

## Study of Aetiology of Optic Atrophy

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### ABSTRACT

#### BACKGROUND

Optic atrophy refers to the late stage changes that take place in the optic nerve resulting from axonal degeneration in the visual pathway between the retina and the lateral geniculate body. There are various aetiologies and classifications of optic atrophy. Therefore, it is crucial to diagnose the aetiology and treat a case of optic atrophy at the earliest, not only for good visual outcome but also for the general health and good quality of life for the patient. We wanted to study the aetiology of optic atrophy.

#### METHODS

A prospective cross-sectional study was conducted in the Department of Ophthalmology, Sri Venkateswara Ramnarain Ruia Government General Hospital, attached to Sri Venkateswara Medical College, Tirupati. A total of 100 cases of optic atrophy fulfilling the inclusion criteria were examined as per protocol and results were analysed.

#### RESULTS

A total of 100 cases was included in the study. Mean age-group of the majority of the patients was 46.88±17.04 years; of these, 62% were males, and 38% were females. Out of 100 cases, 26 cases were due to primary optic atrophy, 35 cases were due to glaucoma, 28 cases were due to consecutive optic atrophy, and 11 cases were due to secondary optic atrophy.

#### CONCLUSIONS

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

#### KEYWORDS

Consecutive Optic Atrophy, Glaucomatous Optic Atrophy, Primary Optic Atrophy, Secondary Optic Atrophy

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## BACKGROUND

Optic Atrophy is the end result of various lesions of the visual pathways from the ganglion cell layer to the lateral geniculate body.<sup>1</sup> Optic atrophy is not a disease. It is a nonspecific morphologic endpoint of any disease that causes damage to the ganglion cells and axons of the optic nerve. Unlike the axons elsewhere in the central nervous system, the axons of retinal ganglion cells have virtually no capacity for regeneration. Shrinkage of the optic nerve is secondary to a loss of parenchyma.<sup>2</sup> This represents loss of retinal ganglion cell axons as well as the ensheathing myelin.<sup>3</sup> No treatment is effective for optic atrophy. The only solution is to control the risk factor where possible. Consequently, proper management of optic atrophy involves early diagnosis and investigation to find the cause wherever possible. Therefore, it is crucial to diagnose the aetiology 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.

Ophthalmoscopic classification of optic atrophy:<sup>4</sup>  
Primary/Secondary/Consecutive/Glaucomatous

### **Aetiological Classification:**<sup>5</sup>

1. Congenital atrophy
2. Consecutive atrophy: secondary to retinal disease and destruction of the ganglion cells.
  - a) Post-inflammatory- as after diffuse chorioretinitis: Ex. Post PRP.
  - b) Degenerative:
    - i. In pigmentary retinal dystrophy
    - ii. In myopia
3. Circulatory atrophy:
  - a) Occlusion of the central retinal artery or the internal carotid artery
  - b) Post-haemorrhagic
  - c) Arteriosclerotic, producing ischaemic optic neuropathy.
  - d) Pernicious anaemia
4. Pressure and Traction atrophy:
  - a) Glaucomatous atrophy
  - b) Oedema of the optic disc
  - c) Aneurysms of the internal carotid artery or neighbouring arteries.
  - d) Bony pressure at the optic foramen
  - e) Tumours, either of the optic nerve or its sheaths
  - f) Inflammatory adhesions, as in basal arachnoiditis.
  - g) Swelling of the optic nerve
  - h) Traction or pressure on the optic nerve in cases of endocrine exophthalmos.
5. Post-Inflammatory atrophy:
  - a) Optic neuritis or a perineuritis derived from the orbital tissues
  - b) Septicaemia
6. As a part of Central Nervous System disease: Multiple sclerosis, neuromyelitis optica, disseminated encephalomyelitis, zoster, encephalitis lethargic, tabes,

dementia paralytica, cerebral palsy, hereditary disease.

7. Metabolic disorders: occurrence of optic atrophy in diabetes of the juvenile type
8. Toxic atrophy: Tobacco, alcohol, lead and many others
9. Traumatic atrophy
10. Atrophy of unknown aetiology: Leber's optic atrophy, hereditary optic atrophy.

### **Differential Diagnosis of Optic Atrophy**

Coloboma of the disc/Optic disc pit/Morning glory syndrome/Medullated nerve fibres/Myopic disc/Hypoplasia of optic disc/Drusen of the disc.

We wanted to determine and study the aetiology of optic atrophy and classify optic atrophy based on ophthalmoscopic appearance.

## METHODS

The present study is a hospital-based cross-sectional study conducted between July 2017 and August 2018 in the Department of Ophthalmology, Sri Venkateswara Ramnarain Ruia Government General Hospital (SVRRGGH), Tirupati. In this study, a total number of 100 cases of optic atrophy were included. They were diagnosed by fundus examination. Later, data was collected by taking a detailed clinical history. Information was taken regarding the demographics, chief complaints, including the duration of the problem and presence of any systemic diseases. The subjects had undergone complete ophthalmological examination, i.e. anterior segment examination with the help of slit lamp and posterior segment examination with the help of direct and indirect ophthalmoscopy. Visual fields and colour vision were performed whenever required and in possible cases. CT scan and MRI of the brain and orbits were done to rule out intracranial space-occupying lesions.

### **Inclusion Criteria**

Patients diagnosed to have optic atrophy on ophthalmoscopic examination.

### **Exclusion Criteria**

Patients not willing to participate in the study.

### **Statistical Analysis**

The data were entered into MS Excel 2007, and basic statistical analysis was done using frequencies, proportions and percentages.

## RESULTS

A total of 100 patients who met the inclusion criteria are included in the present study, during the study period. In the present study, optic atrophy is seen in all age groups, most commonly between 4<sup>th</sup> and 6<sup>th</sup> decades. Mean age of

presentation in the present study is 46.88 ± 17.04 years. In the present study, out of 100 cases, 62 cases are males, and 38 cases are females. In the present study, 30 cases have unilateral optic atrophy, and 70 cases have bilateral optic atrophy. Out of 30 cases of unilateral optic atrophy in the present study, trauma (10 cases), glaucoma (9 cases), circulatory (7 cases) are the major causes, the other causes being post-inflammatory (2 cases), post-PRP (1 case) and papilloedema (1 case). Glaucoma (26 cases) and Retinitis pigmentosa (23 cases) are the major causes of bilateral optic atrophy, intracranial tumours and papilloedema are other causes.

Age-Group in Years	No. of Cases	%
1-10	1	1
11-20	8	8
21-30	12	12
31-40	21	21
41-50	12	12
51-60	22	22
61-70	18	18
>70	6	6
Total	100	100

Table 1. Age Distribution

Gender	No. of Cases	%
Male	62	62
Female	38	38
Total	100	100

Table 2. Gender Distribution

Laterality of Optic Atrophy	No. of Cases	%
Unilateral	30	30
Bilateral	70	70
Total	100	100

Table 3. Laterality of Optic Atrophy

Type of Optic Atrophy	No. of Cases	%
Primary	26	26
Secondary	11	11
Consecutive	28	28
Glaucomatous	35	35
Total	100	100

Table 4. Type of Optic Atrophy

Aetiology of Optic Atrophy	No. of Cases
<b>1. Pressure and traction atrophy</b>	54
Glaucomatous optic atrophy	35
Post Papilloedematous optic atrophy	06
Intracranial tumours without raised ICT	13
<b>2. Consecutive optic atrophy</b>	25
Retinitis pigmentosa	23
Post PRP	02
<b>3. Circulatory optic atrophy</b>	08
CRAO	03
AION	05
<b>4. Post-inflammatory</b>	02
<b>5. Traumatic optic atrophy</b>	10
<b>6. Idiopathic</b>	01
Total	100

Table 5. Distribution of Patients According to the Aetiology of Optic Atrophy

Abbreviations: ICT, intracranial tension; PRP, pan-retinal photocoagulation; CRAO, central retinal artery occlusion; AION, anterior ischaemic optic neuropathy.

Out of 100 cases of optic atrophy, 35% has glaucomatous optic atrophy, 28% has consecutive optic atrophy, 26% has primary optic atrophy, and 11% has secondary optic atrophy. Out of the etiologically established cases of optic atrophy, there is ocular pathology in 80 cases. Glaucoma and retinitis pigmentosa are the most common ocular causes leading to optic atrophy. There is extraocular pathology in 19 cases. Intracranial tumours and

papilloedema constituted the most common extraocular causes for optic atrophy. Out of the 170 eyes of 100 patients with optic atrophy, 14 eyes have visual acuity of No PL (perception of light) at presentation, for which pressure and traction atrophy (glaucoma, papilloedema) is the major cause. 62 eyes have visual acuity of less than CF 1 mt to PL, for which pressure and traction atrophy (glaucoma, papilloedema), and retinitis pigmentosa are the predominant causes.

Aetiology	VA: 6/6-6/18	VA: 6/24-6/60	VA: <6/60- CF 1 mt	VA: <CF 1 mt-PL	VA: No PL	Total
Pressure & traction atrophy	07	19	30	34	08	98
Consecutive atrophy	-	06	22	21	-	49
Circulatory	02	01	03	02	01	09
Post-inflammatory	-	-	02	-	-	02
Traumatic atrophy	-	-	-	05	05	10
Idiopathic	-	-	02	-	-	02
Total	09	26	59	62	14	170*

Table 6. Aetiology in Relation to Visual Acuity at Presentation for Both Eyes

(\* includes total eyes with Optic atrophy)  
Abbreviations: VA, visual acuity; PL, perception of light

## DISCUSSION

### Age Analysis

In the present study, the maximum number of cases are reported in the age-group of 51-60 years (22%), followed by 31-40 years (21%), 61-70 years (18%) and 1-10 years (1%). Mean age in the present study is 46.88±17.04 years, and this is in accordance with the study conducted by Bajracharya K et al<sup>6</sup> study where the mean age was reported as 53.16±18.1 years. In a study by T. S. Oluleye et al.,<sup>7</sup> it was 40.8 years.

### Gender Analysis

In the present study, out of 100 cases, 62 cases are males, and 38 cases are females, with a male to female ratio of 1.6: 1. This difference of gender with male preponderance was also documented in the Chaddah M Ret al<sup>8</sup> study, T. S. Oluleye et al<sup>7</sup> study and Kumar MP et al<sup>9</sup> study, with the male-female ratios being 1.9: 1, 2: 1 and 2: 1 respectively. The reason for the males being more affected with optic atrophy may be attributed to the X-linked recessive inheritance of Retinitis Pigmentosa. Road traffic accidents leading to traumatic optic atrophy may be another reason for the optic atrophy being more common among males.

### Optic Atrophy and Laterality

In this study, out of the 100 cases of optic atrophy, 70 cases are bilateral, and 30 cases are unilateral affecting either of the right or the left eye. This is in accordance with Chaddah M R et al<sup>8</sup> study, T. S. Oluleye et al<sup>7</sup> study and Kumar MP et al<sup>9</sup> study.

### Aetiology of Optic Atrophy

In the present study, all causes of optic atrophy are in accordance with Krishna VM et al<sup>10</sup> study except in Idiopathic

causes. In the present study, Idiopathic causes constitute 1%, but in Krishna VM et al<sup>10</sup> study it was 4%. In the present study, congenital or hereditary causes of optic atrophy are not reported. The probable reason is that these cases have accounted for a few of the cases in the Idiopathic group. Of the 99% cases in which aetiology has been established, 80% of cases have ocular causes, 19% of cases are due to extraocular causes. In 1 case, the cause of optic atrophy could not be determined.

### Optic Atrophy in Glaucoma

In the present study, glaucomatous optic atrophy is seen in 35 patients out of whom 22 are males, 13 are females. Out of the 35 cases, 24 cases are of Primary open-angle glaucoma, 8 are of Chronic angle-closure glaucoma, 2 are of Developmental glaucoma, and 1 is Pseudoexfoliation glaucoma. In the present study, out of these 35 cases of glaucomatous optic atrophy, 91.4% are above 40 years of age. Usually, glaucomatous optic atrophy peaks between 6<sup>th</sup> and 7<sup>th</sup> decades. This is in accordance with the study conducted by Bajracharya K et al,<sup>6</sup> where it was 93.1% in this age group. Out of 70 eyes of 35 patients, 32 eyes have IOP between 21 to 30 mmHg, 36 eyes have IOP between 31 to 40 mmHg, and 2 eyes had IOP between 41 to 50 mmHg. In the present clinical study, glaucoma is the cause of optic atrophy in 35% of cases. This is in accordance with Krishna VM et al<sup>10</sup> study where it was 28%. This finding is not in concordance with Kumar MP et al<sup>9</sup> study and Bajracharya K et al<sup>6</sup> study where it was 16% and 58% respectively.

### Optic Atrophy in Retinitis Pigmentosa

Retinitis Pigmentosa (RP) is found to be the cause of optic atrophy in 23 cases, out of which 15 were males, and the remaining 8 were females. In the present study, optic atrophy due to RP peaked between 3<sup>rd</sup> and 4<sup>th</sup> decades and is consistent with the previous studies. In the present clinical study, retinitis pigmentosa is found to be the cause of consecutive optic atrophy in 23% of cases. This is in accordance with Krishna VM et al<sup>10</sup> study where it was 22%. This finding was not in concordance with T. S. Oluleye et al<sup>7</sup> study where it was 3%.

### Optic Atrophy and Trauma

In this study, trauma is the cause of optic atrophy in 10% of cases. This is in accordance with Chaddah M R et al<sup>8</sup> study, T. S. Oluleye et al<sup>7</sup> study and Krishna VM et al<sup>10</sup> study where it was 7%, 8% and 6% respectively. However, in Kumar P et al<sup>9</sup> study, it was reported as 2%.

### Optic Atrophy and Intracranial Tumours

In this study, there are 15 cases of intracranial tumours causing optic atrophy, of which 13 cases are due to pituitary adenoma, and 2 cases are due to meningioma of the sphenoid. In the present study, 15% of cases are due to intracranial space-occupying lesions. This is in accordance with Chaddah M R et al<sup>8</sup> study and Krishna VM et al<sup>10</sup> study where it was 12% and 8% respectively. This finding was not in concordance with Bajracharya K et al<sup>6</sup> study and Kumar P et al<sup>9</sup> study where it was 4% and 2% respectively.

### Circulatory Optic Atrophy

In the present study, 8% of cases are due to circulatory causes. This is in accordance with Krishna VM et al<sup>10</sup> study where it was 4%. Out of the 8 cases, 5 cases are due to AION, and 3 cases are due to CRAO.

### Strengths of the Present Study

A wide spectrum of the aetiological factors of optic atrophy, which is the end result of various lesions of the visual pathways, was described in this hospital-based study.

### Limitations of the Present Study

Though the sample size is appropriate for describing the aetiologies of optic atrophy, further studies with larger samples can be conducted to determine the significant associated factors with optic atrophy.

### Recommendations from the Present Study

- 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 fibre layer.
- Incidence of Retinitis pigmentosa should be reduced by genetic counselling, discouraging consanguineous marriages and limitation of the number of children in individuals with this disease.
- Early diagnosis and treatment of the aetiological factors like intracranial tumours are needed to prevent subsequent optic atrophy.
- Incidence of traumatic optic atrophy can be reduced by taking preventive measures like using protective helmets while driving, implementing and following strict traffic rules.
- As treatment is not effective once optic atrophy sets in, early diagnosis and timely management of underlying cause is the critical factor to prevent optic atrophy.
- As optic atrophy causes irreversible blindness, patients require visual rehabilitation.

### Summary

1. In this study, optic atrophy was seen in all age groups, but it is less in extremes of age and peaks between 4<sup>th</sup> and 6<sup>th</sup> decades.
2. 70 cases of optic atrophy were bilateral, and 30 cases are unilateral.
3. It is found that out of 100 cases of optic atrophy, 35% are due to glaucomatous, 28% due to consecutive, 26% due to primary and 11% due to secondary optic atrophies.
4. Glaucoma and Retinitis pigmentosa are the commonest ocular causes of optic atrophy.
5. Intracranial tumours and papilloedema constituted the common extraocular causes of optic atrophy.
6. For unilateral optic atrophy, trauma and glaucoma are the major causes and for bilateral optic atrophy, glaucoma and retinitis pigmentosa are the major causes.
7. Out of the 170 eyes of 100 patients with optic atrophy, 14 eyes has visual acuity of No PL (perception of light)

at presentation, for which pressure and traction atrophy (glaucoma, papilloedema) is the major cause, for the 62 eyes which has visual acuity of less than CF 1 mt to PL, retinitis pigmentosa is the predominant cause.

### CONCLUSIONS

Optic atrophy is the end result of various lesions of the visual pathways from the ganglion cell layer to the lateral geniculate body. In this clinical study on optic atrophy, a wide spectrum of aetiological factors is found to be responsible for the development of optic atrophy, but glaucoma (35%) and retinitis pigmentosa (23%) are the commonest causes. 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 fibre layer. Visual loss in progressive cases can also be limited by effective management, and thus blindness can be prevented. Retinitis pigmentosa being genetically transmitted and as there is no effective treatment for this till now, its incidence can be reduced by genetic counselling, discouraging consanguineous marriages and limiting the number of children in individuals with this disease. In the present study, all cases of traumatic optic atrophy are below 40 years of age, and the trauma is due to road traffic accidents in all these cases. 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 also prevent optic atrophy in cases of optic nerve damage due to RTA (road traffic accidents) by early diagnosis and effective medical or surgical management. Early diagnosis and treatment of the aetiological factors like intracranial tumours, meningitis and optic neuritis can prevent or limit visual loss from optic atrophy. Out of the 170 eyes of 100 patients with optic atrophy, 135 eyes had BCVA of less than 6/60, which comes under the category of economical blindness. These patients

require economic rehabilitation. Hence it is important to diagnose early and treat the cause of optic atrophy not only for the better visual function of affected patients but also to reduce the economic burden on the families of these patients.

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