CLINICAL SYMPTOMS AND COMPLICATIONS OF PARS PLANITIS IN CHILDHOOD

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ABSTRACT

Pars planitis is the idiopathic form of intermediate uveitis; it most commonly affects persons between 5 and 40 years of age. In childhood, the disease is largely asymptomatic until in a well advanced stage with markedly reduced vision. Lack of early diagnosis and treatment may lead to permanent visual loss and amblyopia.

Aim: To report on the clinical symptoms, complications, treatment and visual outcome of pars planitis in childhood.

Materials and methods: Ten patients (8 boys and 2 girls) all younger than 16 years were diagnosed with and treated for pars planitis at our clinic between January 1993 and August 2004.

RÉSUMÉ

La pars planite est la forme idiopathique de l'uvéite intermédiaire et affecte le plus souvent les personnes de 5 à 40 ans. Dans l'enfance, cette maladie est la plupart du temps asymptomatique jusqu'à un stade bien evolué où la vision est réduite d'une façon importante. L'absence de diagnostic et de traitement précoce peut mener à une perte permanente de la vision et à l'amblyopie.

But: Nous décrivons les signes, les complications et le traitement de la pars planite dans l'enfance.

Matériels et methodes: Dix patients (8 garçons et 2 filles) tous plus jeune que 16 ans, qui se sont présentés dans notre clinique avec une pars planite entre Janvier à 1993 et Août 2004.

KEY WORDS

Pars planitis, childhood.

MOTS-CLÉS:

Pars planite, enfance.

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INTRODUCTION

Pars planitis is an idiopathic chronic inflammatory disorder affecting the vitreous base, peripheral retina, and ciliary body. It leads to cells and debris in the vitreous with peripheral retinal phlebitis and vitreous 'snowbank' or 'snowball' formation over the pars plana and the peripheral retina (1,2,5,8).

This condition generally affects young patients (between 5 and 40 years of age) and can cause visual loss due to vitreous involvement and cystoid macular oedema. Glaucoma, cataract, epiretinal membrane formation with retinal traction, retinal neovascularisation, vitreal haemorrhage, retinal detachment, rubeosis iridis, cyclitic membrane formation and phthysis bulbi have been described (2,5,8). Sarcoidosis, multiple sclerosis, syphilis, Lyme disease and toxocariasis should be considered in the differential diagnosis.

We emphasise the need for regular follow-up and will discuss the treatment modalities in this patient population.

PATIENTS AND METHODS

We retrospectively reviewed the charts of ten patients (8 boys and 2 girls) all younger than 16 years at the time of diagnosis between January 1993 and August 2004 at the Department of Ophthalmology of the University Hospitals Leuven, Belgium.

At first examination, we recorded age of onset, gender, visual acuity, clinical ophthalmological findings and severity and type of complications. All patients were referred to the Paediatric Department to exclude underlying systemic or infectious diseases.

The diagnosis of pars planitis was based on the observation of snowballs at the pars plana region, anterior vitreous inflammation, cystoid macular oedema, retinal perivasculitis and the absence of an etiologic diagnosis.

A follow-up was planned at short intervals (4-5 weeks) with a complete ophthalmologic examination including best-corrected visual acuity, biomicroscopy, tonometry and fundoscopy. Fluorescein angiography was performed when macular oedema was confirmed on Optical Coherence Tomography (OCT).

During the follow-up period, complications were registered and treated when indicated.

The corticosteroid treatment dose and mode of administration (topically, subconjunctival or systemic) were noted at each visit.

Surgical procedures including lens extraction, strabismus surgery, anterior vitrectomy, cryotherapy and laser photocoagulation were recorded.

RESULTS

Twenty eyes of 10 patients were included in this study.

There were two girls, age 4 and 14 years. The mean age for boys at the time of first diagnosis was 7,7 +/- 2,95 years (range, 4 to 14 years), the overall mean age being 7,38 +/- 1,85 years (range, 4 to 9 years).

Mean follow-up time was 51 months (range, 6 to 132 months).

The presenting symptom in all cases was a decreased visual acuity, detected at a visual screening at school or noticed by teachers or parents. In 3 children there was blinking and rubbing of the eye. One child presented with intermittent convergent strabismus and diplopia.

At diagnosis, the visual acuity (VA) was 20/25 or better in 4 eyes (20%), between 20/100 and 20/25 in 11 eyes (55%), between 20/200 and 20/100 in 4 eyes (20%) and one eye (5%) had a VA of counting fingers.

Biomicroscopy upon the first visit revealed keratic precipitates (4 eyes), anterior chamber cells (10 eyes), cataract (4 eyes), posterior synechiae (4 eyes), vitreous cells in all cases and vitreal veils in 14 eyes. Binocular ophthalmoscopy revealed optic disc hyperaemia (8 eyes), optic disc oedema (6 eyes), macular oedema (13 eyes), periphlebitis (8 eyes) and the presence of snowballs and snowbanking (18 eyes). The ocular involvement was bilateral and asymmetrical in all cases. (Table 1)

Screening for aetiological factors was negative, except for one case with isolated HLA B27 positivity (patient 3). There were, however, no associated systemic symptoms.

At follow-up, the following complications were observed: cataracts (5 eyes, of which 4 eyes

Table 1: *visual acuity, ophthalmological findings and treatment.*

patient	gender	age at diagnosis	eye	VA at diagnosis	Follow-up (Years)	VA at last visit	Findings at diagnosis	corticosteroid treatment	surgical teatment
1	M	9	RE	20/30	3	20/25	MO, S	T, PB	
			LE	20/20		20/20	ST, S	T	
2	F	4	RE	20/30	0,5	20/20	KP, MO	T, PB	
			LE	20/50		20/25	KP, MO	T, PB	
3	M	8	RE	CF	6	20/200	PS, MO, PP, S	T, PB, O	
			LE	20/50		20/20	PS, MO, PP, S	T, PB, O	
4	M	6	RE	20/60	3	20/20	KP, DH, S	T, PB	
			LE	20/40		20/20	KP, DH, S	T, PB	
5	F	14	RE	20/40	10	20/28	DO, MO, VPT, VRT, NV, S	T, PB	
			LE	20/60		20/200	DO, MO, VPT, VRT, S	T, PB, I	LP, CT, CS
6	M	8	RE	20/125	11	20/60	C, MO, VPT, PP, S	T, PB	
			LE	20/125		20/125	C, MO, VPT, PP, S	T, PB	
7	M	6	RE	20/50	1	20/22	DH, MO, VRT, PP, S	T, PB	
			LE	20/20		20/30	DH, MO, VRT, PP, S	T, PB	
8	M	4	RE	20/20	4	20/22	C, PS, DH, DO, S	T, PB, O	CS
			LE	20/40		20/20	PS, DH, DO, S	T, PB, O	
9	M	9	RE	20/25	1	20/25	ST, S	T, PB	SS
			LE	20/100		20/25	ST, S	T, PB	SS
10	M	9	RE	20/60	3	20/25	C, DH, MO, PP, S	T, PB	CS
			LE	20/125		20/30	DH, MO, PP, S	T, PB	

VA: visual acuity; RE: right eye; LE: left eye; CF: couting fingers; MO: macular oedema; S: snowballs; KP: keratic precipitates; PS: posterior synechiae; PP: periphlebitis; DH: disc hyperaemia; DO: disc oedema; VPT: vitreopapillary traction; VRT: vitreoretinal traction; NV: neovascularisation on disc and elsewhere; C: cataract; ST: strabismus; T: topical; PB: parabulbar; O: oral; I: intravitreal; LP: laser photocoagulation of the retina; CT: cryotherapy of the retina; CS: cataract surgery; SS: strabismus surgery

at the time of diagnosis), vitreoretinal traction (4 eyes), vitreopapillary traction (4 eyes), retinal neovascularisation (1 eye), peripheral retinoschisis (1 eye) and vitreous haemorrhage (2 eyes). A high intra-ocular pressure of 28,6 mmHg +/- 1.34 mmHg (range, 27 to 30 mmHg) was noted in 5 eyes, in two patients (4 eyes) it was due to steroid response.

Treatment consisted of topical corticosteroids in all cases. Additional parabulbar injections were administered in 19 eyes (3,6 injections per eye +/- 3.1, range 0 to10 injections). Systemic corticosteroids were prescribed in 2 patients: patient 3 received oral methylprednisolon 9 mg/kg/day with tempering of the dose over a period of 38 days; patient 8 received oral methylprednisolon 1 mg/kg/day with tempering of the dose over one week to prevent recurrence of inflammation during and after cataract surgery. In one case with retinal neovascularisation, an intravitreal injection with triamcinolone, laser photocoagulation and cryotherapy of the retina was applied.

Occlusion therapy for prevention of amblyopia was prescribed in 4 patients. Intraocular lens implantation was necessary in three eyes, with the diagnosis of cataract at the time of presentation. In one of these eyes, an additional anterior vitrectomy and a surgical posterior capsulorhexis were performed.

One patient underwent strabismus surgery.

At the last visit in our outpatient clinic, the VA was 20/25 or better in 13 eyes (65%), between 20/100 and 20/25 in 4 eyes (20%) and between 20/200 and 20/100 in 3 eyes (15%). Visual acuity improved remarkably in all but three eyes (15%) under treatment (Table 1.). Biomicroscopy showed no inflammatory activity in all cases. There was cataract in 3 eyes (15%) and 3 eyes were pseudophakic of which one had posterior capsular opacification.

The ophthalmoscopic fundus findings at a last visit were normal in 4 eyes (20%). Fundus abnormalities included macular oedema in 3 eyes (15%), preretinal membranes in 2 eyes (10%), vitreopapillary traction in one eye (5%), pe-

ripheral retinoschisis in one eye (5%), snowbanking in 4 eyes (20%) and vitreous haze and cells in one eye (5%).
The topical corticosteroid therapy was ongoing in 16 eyes (80%).

DISCUSSION

Pars planitis is an idiopathic eye disease most often presenting in childhood. The typical snowbank formation was examined immunohistochemically by Abu El-Asrar et al. (1). This snowbank formation is a result of extensive tissue repair and remodelling, incorporating collagen type II, a normal component of vitreous, and collagen type I and III which are not found in normal vitreous. Immunohistochemical studies report the presence of Tenascin. Tenascin is a large hexameric extracellular matrix glycoprotein that is found in embryonic tissues, especially at epithelial-mesenchymal junctions, and in developing brain tissue, where it probably plays a role in epithelial-mesenchymal induction and migration. In the retina, it has been reported to be present in choroidal neovascular membranes, in proliferative vitreoretinopathy, proliferative diabetic retinopathy and macular pucker (6,7).
We report upon a group of children, with a variety of vision-threatening complications, all documented in the literature (4,9,10,12,13).
In a retrospective study of 219 patients with uveitis younger than 16 years, pars planitis was diagnosed in 11,87% of children with uveitis (4). Pars planitis was the only type of uveitis with anatomic intermediate localization in children. Not one patient with intermediate uveitis was found to have associated systemic disease. The most frequent complications included cataract (32,7%) and maculopathy (30,7%) and posterior segment neovascularisation (4%). In our patients, the distribution was similar for cataract (40%) and retinal neovascularisation (5%), but maculopathy was present in 65%. The surgical procedures in the study of Kadayifçilar (4) were cataract extraction (14,3%), lensectomy and anterior vitrectomy (2%) and peripheral cryotherapy (22,5%). In our patients, we encountered an equal need for cataract extraction (3 eyes, 15%) and less for cryotherapy (2 eyes, 10%).
In a retrospective study of 118 patients with pars planitis by Lauer et al. (10), vitreous haemorrhage as a complication of pars planitis was encountered in 14% of patients. This complication was more frequent in children than in adults, as children often present at the clinic in a later, florid stage of the disease. This can be explained by the lack of complaints at young age and subsequently presentation in an advanced stage of the disease. Children also exhibit a more exuberant inflammatory and/or fibrovascular response.
In our patient with retinoschisis (or exudative retinal detachment), the size of the schisis waned with time. This finding confirms the exudative aetiology of the detachment, as described by Pollack et al. (13). He and his co-authors assumed that the peripheral retinoschisis and/or exudative retinal detachment may be related to a Coats-like vascular response secondary to chronic inflammation. Treatment of the vascular leakage tended to result in a resolution of the detachment and/or schisis.
In the treatment of pars planitis, systemic immunosuppressive therapy is needed when therapy with systemic corticosteroids is insufficient to control disease progression.
Methotrexate is now widely used in management of uveitis (including intermediate) as a steroid sparing and immunosuppressant drug, despite the lack of controlled trials to positively demonstrate its effectiveness. In a number of case series in inflammatory eye disease and in the rheumatology literature the drug's use has been rigorously proven with a good safety profile (16). Other immunomodulatory treatments that have been used with variable success in the treatment of uveitis include cyclosporine, chlorambucil, azathioprine, tacrolimus (FK506) and tumor necrosis factor antagonists. They can nevertheless be useful in treatment of resistant uveitis. (3, 9, 12, 15) Cyclosporine is claimed to be the most effective treatment. Cyclophosphamide works very quickly but has significant side effects and is therefore not used as first line treatment in children.

CONCLUSION

Our retrospective study consisted of a small number of patients, presenting however a wide

variety of symptoms and complications. Topical corticoids are the basis of treatment and may have to be administered for a long period. In our young patient population, we have not encountered one case that was not responding to systemic corticosteroids. We emphasise the need of rigorous treatment and frequent follow-up (every 4 to 6 weeks), because of fluctuating disease activity and the risk of intraocular pressure rise.

REFERENCES

(1) ABU EL-ASRAR A.M., GEBOES K. – An immunohistochemical study of the 'snowbank' in a case of pars planitis. Ocul Immunol Inflamm. 2002;10:117-23.

(2) AMERICAN ACADEMY OF OPHTHALMOLOGY – Basic and clinical science course 2003-2004: section 9. Intraocular Inflammation and uveitis. San Francisco: American Academy of Ophthalmology; 2003:143-148.

(3) ARELLANES-GARCIA L., NAVARRO-LOPEZ L., RECILLAS-GISPERT C. – Pars planitis in the Mexican Mestizo population: ocular findings, treatment, and visual outcome. Ocul Immunol Inflamm. 2003;11:53-60.

(4) DEV S., MIELER W.F., PULIDO J.S., MITTRA R.A. – Visual outcomes after pars plana vitrectomy for epiretinal membranes associated with pars planitis. Ophthalmology. 1999;106:1086-90.

(5) GILES C.L., BLOOM J.N. – Uveitis in childhood. Duane's Clinical Ophthalmology: Vol.4. Chap.56. pp.8-11.

(6) HAGEDRON M., ESSER P., WIEDEMANN P., HEIMANN K. – Tenascin and decorin in epiretinal membranes of proliferative vitreoretinopathy and proliferative diabetic retinopathy. Ger J Ophthalmol. 1993;2:28-31.

(7) IMMONEN I., TERVO K., VIRTANEN I., LAATIKAINEN L., TERVO T. – Immunohistochemical demonstration of fibronectin and tenascin in epiretinal membranes. Acta Ophthalmol (Copenh). 1991;69:466-471.

(8) KADAYIFCILAR S., ELDEM B., TUMER B. – Uveitis in childhood.J Pediatr Ophthalmol Strabismus. 2003;40:335-40.

(9) KAPLAN H.J., TESSLER H.H., GOLDSTEIN D.A. – American Uveitis Society Winter Symposium, January 1997. Ocul Immunol Inflamm. 1997.5.pp.283-288

(10) LAUER A.K., SMITH J.R., ROBERTSON J.E., ROSENBAUM J.T. – Vitreous hemorrhage is a common complication of pediatric pars planitis. Ophthalmology. 2002;109:95-8.

(11) MERAYO-LLOVES J., POWER W.J., RODRIGUEZ A., PEDROZA-SERES M., FOSTER C.S. – Secondary glaucoma in patients with uveitis. Ophthalmologica. 1999;213:300-4.

(12) MISEROCCHI E., BALTATZIS S., EKONG A., ROQUE M., FOSTER C.S. – Efficacy and safety of chlorambucil in intractable noninfectious uveitis: the Massachusetts Eye and Ear Infirmary experience. Ophthalmology. 2002;109:137-42.

(13) POLLACK A.L., McDONALD H.R., JOHNSON R.N., AI E., IRVINE A.R., LAHEY J.M., LEWIS H., RODRIGUEZ A., RYAN E.H. Jr, SHIELDS C.L. – Peripheral retinoschisis and exudative retinal detachment in pars planitis. Retina. 2002;22:719-24.

(14) POTTER M.J., MYCKATYN S.O., MABERLEY A.L., LEE A.S. – Vitrectomy for pars planitis complicated by vitreous hemorrhage: visual outcome and long-term follow-up. Am J Ophthalmol. 2001;131:514-5.

(15) SLOPER C.M., POWELL R.J., DUA H.S. – Tacrolimus (FK506) in the treatment of posterior uveitis refractory to cyclosporine. Ophthalmology. 1999;106:723-8.

(16) STRAWELL R.J. – Methotrexate in inflammatory eye disease. Ocular Immunology and Inflammation, 2003, 11, 79-82.

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