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Major review

Punctate inner choroidopathy: A review

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ABSTRACT

Punctate inner choroidopathy (PIC), an idiopathic inflammatory multifocal chorioretinopathy that predominantly affects young myopic women, appears to be relatively rare, but there are limited data to support accurate estimates of prevalence, and it is likely that the condition is underdiagnosed. The etiological relationship between PIC and other conditions within the "white dot syndromes" group remains uncertain. We, like others, would suggest that PIC and multifocal choroiditis with panuveitis represent a single disease process that is modified by host factors (including host immunoregulation) to cause the range of clinical phenotypes seen. The impact of PIC on the patient is highly variable, with outcome ranging from complete spontaneous recovery to bilateral severe sight loss. Detection and monitoring have been greatly facilitated by modern scanning techniques, especially optical coherence tomography and autofluorescence imaging and may be enhanced by coregistration of sequential images to detect change over time. Depending on the course of disease and nature of complications, appropriate treatment may range from observation to systemic immunosuppression and antiangiogenic therapies. PIC is a challenging condition

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where treatment has to be tailored to the patient's individual circumstances, the extent of disease, and the risk of progression.

1. Introduction

1.1. Definition

Punctate inner choroidopathy (PIC) is a relatively rare idiopathic inflammatory multifocal chorioretinopathy that most commonly affects young myopic women. Most of these lesions involve the posterior pole, arising at the level of the retinal pigment epithelium (RPE) and inner choroid in the absence of anterior chamber or vitreous inflammation (Fig. 1).[86] Although it may be self-limiting with a favorable outcome, inflammation or neovascularization abutting the fovea may cause permanent visual loss. Depending on the course of disease and development of complications, treatment may range from observation to systemic immunosuppression and intravitreal antiangiogenic therapies.

1.2. Historical background

PIC was first described by Watzke and colleagues in 1984 in a series of 10 young, otherwise healthy myopic women who presented with blurred central vision, photopsia, and paracentral scotomas and had well circumscribed yellow-gray lesions at the level of the inner choroid and retinal pigment epithelium associated with small neurosensory retinal detachments in the macula in the absence of detectable intraocular inflammation.[86] Eight of the 10 patients presented with bilateral lesions, and choroidal neovascularization (CNV) developed in 6 patients. Although initially hypothesized to be secondary to myopia, the episodic nature of recurrences was subsequently acknowledged to be more suggestive of an underlying inflammatory pathogenesis.[1]

1.3. Other names

Punctate inner choroiditis, multifocal inner choroiditis.

2. Epidemiology and demographics

PIC is a relatively rare multifocal chorioretinopathy. It is difficult to make an accurate estimate of the incidence and prevalence of PIC for a number of reasons: first, there is a wide range of presentation and severity, such that many cases may remain unrecognized; second, there is uncertainty over its classification, notably whether it is a distinct entity from multifocal choroiditis with panuveitis (MCP) or part of the same spectrum; and third, in the absence of national registries or reporting systems, estimates are often based on data from single centers for which the size of population and completeness of coverage is uncertain. There is also variable practice between institutions as to whether patients with PIC are under of the care of uveitis subspecialists, medical retina subspecialists, or both, such that estimates drawn from a "single service" cohort may be an underestimate.[1,8,24,35]

While acknowledging these limitations, the following estimates may still be helpful. A retrospective review at the University of Iowa by Brown and colleagues collected 16 cases of PIC over a period of 15 years (1980–1994). Based on an estimate of the population of Iowa of 2.8 million, this would equate to around 0.4 new cases per million population per year.[10] A study in the United Kingdom by Jones reporting the case-mix of the Manchester Uveitis Clinic from 1991 to 2013 found that PIC accounted for 2.8% of all uveitis cases referred to that service. They noted that the incidence of new cases seen from their local catchment area was 5.6/100,000/year for the decade 2003–2013.[35] Based on these figures, an incidence of around 1.6 per million population per year may be estimated. This figure, however, may be an overestimate owing to higher rates of "out-of-area" referrals for complex conditions such as PIC compared to more common forms of uveitis.

Gerstenblith and colleagues evaluated the demographics and clinical features of PIC by analyzing a survey questionnaire completed by 77 patients through the PIC society.[30] Among the respondents, 90% were female, 97% were white, and 85% were myopic with a median refractive error of −7.00 diopters in each eye. The majority of the participants were young, with a median age of 30 years (range, 15–55).

As noted in the Gerstenblith study, PIC tends to mostly affect young women. Other series confirm this profile, with females comprising 92–100% in the United States,[10,62,86] 76–93% in the United Kingdom,[24,35] and 72% in China.[89] Similarly myopes predominate in all countries studied. Myopia was reported in 80–100% of series from the United States, the United Kingdom, and China. In their series of 136 UK patients with PIC, Essex and colleagues[24] reported that the mean spherical equivalent refraction was −4.6 diopters with a range of −14 to +4 diopters. Additionally, Reddy and colleagues noted that patients with PIC had the highest level of myopia of all inflammatory chorioretinopathies.[67]

There is, however, a risk of selection and reporting bias here. Because diagnosis is based on subjective assessment of a clinical pattern rather than a diagnostic test based on etiology, it is likely that clinicians will be more likely to diagnose the condition when seen in a typical patient, and thus the same clinical pattern in an older, male, hyperopic patient may be labeled with an alternative diagnosis (such as "idiopathic," or multifocal choroiditis). This may be exaggerated in surveys of patient societies (such as the Gerstenblith study) where clinician bias may be compounded by patient self-reporting bias. Certainly, PIC is not exclusive to the young, female, myopic population as shown in a number of series and case reports.[7,10,30,35,89]