A study on ocular manifestations of skin disorders- At a tertiary care centre

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A B S T R A C T

Aim & Objective: To study the prevalence, ocular manifestations and also analyse the frequency and distribution of the potential sight threatening lesions in the various type of common skin disorders at tertiary care centre.

Materials and Methods: 40 patients were studied in the Department of ophthalmology from January 2019 to December 2019.

Results: Of the 40 patients that were included in our study,12(30%) had Neurofibromatosis type-1,10(25%) patients had Psoriasis,8(20%) patients had Steven-Johnson syndrome,6(15%) patients had herpes Zoster ophthalmicus and 8(20%) patients ocular cicatricial pemphigoid. Neurofibromatosis type-1 was found to be more common among males and 50% patients presented within the age group of 21-40 years of age. Lisch nodules were seen in all the patients with neurofibromatosis type-1 and nodular neurofibromas were seen 91.7% patients. Conjunctival freckles and sphenoid wing dysplasia were seen in 1 patient. Patients with Psoriasis showed a male predominance accounting for 60% whereas female accounted for 40%.the commonest age group affected by psoriasis 21-40 years seen in 60% of the patients. Among the patients with Psoriasis, blepharoconjunctivitis was the most common manifestation seen in 5(50%) patients followed by keratoconjunctivitis sicca in 3(30%) patients and chronic anterior uveitis in 2(20%) patients. Male preponderance was noted in patients with Steven-Johnson syndrome and majority (62.5%) of the patients were in the age group of 21-40 years. Bilateral, symmetrical involvement was seen in 87.5% of patients with Steven-Johnson syndrome.Drugs were the most common etiological agent seen in 62.5% of patients. Lid manifestations were the most common among those with Stevens-Johnson syndrome accounting for 62.5% patients. Males dominated the clinical scenario among the patients with herpes zoster ophthalmicus accounting for 66.67% of patients with a peak during 21-40 years of age. Preceding the eruption, pain was the commonest prodromal symptom seen in 50% patients followed by burning sensation and fever. Females were affected in 75% patients with ocular cicatricial pemphigoid and they presented beyond 60 years of age with conjunctivitis in 50% patients followed by entropion and trichiasis.

Conclusion: We conclude that, ocular involvement in skin disease is an common feature could be major component for the development of various systemic skin disorders.

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1. Introduction

Ocular involvement in skin disease is a common feature and could be a component of systemic disease as well. The spectrum of involvement of the eye is varied and is largely, dependent on the dermatological disease. This overlap needs to be examined to throw further insight on the nature of the problem as both the ophthalmologist and dermatologist could work in concurrence treating the same. dermatological diseases causing ocular involvement could be due to infections or autoimmune diseases. The reason for this association is multifactorial and will be analysed subsequently. We studied 40 patients with common skin disorders in a tertiary care centre.
1.1. Neurofibromatosis

Neurofibromatosis is a disorder that primarily affects the cell growth of neural tissues. Eyelid neurofibromas tend to develop early in life. When it involves the upper lid, it frequently causes mechanical ptosis. Plexiform neurofibromas cause an “S” shaped deformity of the upper lid.

1.2. Psoriasis

It is a common, chronic, disfiguring, inflammatory and proliferative condition of the skin. In which genetic and environmental influences have a critical role. It is equally common in males and females. It is more likely to appear between the ages of 15-30 yrs.

Ocular lesions such as blepharitis and mucoid conjunctivitis are common. Keratitis in the form of raised, marginal, corneal infiltrates is known to occur. Symblepharon and trichiasis have been reported. Of all the ocular features, conjunctivitis is the commonest ocular manifestation. In majority of the patients conjunctivitis follows the onset of psoriasis. In few cases, it is known to precede the disease.

1.3. Stevens-Johnson Syndrome

It is acute life threatening mucocutaneous reactions characterized by extensive necrosis and detachment of the epidermis.

1.4. Herpes Zoster

Varicella zoster virus is member of the herpes virus family. During the course of varicella, vericella zoster virus passes into the contiguous endings of sensory nerves and is transported centripetally up the sensory fibres to the sensory ganglia. In the ganglia, the virus establishes a latent infection that persists for life.

1.5. Ocular cicatrical pemphigoid

Cicatrical pemphigoid is a rare chronic autoimmune sub epithelial blistering disease characterized by erosive lesions of the mucous membrane and skin that results in scarring.

Patients typically describe the onset of painful, erosive, and/or blistering lesions on one or more mucosal surfaces. The two most frequently involved sites in these patients are the oral and conjunctival mucosae.

Besides, the ophthalmologist plays a crucial role in identifying the ocular manifestations and administering prompt treatment as, in several situations these dermatological diseases may resolve but the ophthalmic complications that develop may persist and cause severe visual loss and blindness.

2. Aims and Objectives

1. To study the prevalence ocular manifestations of common skin disorders in a tertiary care centre
2. To study the frequency and distribution of the potential sight threatening lesions in the various type of skin disorders.

3. Materials and Methods

3.1. Study type

It was an observational Prospective study

3.2. Study population & duration of the study

The materials for this study were collected from outpatients and inpatients of ophthalmology and dermatology departments from January 2019 to December 2019. 4 patients who were referred to the Department of ophthalmology from the department of dermatology and venereology were accepted. Different types of skin diseases were seen, five were studied in detail.

Methodology A preliminary ophthalmic examination was carried out with oblique illumination and slit lamp to assess the ocular involvement. External deformities and adnexal involvement were noted. Extraocular movements were assessed in all patients followed by a detailed check using slit lamp. Examination comprised of evaluation of the anterior segment, vitreous and posterior segment. Indirect ophthalmoscopy and bio microscopy was performed on all patients to examine for retinal involvement. Intraocular tension was recorded with Goldmann applanation tonometer. Other investigations like corneal staining and gonioscopy was done wherever situation demanded. Fundus photographs were taken if retinal involvement was present.

Conservative (medical) management for the patients with ocular involvement was advised. Conservative management included topical cycloplegics, topical steroids, topical antibiotics artificial tears and taping of lids.

4. Results and Analysis

Of the 40 patients that were included in our study, 12 (30%) had neurofibromatosis type-1, 10 (25%) patients had psoriasis, 8 (20%) patients had stevens-Johnson syndrome, 6 (15%) patients had herpes zoster ophthalmicus and 4 (10%) patients had cicatrical pemphigoid (Table 1).

4.1. Neurofibromatosis Type 1

Of 12 patients with NEUROFABROMATOSIS TYPE 1.5(41.67%) were males and 7(58.33%) were females (Table 2)

Among the 12 patients with NEUROFABROMATOSIS TYPE 1.6(50%) patients belonged to the age group of <20
Table 1: Distribution of Skin Disorders

| Primary diagnosis                  | No. Of patients | Percentage |
|-----------------------------------|-----------------|------------|
| Neurofibromatosis                 | 12              | 30%        |
| Psoriasis                         | 10              | 25%        |
| Stevens-Johnson Syndrome          | 8               | 20%        |
| Herpes Zoster Ophthalmicus        | 6               | 15%        |
| Ocular Cicatrical Pemphigoid      | 4               | 10%        |

Table 2: Sex distribution in patients with NEUROFABROMATOSIS TYPE 1

| Sex        | No. of patients | Percentage |
|------------|-----------------|------------|
| Males      | 5               | 41.67      |
| Females    | 7               | 58.33      |

Of the 12 patients with NEUROFABROMATOSIS TYPE 1 in our study we found that 1(8.3%) patient had plexiform neurofibroma and nodular lesions were found in 11 (91.7%) patients.

Table 3: Age distribution in patients with NEUROFABROMATOSIS TYPE 1

| Age group | No of patients | Percentage |
|-----------|----------------|------------|
| <20 years | 3              | 25%        |
| 21-30 years | 6          | 50%        |
| 41-60 years | 2          | 16.67%     |
| >60 years  | 1              | 8.3%       |

4.2. Psoriasis

Out of the 10 patients with psoriasis, 6(60%) patients were males and 4(40%) patients were females (Table 4)

Of the 10 patients with psoriasis, the commonest age group affected was 21-40 years (Table 5).

Table 4: Sex distribution in patients with psoriasis

| Sex        | No. of patients | Percentage |
|------------|-----------------|------------|
| Males      | 6               | 60%        |
| Female     | 4               | 40%        |

Table 5: AGE distribution in patients with psoriasis

| Age group | No of patients | Percentage |
|-----------|----------------|------------|
| <20 Years | 1              | 10%        |
| 21-20 Years | 6           | 60%        |
| 41-60 Years | 2           | 20%        |
| >60 Years  | 1              | 10%        |

Among the 10 patients with psoriasis, blepharonjunctivitis was found to be the most common manifestation seen in (50%) patients. 3 (30%) patients had keratoconjunctivitis sicca and 2 (20%) patients had chronic anterior uveitis (Figure 2)

4.3. Stevens-Johnson Syndrome

Out of 8 patients with Stevens-Johnson Syndrome, 5(62.5%) patients were males and 3 (37.5%) patients were females (Table 6).

Table 6: Sex distribution in patients with Stevens-Johnson Syndrome

| Sex        | No. of patients | Percentage |
|------------|-----------------|------------|
| Males      | 5               | 62.5%      |
| Female     | 3               | 37.5%      |

The following table shows the age distribution of patients with STEVENS-JOHNSON SYNDROME (Table 7).

Table 7: Age distribution in patients with Stevens-Johnson Syndrome

| Age group | No of patients | Percentage |
|-----------|----------------|------------|
| <20 Years | 1              | 12.5%      |
| 21-20 Years | 6           | 62.5%      |
| 41-60 Years | 2           | 25%        |
| >60 Years  | -              | -          |

All patients with Stevens-Johnson Syndrome had bilateral involvement. Bilateral symmetrical involvement was seen in 7 (87.5%) patients and bilateral, asymmetrical involvement was found in 1(12.5%) patient (chart 4)

In our study, the most common etiological agent for Stevens-Johnson Syndrome was drugs, found in 5 (62.5%) patients. 2(25%) patients gave a previous history of viral fever with no history of drug injection. No definitive cause was ascertained in 1(12.5%) patient (Table 8).

On slit-lamp examination, the lid, conjunctiva and corneal complications were seen in 5 (62.5%), 2 (25%) and 1 (12.5%), respectively among the 8 patients with Stevens-Johnson Syndrome.
4.4. Herpes zoster ophthalmicus

Of the 6 patients with Herpes Zoster Ophthalmicus, 4 (66.67%) patients were males and 2 (33.33%) patients were females (Table 9).

Table 9: Sex distribution in patients with Herpes Zoster Ophthalmicus

| Sex     | No of patients | Percentage |
|---------|----------------|------------|
| Males   | 4              | 66.67%     |
| Females | 2              | 33.33%     |

The table below shows the age distribution of patients with HZO in our study (Table 10).

Table 10: Age distribution of patients with Herpes Zoster Ophthalmicus

| Age group | No of patients | Percentage |
|-----------|----------------|------------|
| <20 Years | 1              | 16.67%     |
| 21-20 Years | 3            | 50%        |
| 41-60 Years | 2            | 33.33%     |
| >60 Years | -              | -          |

In our study, pain was the commonest prodromal symptom noticed in 3 (50%) patients, followed by burning sensation in 2 (33.33%) patients and only 1 (16.67%) patient experienced fever.

All patients with HERPES ZOSTER OPHTHALMICUS had unilateral involvement. No definite bilaterality was noticed in our study.

4.5. Ocular cicatricial pemphigoid

Of 4 patients with Ocular Cicatricial Pemphigoid, 3 (75%) patients were females and 1 (25%) patient was male (Table 11).

Table 11: Sex distribution in patients with ocular cicatricial Pemphigoid

| Sex     | No of patients | Percentage |
|---------|----------------|------------|
| Males   | 1              | 25%        |
| Females | 3              | 75%        |

The following table shows the age distribution of patients with ocular cicatricial pemphigoid in our study (Table 12).

Table 12: Age distribution in patients with Ocular Cicatricial Pemphigoid

| Age group | No of patients | Percentage |
|-----------|----------------|------------|
| <20 Years | -              | -          |
| 21-20 Years | -            | -          |
| 41-60 Years | 1            | 25%        |
| >60 Years | 3              | 75%        |

Among 4 patients with ocular cicatricial pemphigoid we found that 2 (50%) presented with conjunctivitis, 1 (25%) patient presented with entropion and 1 (25%) patient presented with entropion and trichiasis.

5. Discussion

40 Patients were studied in the Department of ophthalmology from January 2019 to December 2019. Of the 40 patients, 12 (30%) had Neurofibromatosis type-1, 10 (25%) patients had Psoriasis, 8 (20%) patients had Steven-johnson syndrome, 6 (15%) patients had herpes zoster ophthalmics and 4 (10%) patients had ocular cicatrical pemphigoid.

5.1. Neurofibromatosis type-1

Of the 12 patients with Neurofibromatosis type-1, 5 (41.67) were males and 7 (58.33%) were females. In a study done by Huson sm and Harper, there was a slight female preponderance. This is in contrast to a study conducted by Odehode, in which there was a definite male preponderance affecting 60 males and 38 females with a total of 98 patients.

Among the 12 patients with neurofibromatosis type -1 in our study, 6 (50%) patients belonged to the age group of 20-40 years, 3 (25%) patients belonged to the age group of ≤ 2 years, 2 (16.67%) patients belonged to the age group of 41-60 years and 1 (8.3%) patient was > 60 years of age. The commonest age group with which patients presented to us was between 20-40 years. On comparing this with the study done by Genet et al, the commonest age group affected was > 20 years of age. Of the 12 patients, with Neurofibromatosis type-1, in our study we found that 1 (8.3%) patient had plexiform neurofibroma and nodular lesions were found in 11 (91.7%) patients. However, a study
was conducted by ALO and Massobrio showed that 20% of the patients had plexiform neurofibromas.\textsuperscript{9}

Lisch nodules were found to be the commonest manifestation among all patients with neurofibromatosis type 1, in our study. This finding was supported by the studies done by Huson et al,\textsuperscript{10} Riccardi\textsuperscript{11} and by Goeker\textsuperscript{12} rare findings such as conjunctival freckles and sphenoid wing dysplasia were seen in 1 patient as found earlier in a study conducted by premalatha.\textsuperscript{13} A similar study was done by Mirowitz et al and it confirmed the above findings.\textsuperscript{14}

5.2. Psoriasis

Of the 10 patients with Psoriasis, 6 (60%) patients were males and 4 (40%) were females giving a male to female ratio of 1.5:1. This finding was found to be in concordance with a German study done by Henseler et al,\textsuperscript{15} where the ratio 1.74:1. Smith AE also noted the male preponderance.\textsuperscript{16} The age of the patients ranged from 16 years to 64 years. The commonest age group affected was 21 — 40 years followed by 41 — 60 years. Farber and Nall\textsuperscript{1} found that the average age of onset was 28 years, while in the study on psoriasis done in China by YuiYie,\textsuperscript{17} the average age onset was 36 years. In a recent U.K based study done by Nevitt and Hutchinson,\textsuperscript{18} the mean age of onset was 3 years with the mode in the second decade. This is in contrast to a study done by LomhoIt\textsuperscript{19} who reported the average age of onset as 12 years.

Among the 10 people with psoriasis, in our study we found blepharoconjunctivitis to be the most common manifestation seen in 5 (50%) patients followed by keratoconjunctivitis sicca. Only 2 (20%) patients had chronic anterior uveitis. Catsaru-Catsari\textsuperscript{20} found that blephariconjunctivitis and keratoconjunctivitis sicca were the most common ocular manifestations of psoriasis however, offers et al found in their study that uveitis and blepharitis are the most common ocular manifestation of psoriasis followed dry eye.\textsuperscript{21}

5.3. Stevens- Johnson syndrome

in our study, out of 8 patients with Stevens- Johnson syndrome, 5 (62.5%) patients were males 3 (33.5%) patients were females. We found a male preponderance in our study. This is in concordance with the study done by Letko et al\textsuperscript{22} in 2005 in which they noted a clear male dominance. However, this is an contrast to a study done on ophthalmic complications and management of Stevens Johnson Syndrome by Kompella,\textsuperscript{23} in which they found a female predominance.

The common age group affected with Stevens-Johnson syndrome is between 21-40 years our study. The study done by Kompella also found that majority of the Patients (55.78%) were between 20 and 40 years of age.\textsuperscript{23} All patients with Stevens-johnson syndrome had bilateral involvement. Bilateral, symmetrical was CCI m 7 (87.5%) patients and bilateral, asymmetrical involvement was found in 1 (12.5) Patient. In the study done by Kompella all the patients with Stevens Johnson syndrome had bilateral involvement and most had bilateral, symmetrical presentation,\textsuperscript{23} a study done by Wilkins J and Morrison L also showed similar findings.\textsuperscript{24}

In our study, the most common etiological agent for Stevens- Johnson syndrome was drugs found in 5(62.6%) patients.2(25%) patients gave a previous history of viral fever with no history of drug intake, no definitive cause was ascertained in 1(12.5%) patient. The most commonly identified possible causative factor was drugs in 52 (51.89%) patients, 3(3.15%) patients had a history of viral fever preceding the onset and no definitive causative factor was identified in 37 (38.84%) patients according to the study done by Kompella.\textsuperscript{23} Drugs were found to be the most common etiological agent in the study by Letko E et al and Wilkins J and Morrison also.\textsuperscript{24}

In our study, the patients with Steven Johnson syndrome on slit lamp examination; the complications seen on the lids (62.5%) were crusting, entropion, and trichiasis. The conjunctival complications (25%) found were conjunctivitis and symblepharon cratopathy was seen in 12.5% due to entropion and trichiasis. In the study done by Letko E et al and Kompella,\textsuperscript{23} the lid abnormalities were observed in 87 (91.51%), conjunctival abnormalities in 92 (6.84%) and corneal complications in 93 (97.89%).\textsuperscript{23}

5.4. Herpes zoster ophthalmicus

Of the 6 patients with Herpes Zoster Ophthalmicus, 4 (66.67%) patients were male and 2 (33.33%) were female giving a male to female ratio of 2:1. This found to be in concordance with a study done by Dubey et al in 2005 in South India where the ratio was 1.84:1.\textsuperscript{25}Chaudhary, Sehgal, Nigarn and Mathur also noted this, male preponderance.\textsuperscript{26—29}The commonest age of presentation among the patients with herpes zoster ophthalmicus in our study, was found to be between 21-40 years. 3 (50%) presented within 21-40 years of age followed by 2 (33.33%) patients who presented within 41-60 years of age and 1 (1.67%) was less than 20 years of age. In the study by Dubey et al, the mean age of presentation was 37.65 years with a range of 2-77 years in their study.\textsuperscript{25} Chaudhary found a high incidence in the 2nd and 3rd decade,\textsuperscript{26} whereas Sehgal found a high incidence in the 4th and 5th decade.\textsuperscript{27} In our study, pain was the commonest prodromal symptom noticed in 3 (50%) patients, followed by burning sensation in 2 (33.33%) patients and only 1 (16.67%) patient experienced fever before the eruptions. Dubey et al found 90.65% to have pain in the prodromal period followed by paresthesia in 23.36% and itching in
19.62%–63. No definite laterality was noted in our study. This is in concordance to most of the prevailing studies.25–28

5.5. Ocular cicatricial pemphigoid

Of the 4 patients with OCULAR CICATRIAL PEMPICHOID, 3 (75%) patients were female and 1 (25%) patient was male giving a female to male ratio of 3:1. Similar findings were seen in a study done by Egan CA and Yancey KB in which they had found a female to male ratio of 2.6:1.4 This is in contrast to a study done by John Chang & Peter in 2005, in which there was only a slight female preponderance.30

The commonest age group in which the patient ocular cicatricial pemphigoid presents was >60 years of age. Similar findings were noted by John H chang & peter J mc Cluskey in their study.30 Among 4 patients with ocular cicatricial pemphigoid. We found that 2 (50%) patients presented with conjunctivitis, 1 (25%) patient presented with entropion and 1 (25%) patient presented with entropion trichiasis. Similar findings were observed in the study done by Stephen foster in which he observed that conjunctival abnormalities were the most common manifestation among the patients presenting with ocular cicatrical pemphigoid. Chang and Peter found that lid abnormalities (62.5%) were more common.30

6. Conclusion

In the present study, we conclude;

1. 40 patients were studied in the Department of ophthalmology from January 2019 to December 2019.
2. Neurofibromatosis type-1 accounted for majority of cases followed by psoriasis.Steven-Johnson syndrome, ocular cicatrical pemphigoid and herpes zoster ophthalmicus.
3. Males were more common than females in all the condition except ocular cicatrical pemphigoid and neurofibromatosis type-1.
4. The commonest age group of presentation was between 21-40 years in all the conditions except among patients with ocular cicatrical pemphigoid in the patients presented above 60 years of age
5. The most common ocular manifestation in patients with psoriasis, Is blepharoconjunctis followed by keratonjunctivitis Sicca and chronic anterior uveitis.
6. Most patients with Stevens-Johnson syndrome had bilateral symmetrical involvement
7. The most common etiological agent for Stevens-Johnson syndrome was drugs followed by viral fever.
8. Among the patients with Stevens-Johnson syndrome, lid abnormalities were the most common presentation followed by conjunctival and corneal complications.
9. Conjunctivitis was the most common manifestation of ocular cicatricial pemphigoid followed by lid abnormalities
10. Nodular lesions were more common than plexiform neurofibromas
11. Lisch nodules were the most common ocular manifestation in patients with neurofibromatosis type -1
12. Rare findings such as conjunctival freckles and sphenoid wing dysplia were also seen
13. Preceding the eruptions, pain was the Commonest prodromal symptom followed by burning Sensation and fever among the patients with herpes zoster ophthalmicus.
14. No definite laterality was noted in herpes zoster ophthalmicus.
15. Ocular morbidity was seen to occur maximally in Stevens-Johnson syndrome in the form of symblepheron, entropion and trichiasis, thus obscuring vision.

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9. Conflict of Interest

None.

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