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Ischemic optic neuropathy☆

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A B S T R A C T

Ischemic optic neuropathy is one of the major causes of blindness or seriously impaired vision, yet there is disagreement as to its pathogenesis, clinical features and especially its management. This is because ischemic optic neuropathy is not one disease but a spectrum of several different types, each with its own etiology, pathogenesis, clinical features and management. They cannot be lumped together. Ischemic optic neuropathy is primarily of two types: anterior (AION) and posterior (PION), involving the optic nerve head (ONH) and the rest of the optic nerve respectively. Furthermore, both AION and PION have different subtypes. AION comprises arteritic (A-AION – due to giant cell arteritis) and, non-arteritic (NA-AION – due to causes other than giant cell arteritis); NA-AION can be further classified into classical NA-AION and incipient NA-AION. PION consists of arteritic (A-PION – due to giant cell arteritis), non-arteritic (NA-PION – due to causes other than giant cell arteritis), and surgical (a complication of several systemic surgical procedures). Thus, ischemic optic neuropathy consists of six distinct types of clinical entities. NA-AION is by far the most common type and one of the most prevalent and visually crippling diseases in the middle-aged and elderly. A-AION, though less common, is an ocular emergency and requires early diagnosis and immediate treatment with systemic high dose corticosteroids to prevent further visual loss, which is entirely preventable.

Controversy exists regarding the pathogenesis, clinical features and especially management of the various types of ischemic optic neuropathy because there are multiple misconceptions about its many fundamental aspects. Recently emerging information on the various factors that influence the optic nerve circulation, and also the various systemic and local risk factors which play important roles in the development of various types of ischemic optic neuropathy have given us a better understanding of their pathogeneses, clinical features and management. This knowledge should help us not only to manage them better but also to reduce their incidence. For example, clinically, the evidence that about 40% of NA-AION eyes experience spontaneous improvement in visual acuity and that systemic steroid therapy during early stages in both NA-AION and NA-PION has a significant beneficial effect for visual outcome are encouraging developments. This review discusses the current concepts on various issues related to various types of ischemic optic neuropathy.

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1. Introduction

Ischemic optic neuropathy constitutes one of the major causes of blindness or seriously impaired vision among the middle-aged and elderly population, although no age is immune. Its pathogenesis, clinical features and management have been subjects of a good deal of controversy and confusion. I have conducted basic, experimental and clinical research on the blood supply of the optic nerve and on various aspects of ischemic optic neuropathy since 1955. This review is based on the cumulative information drawn from those studies, as well as from a PubMed search of the literature on the subject.

2. Terminology

Before 1974, this condition was described under different eponyms, including optic neuritis, arteriosclerotic papillitis, senile papillopathy, papillary apoplexy, vascular pseudo-papillitis, opticomalacia, ischemic neuritis of papilla, ischemic papillopathy and ischemic optic neuritis and so on (Hayreh, 1975a). Since studies have shown that it is an acute ischemic disorder of the optic nerve, the proper terminology for this disease is "ischemic optic neuropathy". Based on the blood supply pattern of the optic nerve, and my clinical and experimental studies, in 1974 I defined ischemic optic neuropathy into the following two distinct clinical entities.

2.1. Anterior ischemic optic neuropathy (AION)

This is due to ischemia of the anterior part of the optic nerve, which is supplied by the posterior ciliary artery (PCA) circulation (Hayreh, 1969, 1995, 2001b) (Fig. 1A). In view of that I named it "anterior ischemic optic neuropathy" (Hayreh, 1974b).

2.2. Posterior ischemic optic neuropathy (PION)

I first described this clinical entity in 1981 (Hayreh, 1981b); it is due to ischemia of a segment of the posterior part of the optic nerve, which is supplied by multiple sources but not the PCA (Figs. 1B and 2).

Of the two types, AION is far more common than PION. Pathogenetically and clinically AION and PION are quite distinct clinical entities; thus, the common practice of calling AION simply "ischemic optic neuropathy" is incorrect, and "ischemic optic neuritis" is worse still, since there is no evidence of inflammation.

From the basic scientific facts about the disease process, one can logically deduce its pathogenesis, clinical features and management. The basic sciences are the foundation of Medicine. To comprehend the scientific basis of the pathogeneses, various clinical features and management of AION and PION, the first essential is to have a good understanding of the various basic scientific issues involved. Since this is an ischemic disorder of the optic nerve, the