INTRODUCTION

Hair shaft abnormalities comprise a group of congenital or acquired disorders which involve the hair shaft ***(Smith et al., 2005)***.

The hair shaft is a unique structure in terms of its strength, resilience, and resistance to the environment. Any defects in this normal structure can lead to variations in physical properties, such as shape and strength. An alteration in these properties is clinically noticed as a change in strength (fragility, or inability to grow hair long), texture, appearance, and manageability ***(Mirmirani et al., 2011)***.

Abnormal hair shafts usually lack macroscopic features, which would enable easy diagnosis. Thus, the usual diagnostic method is light microscopy and every time about 50 hairs are plucked to decrease the risk of missing a hair with the characteristic abnormality ***(Itin and Fistarol, 2005)***.

A number of genetic disorders are characterized by hair abnormalities. Hair changes may be a significant finding or even the initial presentation of a syndrome giving the clue to the diagnosis, as in diseases such as trichothiodystrophy or Netherton syndrome ***(Furdon and Clark, 2003)***.

Hair shaft disorders usually lye into two broad categories. Those with increased fragility namely; trichorrhexis nodosa, monilethrix, pili torti, trichorrhexis invaginata and trichothiodystrophy, and those without increased fragility as;

pili annulati, woolly hair and uncombable hair syndrome ***(Mirmirani et al., 2011)***.

In a different classification system, hair shaft abnormalities are divided into four major categories including: (a) fracture (e.g. trichothiodystrophy, trichorrhexis invaginata); (b) irregularities (e.g. congenital hypotrichosis, loose anagen syndrome, uncombable hair, pili annulati, monilethrix, bubble Hair); (c) twisting (e.g. pili torti, woolly hair, trichonodosis); and (d) extraneous matter on the hair shaft (piedra, pediculosis, peripilar cast) ***(Whiting, 2006).***

Dermoscopy, also known as dermatoscopy, epiluminescence microscopy or incident light microscopy, is a relatively new noninvasive diagnostic technique for the in vivo observation of skin lesions ***(Lacarrubba et al., 2010)***.

Dermoscopes are modified magnifying devices that permit the visualization of pigmented structures or vessels in the epidermis and superficial dermis and generally employ x10 magnifications ***(Micali et al., 2011)***.

Although they have been used mainly in the diagnosis of pigmented skin lesions ***(Akay et al., 2010).*** Their use has been later applied for the diagnosis of non-pigmented skin lesions including skin tumors, inflammatory and infectious diseases ***(Zalaudek et al., 2006).***

More recently, dermoscopy has proven extremely useful in aiding the diagnosis and follow-up of hair and scalp diseases. The term "trichoscopy" was first suggested in 2006 by

Rudnicka and Olszewska for dermoscopy and or videodermoscopy of hair and scalp diseases ***(Rudnicka et al., 2006).***

The method was developed by groups of dermatologists directed by: Rudnicka in Poland, Tosti and Micali in Italy and Inui ***(Rudnicka et al., 2008). Inui (2011)*** in Japan. In 2004, Lacarrubba and coworkers ***(Lacarruba et al., 2004).***

Olszewska and Rudnicka(2005),were the first who described videodermoscopic features of alopecia areata.

First used videodermoscopy for evaluation of androgenic alopecia. Later in 2006, Ross and coworkers ***(Ross et al., 2006).*** Specified videodermoscopy features of different acquired hair and scalp diseases.

Moreover, in 2008 ***Rakowska and coworkers***, first showed usefulness of trichoscopy in diagnosing children with congenital hair shaft abnormalities. It was shown that this method is helpful in diagnosing monilethrix, Netherton syndrome and other genetic diseases. In view of the importance of diagnosing the congenital hair shaft abnormalities as they may serve as a clue for the associated genetic syndrome, we assume that studying the dermoscopic characteristics of the various genetic hair shaft abnormalities may represent a significant advance in this field ***(Rakowska et al., 2008).***

AIM OF THE WORK

The aim of this thesis is to evaluate the role of hand-held dermoscopy in assessing congenital hair shaft abnormalities in comparison to light microscopy.

Chapter (1)

DERMOSCOPY

A dermoscope is a non-invasive, diagnostic tool which visualizes subtle clinical patterns of skin lesions and subsurface skin structures not normally visible to the naked eye. Some dermoscopic patterns are observed consistently with certain diseases and these then could be used for their diagnosis. Hence, this office procedure may obviate the need for a skin biopsy for diagnosis and for follow-up. The facility of storage of images and the results being immediately available are added advantages. Basically, a dermoscope is functionally similar to a magnifying lens but with the added features of an inbuilt illuminating system, a higher magnification which can be adjusted, the ability to assess structures as deep as in the reticular dermis, and the ability to record images ***(Nischal and Khopkar, 2005).***

Over the past years, dermoscopy has been known by a variety of names, including skin surface microscopy, epiluminescence microscopy, incident light microscopy, dermatoscopy, and video dermatoscopy. The term "dermoscopy," however, first used by Friedman et al in 1991, currently enjoys the greatest international consensus ***(Argenziano et al., 2003).*** Therefore, this term was preferably applied throughout this study.

❖ History

The 1980s can be considered as the heyday of dermoscopy, with the definition of criteria for dermoscopy and the first Consensus Conference on Skin Surface Microscopy ***(Bahmer et al., 1990).***

However, the origin of the dermoscopic technique goes back as far as the 17th century, when Kohlhaus in 1663 first examined ungual matrix vessels under the microscope ***(Gilje et al., 1953)***.

Progress in dermoscopy went faster where ***Fritsch and Pechlaner*** in ***1981*** distinguished benign from malignant skin lesions according to the characteristics of the pigmented net of the lesions.

Pehamberger et al. in ***1987*** introduced the analysis of patterns for the diagnosis of pigmented cutaneous lesions.

Soyer et al. (1989) established a correlation between dermoscopic and histopathologic structures.

In the same year, the 1st Consensus Conference on Skin Surface Microscopy was held in Hamburg, Germany where a terminology for dermoscopy was defined.

In ***1990 Kreusch and Rassner*** published the first Dermoscopy Atlas.

From the 1990s onwards several dermatology research groups developed several different diagnostic methods for analyzing dermoscopic images. At the same time hand held dermatoscopes became widely commercially available for purchase enabling an increased uptake in their use around the world ***(Peris et al., 2002)***.

From the year 2000 to the present, interest in this technology has been increasing, with the global spread of dermoscopy and the production of several courses, books, publications, and symposia on the theme, as well as the founding of the International Society of Dermoscopy ***(Rezze et al., 2004).***

In 2001, a California medical device manufacturer, 3Gen, introduced the first polarized dermoscope, the DermLite. Polarised illumination, coupled with a cross-polarised viewer, reduces (polarised) skin surface reflection, thus allowing visualisation of skin structures (the light from which is depolarised) without using an immersion fluid. Examination of several lesions is thus more convenient because physicians no longer have to stop and apply immersion oil, alcohol, or water to the skin before examining each lesion. With the marketing of polarised dermoscopes, dermoscopy increased in popularity among physicians worldwide. Although images produced by polarised light dermoscopes are slightly different from those produced by a traditional skin contact glass dermoscope, they

have certain advantages, such as vascular patterns not being potentially missed through compression of the skin by a glass contact plate ***(Peris et al., 2002).***

❖ Principle

The basic principle of dermoscopy is transillumination of a lesion and studying it with a high magnification to visualize subtle features ***(Stolz et al., 1994).*** Normally, light is reflected, dispersed or absorbed by the stratum corneum due to the differences in refractive index and optical density between skin and air. Thus, the skin appears opaque and the underlying structures cannot be adequately examined ***(Kenet et al., 1993).***

Most of the light incident on dry, scaly skin is reflected, but smooth, oily skin allows most of the light to pass through it, reaching the deeper dermis. This principle has been harnessed to improve the visibility of subsurface skin structures by employing application of linkage fluids over the lesions to be studied to improve the translucency of the skin (Figure1). Various linkage fluids used are oils (immersion oil, olive oil and mineral oil), water, antiseptic solutions and glycerin. Immersion oil is not used anymore because it contains chlorinated paraffin and dibutyl phthalate which have teratogenic, fetotoxic, and carcinogenic effects ***(Nischal and Khopkar, 2005).***

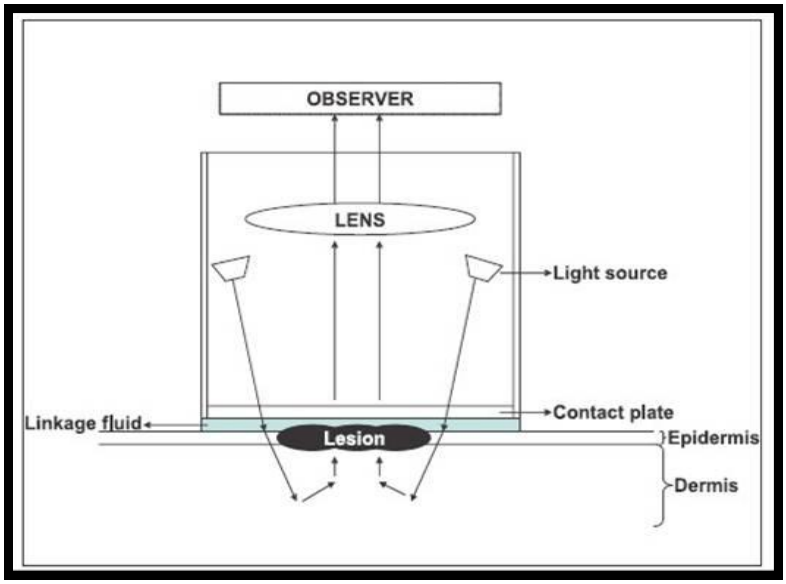

Figure (1): Optics of dermoscope ***(Nischal and Khopkar, 2005).***

Water or antiseptic solutions evaporate quickly and hence are less preferred than oils. Liquid paraffin has been used, which is inexpensive, safe and easily available, with good results. Glass has a refractive index (1.52) similar to that of skin (1.55) and hence when placed over oil-applied skin, further enhances transillumination of the lesion ***(Rabinovitz, 1998).***

Alcohols, due to their low viscosity, amphipathic properties, disinfectant capabilities and most importantly image clarity, are the best immersion liquids to use. 70% ethanol is chosen in offices because it has fewer odors than isopropanol and does not leave crystal deposits after evaporation like alcoholic disinfectants containing chlorhexidine ***(Katz and Rabinovitz, 2001).***

New hand-held dermoscopes are now available on the market that are provided with polarized light, rendering the fluid placed on the lesion unnecessary for inspecting pigmented skin structures ***(Nischal and Khopkar, 2005).***

❖ The essential components of a dermoscope are:

a) Achromatic lens: Most instruments provide 10 x magnifications, but higher magnifications can be achieved with special lenses ***(Pehamberger et al., 1987).***

b) Inbuilt illuminating system: Halogen lamps, which are oriented at an angle of 20 degree, are placed within the handheld piece. The colour contrasts of lesions are altered by the yellow light of halogen lamp. Light emitting diodes (LED) (Delta 20©, DermLite©) provide high intensity white light and consume 70% less power than halogen lamps. Illumination can be altered by turning off a set of LEDs. They are also designed to emit lights of different colors for better visualization of the skin as penetration of the skin by light is proportional to the wavelength of light (DermLite MS©) ***(Nischal and Khopkar, 2005).***

c) Power supply: Handheld instruments are usually powered by batteries, e.g. lithium ion battery, rechargeable lithium battery, AA battery, and rechargeable handles ***(Mayer, 1997).***

Additional facilities in some dermoscopes are an inbuilt photography system either an attachable conventional or digital camera or an inbuilt camera, and supporting software, for the capture, storage, retrieval and even interpretation of images ***(Kittler et al., 2002).***

❖ Types Of dermoscopy instruments:

Marghoob et al. (2003) have exhaustively reviewed various models of dermoscopes For simplicity, dermoscopic instruments can be grouped as:

a) **Dermoscopes without image capturing facility:** A dermoscope is a hand-held, otoscope-like instrument that lacks an inbuilt camera or any other image capture facility. However, cameras can be attached to some of these instruments with an adaptor, e.g. DermoGenius Basic©. Dermalite MS© (multispectral) incorporates four different colored polarized light. White, blue (surface pigmentation), yellow (superficial vessels), and red (deep pigment and vessels). Other models of this type are Delta 10©, Mini 2000 Dermooscope©, Episcope©.

b) **Dermoscopes with image capturing facility:** These instruments have either an inbuilt image capture system or have a camera attached for dermoscopic photography. Also, whole body photography (body mapping) is possible with an apparatus like Mole Max I. Other instruments of

this category are Delta 20© **(Figure2a)**, Dermascope© **(Figure2b)**, Dermaphot© **(Figure2c)**, Dermlite Foto© and Video episcope©. Both clinical and dermoscopic pictures of 10x magnification can be taken. A videodermatoscope (Video dermascope©, Videocap 100©) has a high resolution camera fitted to the hand piece. The image is seen on the computer screen and small videos can be taken with this instrument.

c) **Dermoscope with image capture facility and analytical capability:** These instruments are mainly used in western countries where the concern of melanomas is a driving force to improve dermoscopes for clinical diagnosis and preoperative assessment of pigmented lesions. Archived images of the patient can be compared with new ones and any significant change in lesion produces different colour signals, e.g. DermoGenius MoleMap©, Fotofinder dermoscope©, Molemax II©.

(a) **(b)** **(c)**

Figure (2): Dermoscopes with image capturing facility ***(Nischal and Khopkar, 2005).***

Uses of dermoscopy

Dermoscopes are largely used in white skinned individuals for the study of melanocytic nevi and melanoma. However, it can be used to diagnose other conditions too, e.g. psoriasis, lichen planus, dermatofibroma, Darier's disease, cicatricial alopecia, seborrheic keratosis and urticarial vasculitis. It can be also used in calculation of the follicular density in the donor area before follicular unit hair transplantation ***(Nischal and Khopkar, 2005).*** It is also used in monitoring adverse effects of potent topical corticosteroids in psoriasis ***(Vazquez-Lopez and Marghoob, 2004).***

1. Dermoscopy in inflammatory skin diseases

The most significant dermoscopic vascular pattern of psoriasis are red dots and globules, arranged in a homogenous, regular or ring-like fashion(dermoscopically known as red globular rings) in a light-red background **(Figure 3)** ***(Vazquez-Lopez et al., 2010).***

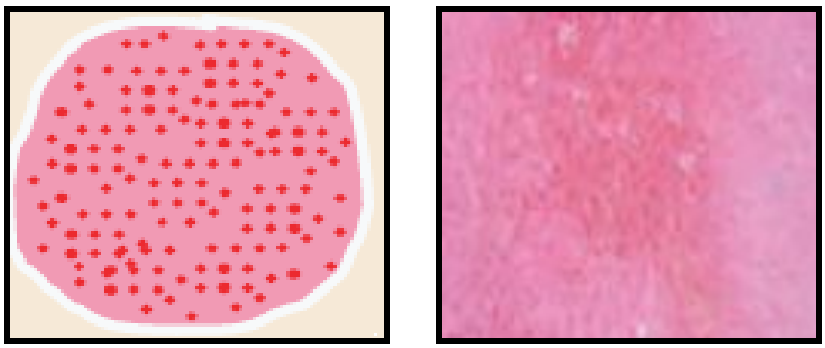

Figure (3): Psoriasis ***(Vazquez-Lopez et al., 2010).***

Dermoscopically, lichen planus reveals dotted vessels arranged in a peripheral arrangement in a dull red background colour. White crossing lines (Wickham striae) are exclusively seen in lichen planus**.** Dermatoscopic evaluation of lichen planus provides an easy and rapid recognition of Wickham striae, which is predictive for the disease, along with other deeper structures including gray-blue dots, cysts, or vascular structures generally not visible to the naked eye **(Figure 4)** ***(Vázquez-López et al., 2010).***

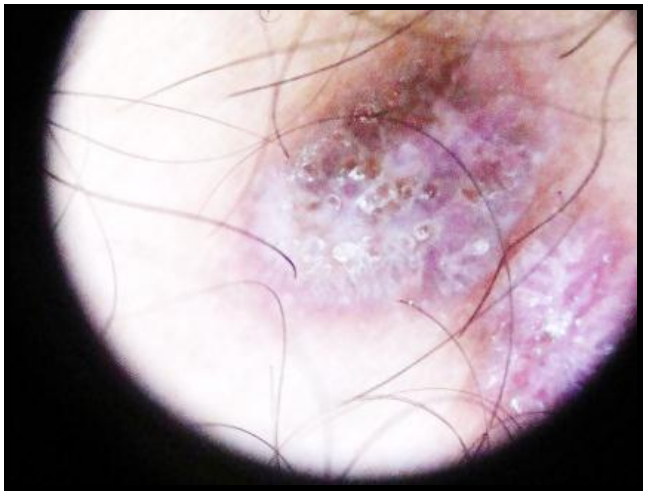

Figure (4): Lichen planus ***(Vázquez-López et al., 2010).***

Dermoscopic features of seborrheic dermatitis are characterized by arborizing vessels and atypical red vessels with the absence of red dots and globules. The arborizing vessels and atypical red vessels in seborrheic dermatitis represent ectatic subpapillary plexus in slightly hyperplastic rete ridges. In seborrhoeic dermatitis, the vessels proliferate horizontally through

the subpapillary plexus with perivascular inflammation. Featureless areas devoid of any particular vascular patterns are also frequently observed in seborrhoeic dermatitis ***(Kim et al., 2011).***

Dermoscopy is used in assisting the early diagnosis of a depigmentation condition (localized vitiligo). A pattern of depigmentation with residual reservoirs of perifollicular pigments is clearly visualized. This pattern is not seen in other disorders of depigmentation. Such pattern signifies focally active or re-pigmenting vitiligo and thus, clearly serves as a useful guide for the cases wherein there exists a doubt about the possible diagnosis ***(Chuh and Zawar, 2004).***

Dermoscopy allows better visualization of the typical clinical morphologic findings of porokeratosis, namely, the characteristic annular whitish-yellowish structure demarcating a central scar-like area and surrounded by a minimal peripheral vascularization ***(Delfino et al., 2004).***

Colors of striae distensae are often different from that of the surrounding skin. A close look using dermoscopy shows distinct patterns of melanized networks at these sites. There are four distinct types, namely striae albae, striae rubrae, striae caeruleae and striae nigrae. The latter hyperpigmented type of striae distensae is specifically identified by epiluminescence examination in dark-skinned subjects. The fine-melanized honeycomb network present on the adjacent intact skin is