrome in 62 patients, in whom a capillary anomaly appeared in 63%, Schwartz's syndrome in 13% and post-medication and post-vaccination uveitis in 6.5%. 5% of patients suffered from pigment dispersion, a further 5% from dystrophy and degeneration of the retina, the remaining 7.5% are equally divided between patients with ocular ischemic syndrome, patients following contusion of the eyeball, patients with juvenile xanthogranuloma, osteoma or postoperative chronic endophthalmitis (non-published data, see graph, table).

There is also a possible coincidence of RP with uveitis: with Fuchs heterochromic iridocyclitis [15, 17], Behcet's disease [1] or Coats disease [2]. These however are rare findings.

Willermain et al. published a case of a patient with birdshot chorioretinopathy HLA-A29 positive, in whom a rarely described hyperpigmentation of cream lesions imitating the finding of bone cells in RP occurred after 6 years of treatment with immunosuppressants [26].

CONCLUSION

We classify RP erroneously considered uveitis amongst benign masquerade syndromes. Deteriorated CVA, constriction of the visual field and nyctalopia may occur in the patient, as was the case in our patient. A finding of bilateral subcapsular cataract, macular oedema, straightened capillaries and concentric constriction of the visual field supports this diagnosis. For final confirmation it is appropriate to supplement ERG or molecular-genetic examination detecting causal mutation.

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