Aetio Pathogenesis Of Optic Atrophy: Case Study

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Abstract: *The term optic atrophy describes a group of clinical conditions which have an abnormal pallor of the disc as a common physical sign. Optic atrophy is not a disease; it is the end result of any pathologic process that damages the retinal ganglion cells and axons of the retinogeniculate pathway. In other words, optic atrophy is defined as a condition characterized by the loss of conducting function of the optic nerve.*

Keywords: optic nerve,optic atrophy,pathology

1.Introduction

In addition to being a sequel of many diseases, primarily of the optic nerve, optic atrophy is also frequently an aftermath of varied disorders of the retina, optic chiasma and optic tract[1].

Therefore it is important to diagnose the etiology and treat a case of optic atrophy at the earliest not only for good visual outcome but also for the general health and life of the patient[2].

2.Aim of the study

Fifty clinically established cases of optic atrophy who attended to the Department of Ophthalmology, Government General Hospital, Kakinada during 2011-2013 were taken into the study.This study is made not only to know the aetiology of optic atrophy but also evaluate the preventable causes of optic atrophy like glaucomatous optic atrophy and optic atrophy due to ICSOL, in which early diagnosis and prompt treatment either may delay or stops the disease process to progress towards optic atrophy.

3.Anatomy of the optic nerve

The Optic nerve, ensheathed in pia, runs as a flattened band from the anterolateral angle of somewhat quadrilateral chiasma forwards and laterally and slightly downwards to optic foramen. It begins anatomically at the optic disc but physiologically and functionally within the ganglion cell layer that covers the entire retina[3].

The total length of the nerve is about 3.5 to 5.5 cm. It can be divided into four parts.

1. Intraocular nerve head : 0.7-1 mm
2. Intraorbital : 25-30 mm
3. Intracanalicular : 6-10 mm
4. Intracranial : 10 mm

It's diameter ranges from 3 mm in orbit to 7 mm near the chiasma.

Where it leaves the eye, the centre of optic nerve is just above and 3 mm medial to the posterior pole of globe.

Within the orbit, it is round in cross section. As it passes to the optic foramen it takes a sinuous course allowing

for ocular movements. Just posterior to the sclera the optic nerve acquires a dural sheath that is contiguous with the periorbita of the optic canal and arachnoid membrane that supports and protects the axons and is contiguous with the arachnoid of the subdural intracranial space through the optic canal. It also acquires the myelin coating just posterior to lamina cribrosa, which increase it's diameter to 3 mm (6 mm with optic nerve sheath) from 1.5 mm of the optic disc[4].

In the optic canal it is oval in cross section. The endosteal layer of duramater merges with the periosteum, so that both here and in the orbit, the nerve is covered by all meninges and is bathed with CSF.

As the nerve leaves the optic canal, it is pyriform in shape with a rounded end medially. And it becomes a flat band as it inclines posteromedially and slightly upwards to the chiasma.

Although we speak of the optic nerve, it is really not a nerve, but essentially a fiber tract joining two portions of the brain. The evidence for this is incontrovertible. Firstly it is an outgrowth of the brain. Secondly it's fibers possess no neurolemmal cells, though other satellite cells occur. Thirdly it is surrounded by the meninges, unlike any peripheral nerve. Fourthly, (and most cogently) the "primary" and "secondary" sensory neurons of the pathway are both in the retina, the ganglion cells corresponding, for example to those in the gracile and cuneate nuclei of medulla oblongata.

Intra ocular nerve head (optic disc, optic papilla)

The intraocular portion of the optic nerve extends from its anterior surface in contact with the vitreous to a plane, which is level with that of the posterior sclera surface.The choroid ends abruptly here, as do all elements of the retina except its axo The axons bend at a right angle into the nerve head and p posteriorly through the scleral canal.

4.Observations and Analysis

Table no: 1:**Age distribution of current sample of optic atrophy**

Age in years	Number of cases		
	male	female	Total
1-10	2	1	3
11-20	3	3	6
21-30	3	6	7
31-40	5	5	10
41-50	3	3	6
51-60	7	5	12
61-70	3	1	4
71-80	1	1	2
			50

Observation: optic atrophy is seen in all age groups but it is less in extremes of age and peaks between 4^{th} and 6^{th} decades.

Table No. 2 : **Gender distribution of current sample of optic atrophy**

Gender	Number of cases	percent
Male	**27**	**54%**
female	**23**	**46%**

Observation: In the present study males are more affected with optic atrophy than females

Table No. 3 Etiology of optic atrophy

SI no	Cause	No. of cases	
			Total
1	**Pressure and traction atrophy**		**24**
	a) Glaucomatous optic atrophy	14	
	b) papilledema and optic atrophy	5	
	c) intracranial tumors without raised ICT	4	
	d) Tumors of optic nerve and its sheath	1	
2	**Consecutive optic atrophy**		**14**

	a) difuse chorioretinitis	2	
	b) degenerative		
	i. pigmentary retinal dystrophy	11	
	ii. Myopic degeneration	1	
3	**Circulatory atrophy**		**2**
	CRAO	2	
4	**Post inflammatory**	3	**3**
6	**Traumatic atrophy**	3	**3**
7	**Toxic atrophy**		**1**
	tobacco, alcohol	1	
8	**Metabolic (nutritional)**	1	**1**
9	**Idiopathic**	2	**2**
			50

Observation: In the current study, glaucoma and retinitis pigmentosa are the most common causes of optic atrophy. Next common causes being intracranial tumors, post inflammatory, traumatic, circulatory and idiopathic.

Table No. 4 Optic atrophy and laterality

RE	LE	BE	Total
5	6	39	50

Observation : Out of the 50 cases of optic atrophy 39 cases are bilateral optic atrophy and 11 cases are unilateral affecting one eye.

Table No. 5 Unilateral optic atrophy – etiology

Etiology	No. of cases
Trauma	3
Post inflammatory	3
Circulatory	2
Chorioretinitis	2
Tumors of optic nerve and its sheath	1
Total	**11**

Observation: Out of the 11 cases of unilateral optic atrophy in the present study, trauma (3 cases), and post inflammatory (3 cases) are the major causes and other causes being circulatory (2 cases), tumors of optic nerve and its sheath (1 case) and chorioretinitis (2 cases).

Table No.6: Bilateral optic atrophy – etiology

Etiology	No.of cases
Glaucoma	14
Retinitis pigmentosa	11
Papilloedema	5
Intracranial tumors without raised ICT	4
Myopic degeneration	1
Toxins	1
Metabolic (nutritional)	1
Idiopathic	2
Total	**39**

Observation: In this study glaucoma (14 cases) and retinitis pigmentosa (11 cases) are the major ocular causes of optic atrophy. Intracranial tumors and papilloedema being the next common cause

Table No. 7: Ocular causes of optic atrophy

Diagnosis	No. of cases
Glaucoma	14
Retinitis pigmentosa	11

Circulatory atrophy	2
Postinflammatory	3
Trauma	3
Chorioretinitis	2
Myopic degeneration	1
Total	**36**

Observation: Out of the etiologically established cases of optic atrophy there is ocular pathology in 36 cases. Glaucoma and retinitis pigmentosa are the most common ocular causes leading to optic atrophy.

Table No. 8:**Extra ocular causes of optic atrophy**

Etiology	**No.of cases**
Intracranial tumors without raised ICT	4
Papilloedem and optic atrophy	5
Tumors of optic nerve and its sheath	1
Toxic atrophy	1
Metabolic	1
Total	**12**

Observation: In the current study on optic atrophy, out of 50 cases,12 cases are due to extra ocular causes. Intracranial tumors, papilloedema (due to intracranial tumors or meningitis) constituting the common extra ocular causes of optic atrophy.

Table no. 9: **Age / gender distribution of glaucomatous optic atrophy**

Age in years	**Number of cases**		
	male	**female**	**total**
1-10	-	-	-
11-20	-	1	1
21-30	-	-	-
31-40	1	-	1
41-50	1	2	3
51-60	4	1	5
61-70	2	1	3
71-80	-	1	1
Total	**8**	**6**	**14**

Observation: In the current study, glaucomatous optic atrophy is seen in 14 patients out of whom 8 are males, 6 are females. All the cases except one (congenital glaucoma) belong to the age groups of 3rd to 6th decades. It peaks between 5th and 7th decades.

Table No. 10:**Age / Gender distribution of consecutive optic atrophy due to**

Retinitis Pigmentosa

Age in years	**Number of cases**		
	Male	**female**	**Total**
1-10	-	-	-
11-20	1	-	1
21-30	2	1	3
31-40	2	2	4
41-50	1	1	2
51-60	-	1	1

61-70	-	-	-
Total	**6**	**5**	**11**

Observation: In the current study out of 11 cases of consecutive optic atrophy due to Retinitis Pigmentosa, 6 are males, 5 are females and it peaks between 3rd and 4th decades

Observation: Out of the 89 eyes of 50 patients with optic atrophy, 32 eyes have visual acuity of NO PL at presentation of which pressure and traction atrophy (glaucoma, papilledema, intracranial tumors) is the major cause. Other causes for such total loss of visual acuity being RP, trauma, circulatory and post inflammatory. 29 eyes have visual acuity of less than CFlmt to PL, among which RP is the predominant cause and next common cause being glaucoma.

5.Discussion

The current study on optic atrophy is a prospective, hospital based study conducted in the department of ophthalmology, Government General Hospital, Kakinada during the period of 2011-2013. This study included 50 cases

Table No. 11:**Visual acuity at presentation**

	616 - 6/18	**6/24 - 6/60**	**<6/60 - CFlmt**	**<CFlmt - PL**	**NO PL**
Pressure & traction atrophy	3	8	6	11	19
Consecutive atrophy	-	2	5	12	7
Circulatory	-	-	-	1	1
Post inflammatory	-	-	-	2	1
Traumatic atrophy	-	-	-	-	3
Toxic atrophy	-	2	-	-	-
Metabolic (nutritional)	-	2	-	-	-
Idiopathic	-	-	-	3	1
Total	**3**	**14**	**11**	**29**	**32**

of optic atrophy that attended the ophthalmic OPD directly or referred from other departments like Paediatrics, general medicine, neuromedicine or neurosurgery.

The results of this study are discussed and analyzed as follows.

Age and Gender distribution of optic atrophy

In the present study, optic atrophy was found in all age groups with its peak occurrence between 4th to 6th decades. Out of the 50 cases, 3 cases belonged to pediatric age group.

The gender distribution of the current sample shows a male preponderance with 54% of cases belonging to male gender. The preponderance of optic atrophy in males may be due to Retinitis Pigmentosa being more common in males because of X- linked recessive inheritance. And also may be due to males being more frequently involved in road traffic accidents leading to traumatic optic atrophy.

Etiology of optic atrophy

One hundred clinically established cases of optic atrophy included in this study are grouped according to Duke-Elder's classification.

1. Pressure and traction atrophy 24
2. Consecutive optic atrophy 14
3. Circulatory atrophy 2

4. Post inflammatory 3
5. Traumatic optic atrophy 3
6. Toxic atrophy 1
7. Metabolic 1
8. Idiopathic 2

In 4% cases the cause of optic atrophy could not be determined. In our study we could not find congenital or hereditary causes of optic atrophy. These causes may account for few of the cases of idiopathic group.

Of the 96% cases in which etiology was established, 72% cases have ocular causes, 24% cases are due to extra ocular causes

Optic atrophy and visual acuity

Out of 89 eyes of 50 patients included in this study, 32 eyes have no perception of light and 29 eyes have visual acuity between <CFlmt and PL. 19 eyes having visual acuity of less than CFlmt belong to pressure and traction optic atrophy of which glaucoma is the major cause, next common causes being intracranial tumors, papilledema.

Optic atrophy and visual field defects

In this study 17 eyes having visual acuity of 6/60 or better are subjected for visual field examination. In 9 eyes with glaucoma, the following field defects were noted: arcuate scotomas in 7 eyes, tubular vision in 1 eye and 1 patient was not cooperative. Out of 5 eyes of RP having visual acuity 6/60 or better, 3 eyes have peripheral constriction of fields and in 2 eyes the visual fields were not recorded as the patients

were not co-operative. Centrocaecal scotoma was found in 4 eyes in patients of toxic atrophy and metabolic atrophy. Concentric constriction of visual fields was noted in a patient with papilledema.

Optic atrophy in glaucoma

Glaucoma being the predominant cause of optic atrophy (either complete or partial) in this study is further analyzed in detail. Out of the 14 cases, 8 are males and 6 are females. Glaucomatous optic atrophy peaks between 5^{th} and 7^{th} decades. Out of the 14 cases one case was of congenital glaucoma with total glaucomatous optic atrophy, 8 cases are of primary open angle glaucoma, 3 cases are of pseudo exfoliation glaucoma and 2 are of chronic angle closure glaucoma.

Out of the 14 cases, 7 cases have complete optic atrophy in both eyes; 5 cases have one eye complete and other eye partial optic atrophy and 2 cases have both eyes partial optic atrophy (reduced visual acuity or field defects)

Out of 28 eyes of 14 patients, 10 eyes have IOP between 24 and 30 mmHg, 16 eyes have IOP between 3land 40 mm Hg and 2 eyes have IOP between 4l and 50 mm Hg.

Patients are treated with topical antiglaucoma medication like Timolol 0.5% e/d, or Betoxolol 0.5%e/d, or Dorzolamide 2% e/d (or)

BrimonidineO.2% e/d (or) Latanoprost 0.005% e/d either with single drug or two drugs. Oral carbonic anhydrase inhibitors were given wherever necessary and were regularly followed every month. The visual outcome was assessed after 3 and 6 month and the visual acuity remained stable in all the patients.

This workup about glaucoma shows that because of it's asymptomatic nature and painless slow progressive loss of vision patients

are presenting to the ophthalmologist in advanced stage of disease either in one eye or both eyes. This emphasizes the fact that

all patients above 40 yrs should be regularly screened for glaucoma and treated if necessary to prevent blindness from it.

Optic atrophy in Retinitis pigmentosa

Retinitis Pigmentosa was found to be the cause of optic atrophy in 11 cases, out of which 6 are males and 5 are females. The relative male preponderance may be due to X-linked recessive pattern of inheritance in some cases apart from AD & AR inheritance.

In this study optic atrophy due to RP peaked between 3rd & 4th decades and was consistent with the previous studies.

Optic atrophy and intracranial tumors

In this study there are cases 4 of intracranial tumors causing optic atrophy: of which 2 are craniopharyngiomas, one case is pituitary adenoma and one case is meningioma of sphenoid. All the 4 cases have chronic headache for more than two years and gradual loss of vision in both eyes, but were using analgesics for symptomatic relief of pain. They presented to our department in advanced stage of optic atrophy and there was no visual improvement even after surgical management in 3 cases.

This shows that patients with chronic headache and visual disability should be thoroughly examined clinically with great attention towards any pallor of optic disc and examine the visual fields to detect any defects suggestive of compression of optic nerve, chiasma or optic tract. Early diagnosis and management in such cases will prevent blindness and it is important for the life of the patient as well.

Optic atrophy and trauma

In this clinical study trauma was the cause of optic atrophy in 6%

of cases. All the three patients sustained injuries in road traffic accidents and had sudden loss of vision after accidents. In all cases there is unilateral complete primary optic atrophy.

In individuals who sustain injuries in RTA, optic nerve damage should be suspected when the patient has decreased visual acuity (in conscious patients) and / or relative afferent pupillary defect and recording the visually evoked potentials. The nerve damage is produced by shearing forces induced in the relatively immobile canalicular portion of optic nerve by the movement of nerve. There may be direct damage by disruption of nerve fibers or indirect damage by disruption of blood supply. Edema and hemorrhage can also induce nerve damage by compressing the nerve within the optic canal. In addition in a fracture of the wall of the optic canal, bone fragments can directly exert pressure on the optic nerve. Rarely, blunt trauma can lead to optic nerve damage in the orbit, producing an optic nerve head avulsion.

Early institution of therapy with I.V. Methyl prednisolone for 3 days and tapering doses of oral corticosteroids or surgical decompression of optic canal may preserve useful vision in such cases.

Optic atrophy and laterality

Out of the 50 cases, 39 cases have bilateral optic atrophy and 11 cases have unilateral optic atrophy.

Glaucoma (14 cases) and RP (11 cases) account for the major causes of bilateral optic atrophy. Next common causes being intracranial tumors (4 cases), optic atrophy due to papilludema (5 cases), toxic (1 case), metabolic (1 case) and due to myopic degeneration (1case).

The causes of unilateral optic atrophy being circulatory (2 cases), post inflammatory (3 cases), trauma (3 cases), chorioretinitis(2 cases) and tumors of optic nerve and its sheath (1 case).

Circulatory Optic atrophy

In the present study 4% of cases are due to circulatory causes. All cases are due to CRAO.

It is important to diagnose and work up in detail a case of circulatory optic atrophy because in most of the cases underlying cause would be atherosclerosis related

thromboembolism and the patients would be hypertensives or diabetics, which if not treated has high risk of developing stroke or MI.

Toxic optic atrophy

In this study 2% of cases belong to toxic atrophy. All cases are due to tobacco-alcohol. These two patients improved after treatment.

Visual prognosis in optic atrophy

Optic atrophy has varied etiology. Visual prognosis depends upon etiology, the stage of disease and also on the time & effectiveness of management of the cause.

Glaucoma is one of the important and major cause of optic atrophy in which early diagnosis and treatment can stop the progression of disease and thus visual loss can be prevented. As glaucoma leads to painless,

slow progressive loss of vision which is irreversible, regular screening of the population above 40 years for glaucoma and its effective treatment can reduce the incidence of blindness form it.

Next common cause of optic atrophy in this study is retinitis pigmentosa, which is genetically transmitted. As there is no effective treatment till date, the incidence of RP can be reduced by genetic counseling, discouraging consanguineous marriages and limitation of number off-spring in the married individuals with RP.

Other causes leading to optic atrophy like intracranial tumor, optic neuritis, meningitis etc. should be diagnosed and treated at the earliest to prevent visual loss.

6.Conclusion

Optic atrophy is morphologic sequel of any disease that causes damage to ganglion cells and axons of the retinogeniculate pathway characterized by pallor of the disc and loss of conducting function of the optic nerve, resulting in decreased visual acuity or defects in visual field.

In this clinical study on optic atrophy a wide spectrum of etiological factors are found to be responsible for the development of optic atrophy, but glaucoma (28%) and retinitis pigmentosa (22%) are the commonest[5].

Optic atrophy due to glaucoma can be prevented by screening all individuals above 40 years for glaucoma and treating at the earliest evidence of damage to the nerve fibers. Visual loss in progressive cases can also be limited by effective management and thus blindness can be prevented[6].

In case of traumatic optic atrophy all individuals in this study are below 40 years and are due to road traffic accidents. Incidence of traumatic optic atrophy can be reduced by taking preventive measures like using protective helmets while driving, implementing and following strict traffic rules. We can limit the optic atrophy in case of optic nerve damage due to RTA by early diagnosis and effective medical or surgical management[7].

Retinitis pigmentosa being genetically transmitted and as there is no effective treatment for this till now, only its incidence should be reduced by genetic counseling, discouraging consanguineous marriages, limitation of number of children in individuals with this disease.

Early diagnosis and treatment of the etiological factors like intracranial tumors, meningitis, optic neuritis, and toxic atrophy can prevent or limit visual loss from optic atrophy.

In this clinical study on optic atrophy out of 50 cases, 39 cases are bilateral Optic atrophy. Hence requires visual rehabilitation.

In this clinical study on optic atrophy, 68% were economically blind i.e. visual acuity is less than 6/60 and requires rehabilitation

It is important to diagnose early and treat the cause of optic atrophy not only for better visual function but also for the life of the patient.

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