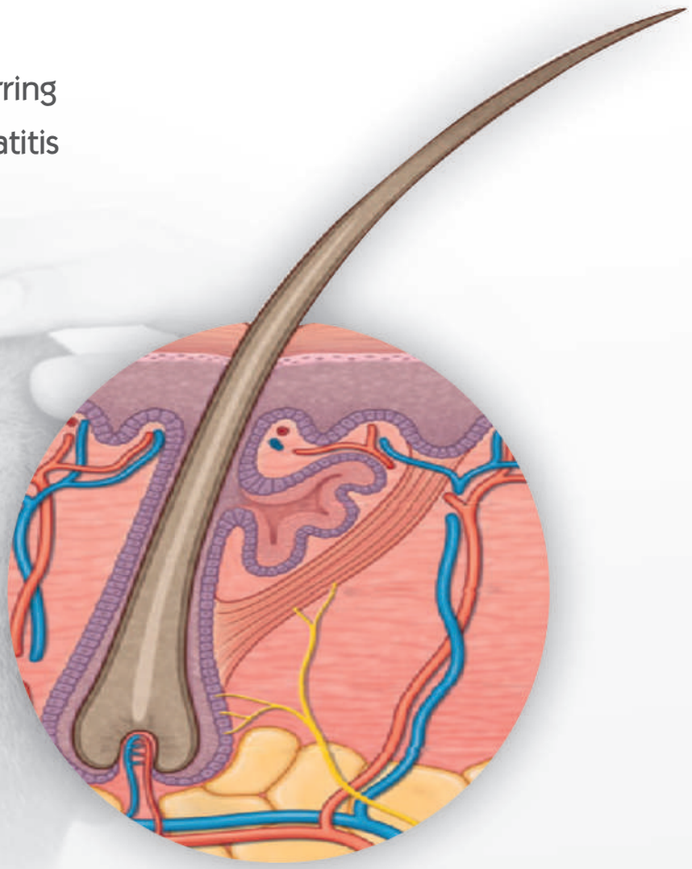
Chapter

1

Hair and Scalp

- Alopecia: Scarring and Non-Scarring
- Infections and Seborrheic Dermatitis
- Hirsutism
- Hair Shaft Disorders
- Tumors
- Miscellaneous Disorders



ALOPECIA: SCARRING AND NON-SCARRING

ALOPECIA AREATA

Alopecia areata (AA) is an autoimmune disorder that affects the hair follicles causing non-scarring hair loss. AA classically presents as asymptomatic well-demarcated round smooth patches of non-scarring hair loss. The periphery of the patches shows "exclamation mark" hair which are broad distally and tapered proximally and indicate active disease.

AA can be classified into the following clinical types based on the extent of hair loss:

a. Patchy (Figs 1.1a and b): There is partial loss of scalp hair.
b. Alopecia totalis (AT): Hundred percent of scalp hair are lost (Fig. 1.1c).
c. Alopecia universalis (AU): Body hair are also lost.

Less common presentations are:

a. Ophiasis: Band-like areas of hair loss in parieto-temporo-occipital regions of scalp (Figs 1.1d and e).
b. Ophiasis inversus/sisaipho: Hair loss in fronto-parieto-temporal areas.
c. Diffuse: Diffuse thinning over part or whole of scalp.

AA can be associated with nail changes like trachyonychia, Beau's lines, koilonychia, leukonychia. Other autoimmune diseases, e.g. thyroid disorders and vitiligo may be associated. However, routine screening for autoimmune disorders is not indicated except in patients with AT, AU, those with long duration of disease and older age at onset.

Management

Diagnosis is mainly clinical. Scalp biopsy is required only in doubtful cases and shows a classical 'swarm of bees' appearance due to a peribulbar lymphocytic infiltrate. Spontaneous regrowth can occur in up to 50% of patients in one year. Hence, patients should be counselled and offered the option of "no treatment" in localized disease.

Topical Agents

Topical treatments cause regrowth in the treated areas but do not affect the activity of disease. Options available are:

i. *Corticosteroids*: Intralesional steroids (triamcinolone) can be given at a concentration of 5 mg/ml for scalp and 2.5 mg/ml for eyebrows. It forms the first line of treatment for adult patients with less than 50% scalp involvement. Injections have to be repeated every 4 to 6 weeks. Topical steroids can be used but are generally less effective.
ii. *Minoxidil*: Used at a concentration of 5%, it forms an effective therapy for AA.
iii. *Anthralin*: Used as short contact therapy. Effective, if used properly under supervision.

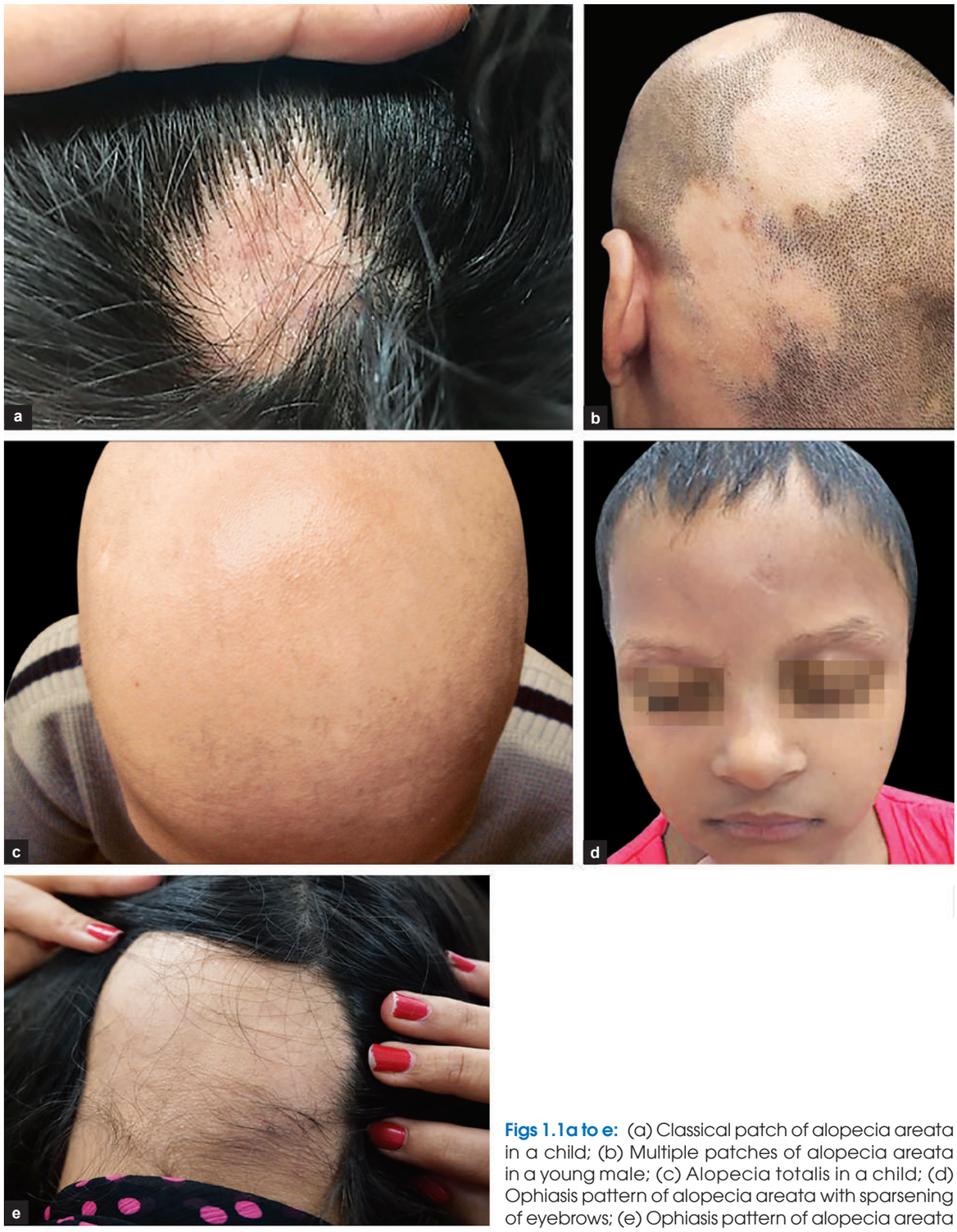
Systemic Agents and Phototherapy

Other treatment options are topical immunotherapy and photochemotherapy. Rapidly progressive and extensive disease requires systemic treatment. Various options available are corticosteroids, cyclosporine, sulfasalazine, methotrexate, azathioprine and biological agents.

In our experience, steroids are not needed in majority of cases and should be avoided as they lead to side effects even with the "so-called" oral pulse treatment.

Cyclosporine is useful but we have noted rapid relapses in severe cases.

Azathioprine, methotrexate and tofacitinib (JAK STAT inhibitor) are useful agents but should be given under expert supervision.



Figs 1.1a to e: (a) Classical patch of alopecia areata in a child; (b) Multiple patches of alopecia areata in a young male; (c) Alopecia totalis in a child; (d) Ophiasis pattern of alopecia areata with sparsening of eyebrows; (e) Ophiasis pattern of alopecia areata

TRACTION ALOPECIA

Prolonged traction on the hair due to tight hairstyles can lead to damage to follicles, causing hair loss. Use of rollers can also result in traction alopecia (TA). The hair loss is initially non-scarring but can progress to scarring owing to atrophy of follicles due to prolonged traction. TA should be differentiated from ophiasis variant of AA.

TA presents as geometric areas of hair loss on the sides of scalp, which are sites of maximum traction (Figs 1.2a and b). "Fringe sign" that is a fringe of hair at the margin of the alopecic patch can help in diagnosis.

Management

Patient needs to be counselled regarding modification of hairstyle practices. However, if atrophy develops, the condition becomes permanent.

TRICHOTILLOMANIA

Trichotillomania (TTM) has been reclassified as an obsessive-compulsive disorder according to the American Psychiatric Association Classification DSM-V.

It is characterized by habitual hair pulling with a rising tension prior to pulling of hair and a sense of relief after the act or behaviour.

Patients usually pull scalp hair though eyebrows, eyelashes, pubic region and trunk can also be targeted. TTM presents with bizarre shaped irregular patches of incomplete alopecia that have hair of varying lengths. These are typically seen on approachable areas of scalp with uninvolved areas appearing completely normal (Figs 1.2c to f). Hair pull test is negative. The patients manipulate the hair (e.g. twisting) prior to pulling. There may be accompanying trichophagy which is the swallowing of pulled hair. This can lead to intestinal obstruction. TTM, when present in adults, is usually associated with stress or personality disorder.

The condition should be differentiated from other causes of non-scarring hair loss, e.g. alopecia areata and tinea capitis. Presence of hair of varying lengths, absence of exclamation mark hair and scaling are diagnostic pointers. Doubtful cases may be helped with a scalp biopsy that shows deformed hair and empty hair follicles.

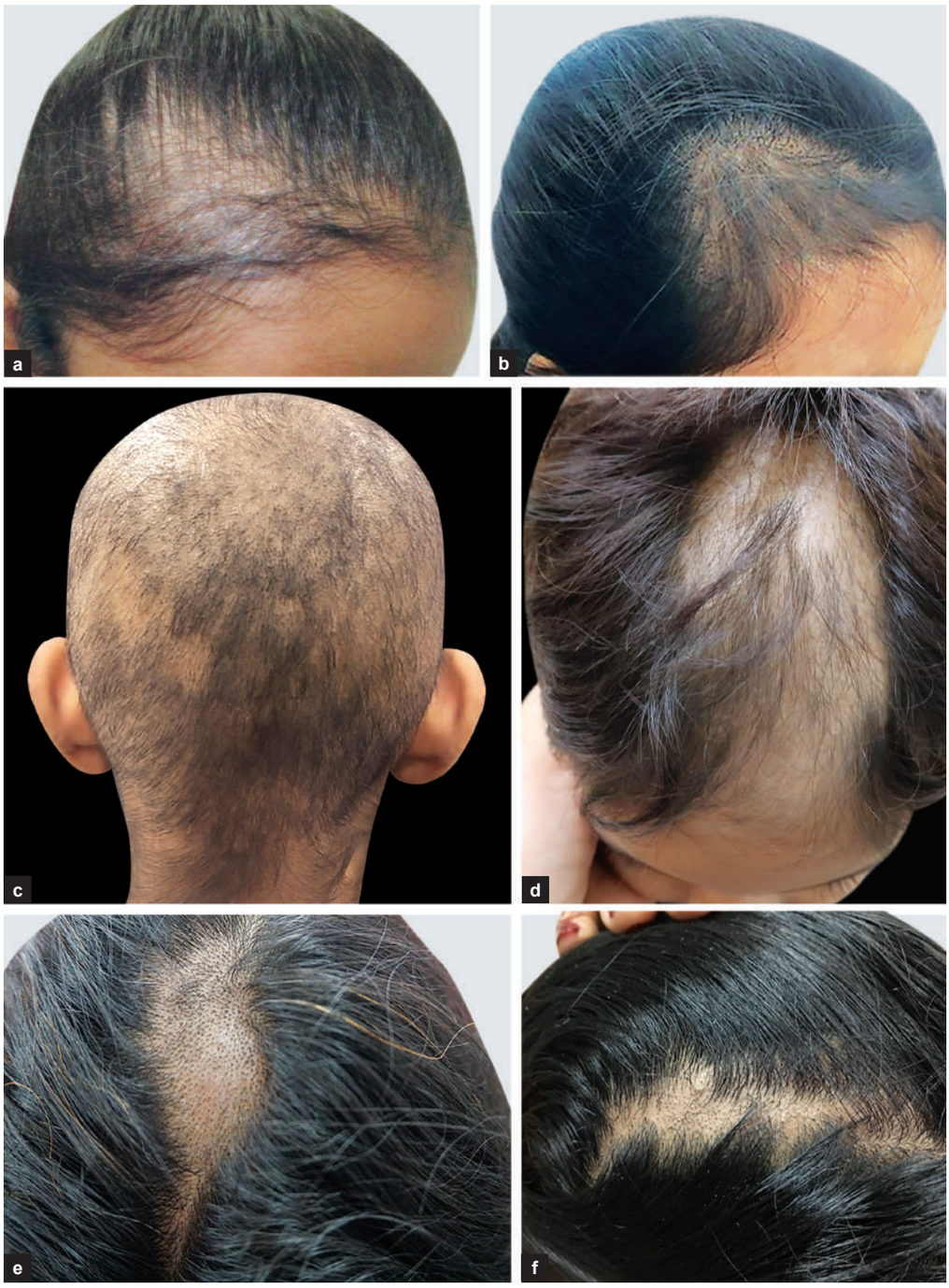
Management

Lip licking, hair pulling (trichotillomania), and nail-biting are common childhood habits, and have a good prognosis in this age group. The same in adults is associated with emotional stress, and tensions involving family or work may precede the onset.

In some cases, these disorders may also be associated with learning difficulties and psychiatric conditions such as personality, eating, or mood disorders. Although patients may know what they are doing, they are unable to stop the damaging behaviour.

Behaviour modification therapy is the mainstay of treatment. Pharmacologic therapy has been tried with variable success rates. Selective serotonin reuptake inhibitors (SSRI) which are a class of anti-depressants are most commonly used. Fluoxetine, clomipramine, sertraline, and paroxetine are the options available.

Antipsychotics (olanzapine) have been used in a few case series. Recently, N-acetylcysteine has been tried with good results.



Figs 1.2a to f: (a) Traction alopecia seen usually on the frontal hair margin; (b) Traction alopecia in a girl who tied her hair tightly; (c) Severe trichotillomania in a 15-year-old girl with family conflict; (d) Trichotillomania in a child; (e) A female patient with trichotillomania. There are broken hair in the centre; (f) A 10-year-old girl with trichotillomania. Note broken hair and the irregular margins

TELOGEN EFFLUVIUM

Telogen effluvium (TE) is one of the most common causes of diffuse hair loss. It occurs due to the disruption of hair cycle causing increased synchronised shedding of telogen hair. It can be broadly classified into acute and chronic TE.

Acute TE: It occurs 2–3 months after a triggering event, e.g. high fever, surgery, crash dieting, and postpartum. It resolves within a few months in majority of cases.

Chronic TE: Chronic TE (CTE) is an idiopathic condition affecting middle-aged women. Chronic diffuse telogen hair loss (CDTHL) refers to telogen effluvium secondary to organic causes, e.g. thyroid disorder, iron deficiency anemia, etc.

Both CTE and CDTHL present with hair shedding that lasts longer than 6 months. There is no widening of central partition or miniaturization of hair. They have a prolonged fluctuating course. On examination, hair are normal in thickness (Fig. 1.3a). A few patients may have bitemporal recession (Figs 1.3b, c and e). Hair pull test is positive. Patients usually come with a collection of hair which can be used to assess the severity of the disease (Fig. 1.3d).

Management

Thorough work-up to rule out organic causes should be done. These include full blood count, thyroid function tests, iron studies and other tests based on the patient's history. The role of various vitamins and supplements (e.g. biotin) in patients without any evidence of deficiency is doubtful. In cases of iron deficiency, iron replacement should be started.

Only in case of chronic telogen effluvium does minoxidil 5% help.

A few steps are listed below that help to delineate the common causes.

- Assessment of dietary habits and determination of iron saturation and ferritin are the simplest ways to determine nutritional status.
- Its important to note that iron supplements may not always reverse hair loss, thus addressing nutritional imbalance as a whole is useful.
- Sources of blood loss, such as menstrual bleeding and gastrointestinal (GI) blood loss, should be investigated.
- Simple cause like hypothyroidism, allergic contact dermatitis to hair dyes, and renal dialysis with secondary hypervitaminosis A should be ruled out.
- A long list of drugs are associated (amphetamines, captopril, carbamazepine, cimetidine, danazol, enalapril, metoprolol, propranolol) with CTE.
- Also effluvium in infants may occur between birth and the first 4 months of age. Usually, regrowth occurs by 6 months of age. Telogen counts by Kligman in six infants varied from 64–87%. He also found a tendency for the alopecia to occur in the male-pattern distribution.

Figs 1.3a to e: (a) Chronic telogen effluvium; (b, c) Bitemporal recession in telogen effluvium; (d) Daily hair count: Collected hairs of 7 days; (e) Temporal recession in a girl with chronic telogen effluvium due to iron deficiency anemia

MALE ANDROGENETIC ALOPECIA (MALE-PATTERN HAIR LOSS)

Male androgenetic alopecia (MAGA) is the most common cause of alopecia in men worldwide. The condition is genetically determined with a polygenic inheritance. There is hormone-induced miniaturization of terminal hair into vellus hair due to action of dihydrotestosterone (DHT), that is a metabolite of testosterone formed by action of 5α-reductase.

The Norwood-Hamilton classification describes the grades of MAGA depending on the extent of hair loss. It ranges from I to VIII with type I representing normal prepubertal scalp with terminal hair (Fig. 1.4a). There is gradual temporal recession leading to an M-shaped hairline (Figs 1.4b and c). Hair in the occipital area are unaffected.

Management

The diagnosis is mainly clinical but can be aided by dermoscopy in early cases.

Topical minoxidil forms the first line of treatment. It is available in 2 or 5% solution and applied 1 ml twice daily. Hair growth starts at around 3–4 months and peaks at 2 years of use. Other topical products available are aminexil, various vitamins and minerals, prostaglandin analogs. However, scientific data on their efficacy is lacking.

Systemic Agents

Oral finasteride is FDA approved for the treatment of MAGA and is used at a dose of 1 mg per day. It is a type II 5α-reductase inhibitor that decreases production of DHT. It is generally combined with topical minoxidil and gives good results.

FEMALE-PATTERN HAIR LOSS (FPHL)

Androgenetic alopecia in females is referred to as female-pattern hair loss owing to the uncertain relationship between androgens and this condition. There is alteration of hair cycle dynamics leading to miniaturization of hair follicles that results in transformation of terminal hair into vellus hair. The duration of anagen shortens from 3 years to a few months or weeks.

Affected females present with gradual thinning of hair with or without history of increased shedding. There is decrease in hair density over the mid-frontal scalp leading to widening of central partition with intact frontal hairline. Three stages have been defined by Ludwig based on the extent of involvement with stage I showing minimal widening and stage III with severe involvement (Fig. 1.4d).

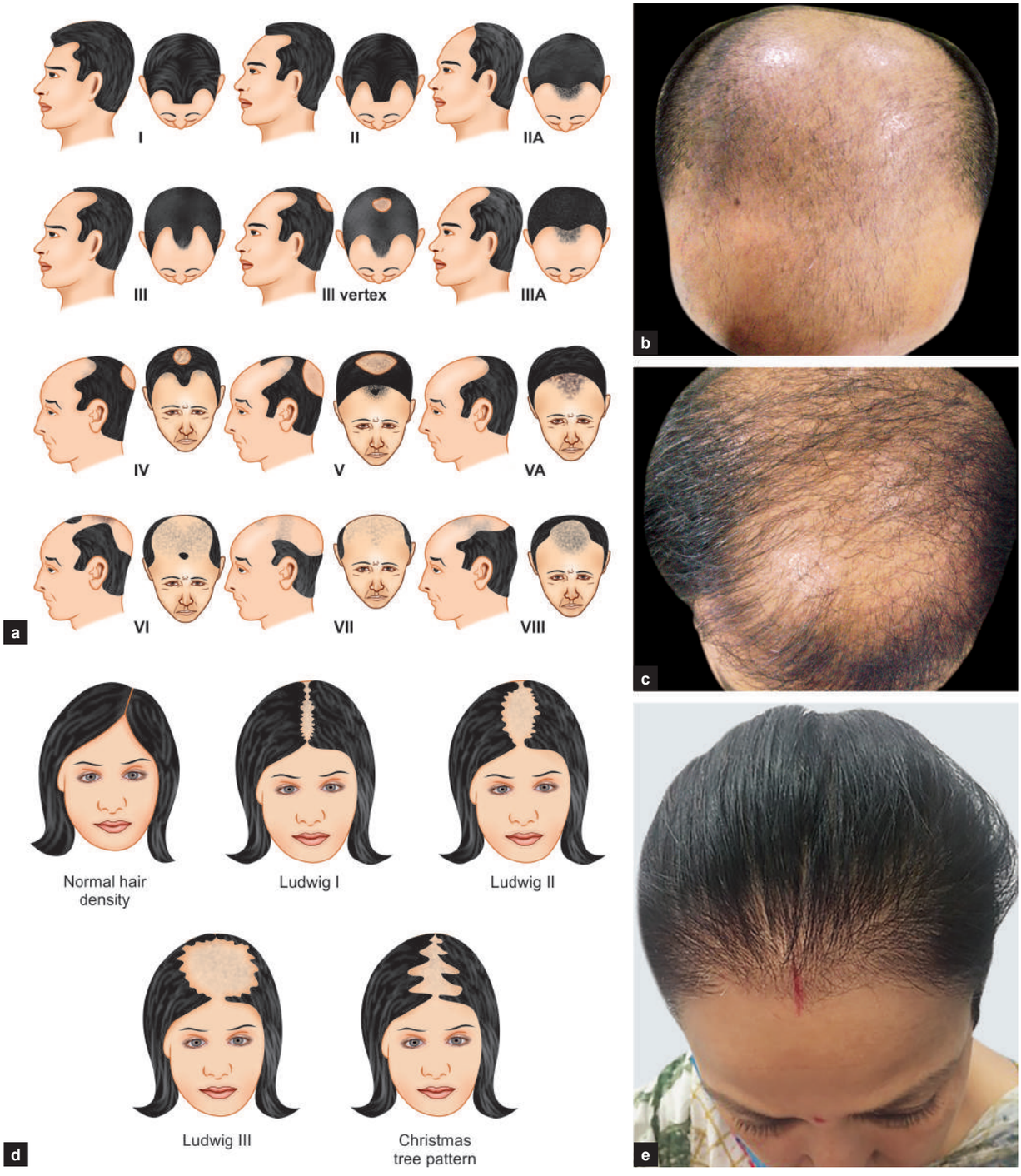
Representative cases are depicted in Figs 1.4d and e.

Management

History and investigations are aimed at ruling out other causes of diffuse hair loss like nutritional, metabolic and endocrine causes. Women with signs of hyperandrogenism, like hirsutism, moderate to severe acne, irregular menstrual cycles, should be screened for androgen excess. Treatment involves use of various topical and oral agents—minoxidil 2% solution and 5% foam have been approved by the FDA for use in FPHL.

Various antiandrogens and 5α-reductase inhibitors used are finasteride, cyproterone acetate, drosperinone and flutamide used with oral contraceptive pills (OCPs).

Patients with extensive hair loss may be offered hair transplantation. However, the prerequisite is a high donor density. Various other drugs (biotin, aloe vera, Chinese herbal extract, aminexil, zinc), though prescribed widely lack scientific evidence of efficacy. This is also true for the various "magical" hair oils advertized in the media.



Figs 1.4a to e: (a) The Norwood-Hamilton classification of male-pattern alopecia; (b) A male patient with Norwood-Hamilton stage V alopecia; (c) A patient with Norwood-Hamilton stage V with preserved frontal hair margin; (d) A depiction of various stages of pattern alopecia in females; (e) Female-pattern hair loss in a 45-year-old female

SCARRING ALOPECIA

This is a serious problem which should be treated and diagnosed early as it can lead to an irreversible hair loss, if untreated.

Common Causes

- *Trauma*: Including radiotherapy.
- *Tumours*: Benign: naevus sebaceous—present at birth and malignant: Basal cell carcinoma (BCC), squamous cell carcinoma (SCC), metastatic deposits.
- *Discoid lupus erythematosus* (*DLE*): DLE usually presents as erythematous (Fig. 1.5a), atrophic, and alopecic patches. There is marked follicular hyperkeratosis (Fig. 1.5b), hyperpigmentation, hypopigmentation (Fig. 1.5c), and telangiectasia. The lesions can heal with depigmentation.

LICHEN PLANOPILARIS (LPP)

It is a type of primary cicatricial (scarring) alopecia characterized by loss of follicular ostia and replacement of hair follicles with fibrous tissue on histopathology.

In early stage, crown and vertex show follicular violaceous papules and perifollicular erythema. These progress to form areas of alopecia which are small and irregularly shaped (Figs 1.5d and e). There may be associated itching or burning sensation. Associated lesions of lichen planus can be seen in a few cases.

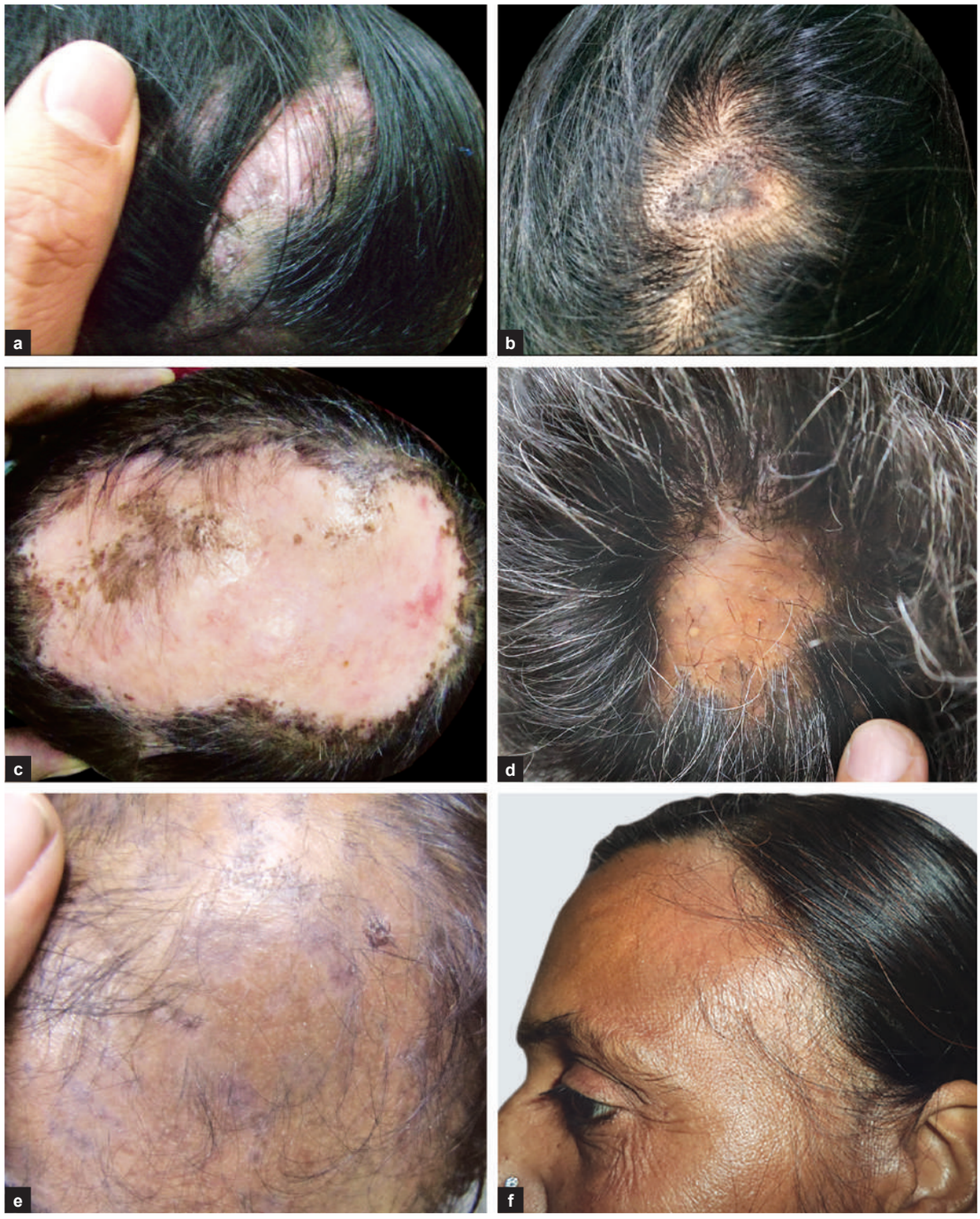
Management

Scalp biopsy should be done to assess activity of disease. In cases with active disease, main aim is to prevent progression and scarring with use of topical and systemic steroids. Intralesional steroids injected into the hair-bearing areas can help in arrest of disease. Other treatment modalities that have been used are antimalarials (hydroxychloroquine), cyclosporine and thalidomide. Pioglitazone (15 mg for 3 months) has been tried in a few cases. In cases with inactive disease, patients may be offered hair restoration surgery.

FRONTAL FIBROSING ALOPECIA

This is seen in older women with a band-like frontotemporal alopecia, hypopigmentation and atrophy (Fig. 1.5f), often with "kinky hairs". It is regarded as a variant of lichen planopilaris due to the similarity between the two conditions clinically and histopathologically.

The treatment involves a combination of pioglitazone and dutasteride. Other treatment options include oral and intralesional steroids, minoxidil and oral isotretinoin.



Figs 1.5a to f: (a) A patient with DLE on the scalp; early stage with erythematous plaques; (b) A patch of discoid lupus erythematosus with hyperpigmentation and hyperkeratosis; (c) A late stage of DLE with depigmentation of the scalp with scarring; (d) Scarring alopecia with perifollicular casts and a purplish hue of the scalp; (e) Multiple violaceous plaques with loss of hair; (f) Frontal fibrosing alopecia

INFECTIONS AND SEBORRHEIC DERMATITIS

Seborrheic dermatitis (SD) is a chronic and relapsing inflammatory condition that affects the seborrheic areas of the body such as the scalp, face and upper chest. Various factors are involved in its pathogenesis. These are Malassezia colonization, increased sebaceous secretions, host immune response, stress and genetic factors. However, the key pathogenetic factor is the alteration of Malassezia from a commensal to pathogen.

SD manifests as "cradle cap" in infants up to 3 months of age and is benign and self-limiting. There is a peak in adolescents and adults with higher prevalence in men. Dandruff is a mild variant of SD that presents as scaling without inflammation (Fig. 1.6a).

A more severe form can present as greasy, erythematous patches on scalp that may be associated with itching (Fig. 1.6b). This can extend to the face (Fig. 1.6c) with marked involvement of sebum-rich areas of face, like nasolabial folds, eyebrows, preauricular areas and anterior chest with scaly erythema, thin papules and plaques associated with itching.

A very common misdiagnosis is psoriasis where the scales are prominent, the plaque is well defined and the border extends beyond the frontal hair margin (Fig. 1.6d).

Management

Ketoconazole shampoo is the first line of treatment for seborrheic dermatitis. Cream preparation can be used for cutaneous lesions. Miconazole can also be used. Other antifungal agents used as shampoos are ciclopirox, zinc pyrithione, and selenium sulfide. Severe cases of SD need combination with topical steroids to decrease signs of inflammation.

Oral antifungal agents (terbinafine, fluconazole, itraconazole) are rarely used due to lack of scientific evidence of efficacy. Isotretinoin can be used in refractory cases in doses of 0.1–0.5 mg/kg/day. It reduces sebum secretion and is anti-inflammatory.

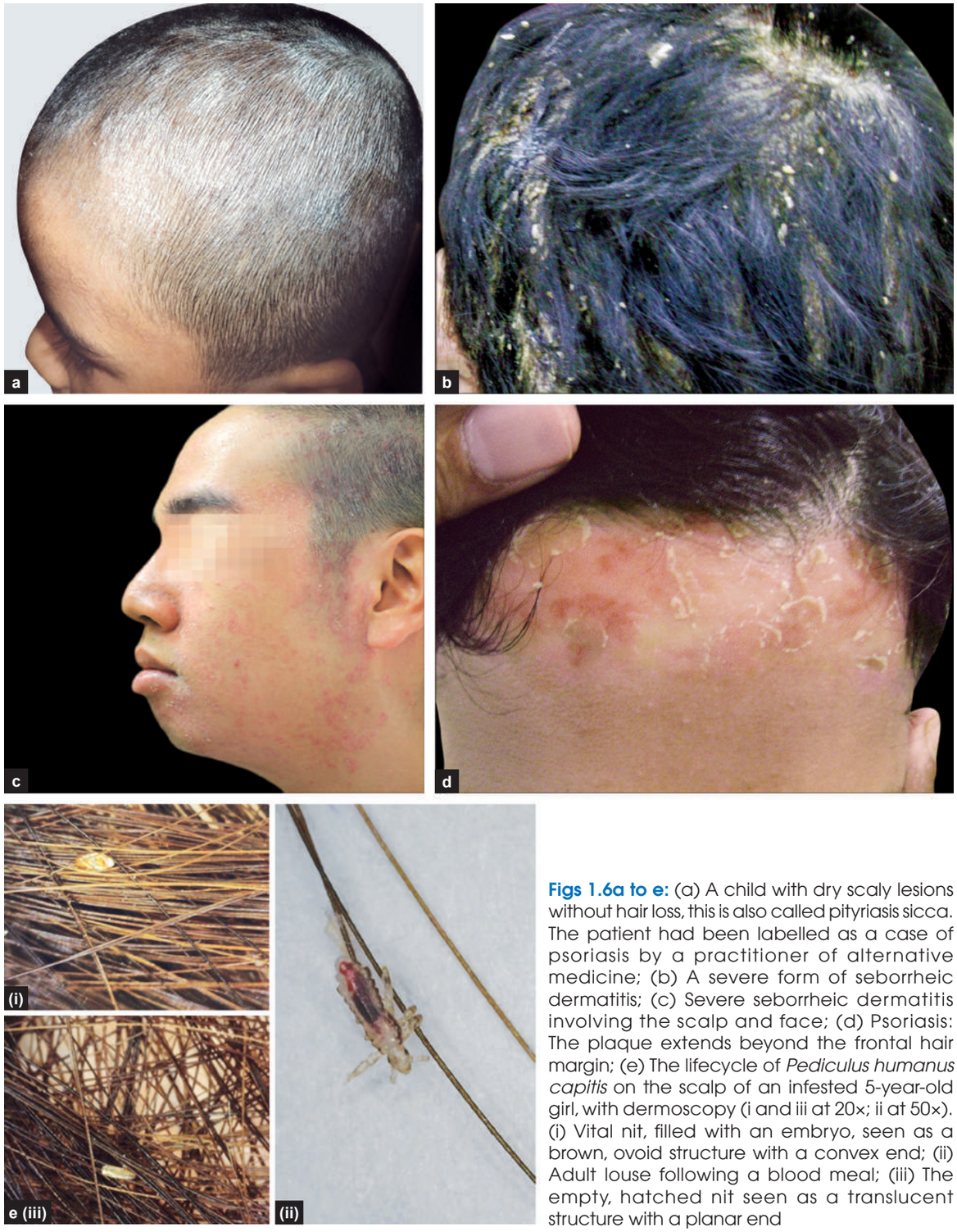
PEDICULOSIS CAPITIS/HEAD LICE

It is caused by *Pediculus capitis* (head louse), which are bloodsucking, wingless insects that live on the scalp hair (Fig. 1.6e). Patients present with complaints of intense pruritus of the scalp. There may be excoriations, erythema and secondary infection. Posterior cervical lymphadenopathy may be present.

Diagnosis can be made by identification of nits which are eggs of size 0.8 mm firmly attached to hair. Adult lice can be seen sometimes.

Management

- Permethrin 1% cream rinse (patients aged ≥2 months) is used for treatment of head lice. It is applied after shampooing and drying the hair. Medication should be kept for at least 10 minutes and should be washed with plain water. Shampooing should be avoided in the next 24 hours. Application should be repeated after 7–10 days.
- Other treatment options are ivermectin 0.5% lotion (≥6 months), oral ivermectin (200 µg/kg single dose repeated after 7 days, 400 µg/kg for >15 kg), malathion gel (children aged ≥2 years).
- Administration of 3 doses of ivermectin (1 dose/wk at weekly intervals) may be needed to eradicate heavy infection.
- Non-pesticidal products that have been claimed to be effective are petroleum jelly, hair pomade, vegetable oil and essential oils. These products do not kill the lice but only slow their movement which helps in combing them out easily. Family members should be examined and given treatment, if there is evidence of infection. Hair care items like combs, hats, brushes should not be shared. Metal combs are better than plastic combs in combing the lice/nits out.



Figs 1.6a to e: (a) A child with dry scaly lesions without hair loss, this is also called pityriasis sicca. The patient had been labelled as a case of psoriasis by a practitioner of alternative medicine; (b) A severe form of seborrheic dermatitis; (c) Severe seborrheic dermatitis involving the scalp and face; (d) Psoriasis: The plaque extends beyond the frontal hair margin; (e) The lifecycle of *Pediculus humanus capitis* on the scalp of an infested 5-year-old girl, with dermoscopy (i and iii at 20×; ii at 50×). (i) Vital nit, filled with an embryo, seen as a brown, ovoid structure with a convex end; (ii) Adult louse following a blood meal; (iii) The empty, hatched nit seen as a translucent structure with a planar end

TINEA CAPITIS

T. capitis is infection of the scalp and hair with dermatophytes (*Trichophyton* spp and *Microsporum* spp). It is common in school-going children and spreads through contact and fomites. It can present as:

i. *Gray patch T. capitis*: It presents as circular areas of partial alopecia with gray scales on the surface (Fig. 1.7a(i) and a(ii)). The hair are brittle and easily pluckable.

ii. *Black dot T. capitis* (Fig. 1.7b): Characterized by areas of hair loss with black dots on the skin surface which occur due to breakage of hair close to the scalp surface.

iii. *Kerion* (Fig. 1.7c): It is an inflammatory variant of T. capitis characterized by erythematous, boggy, areas of alopecia with pustule formation. There may be associated occipital lymphadenopathy.

Management

T. capitis requires systemic treatment in the form of oral antifungal agents. Griseofulvin is the drug of choice and is used in a dose of 10–15 mg/kg/day in divided doses for a duration of 6–8 weeks. Other options are terbinafine, itraconazole and fluconazole. Griseofulvin is superior for *Microsporum* infections, but terbinafine is superior for *Trichophyton* infections.

- Griseofulvin ultramicrosize 10–15 mg/kg/day or microsize 20–25 mg/kg/day qd PO for 6–12 wk (taken with milk or fatty foods to augment absorption). For kerion, treat concurrently with prednisone (1–2 mg/kg/day for 1–2 wk).
- Terbinafine can be used for only 2–4 wk. Terbinafine dosing is 62.5 mg/day (<20 kg), 125 mg/day (20–40 kg), or 250 mg/day (>40 kg).
- Alternatives: Itraconazole oral solution 5 mg/kg od or fluconazole.
- 2.5% selenium sulfide shampoo, or 2% ketoconazole shampoo, 2–3 times/week should be used concurrently to prevent recurrences. Other family members including siblings should be screened for infection and treated.

SCALP FOLLICULITIS/PYODERMA OF SCALP

It is characterized by small, very itchy pustules on the scalp, often most troublesome on the frontal hairline (Fig. 1.7d). The treatment is similar to pyoderma. It is advised that the treatment of severe infections should be aggressive, as in some cases long-standing hair loss may ensue (Fig. 1.7e).

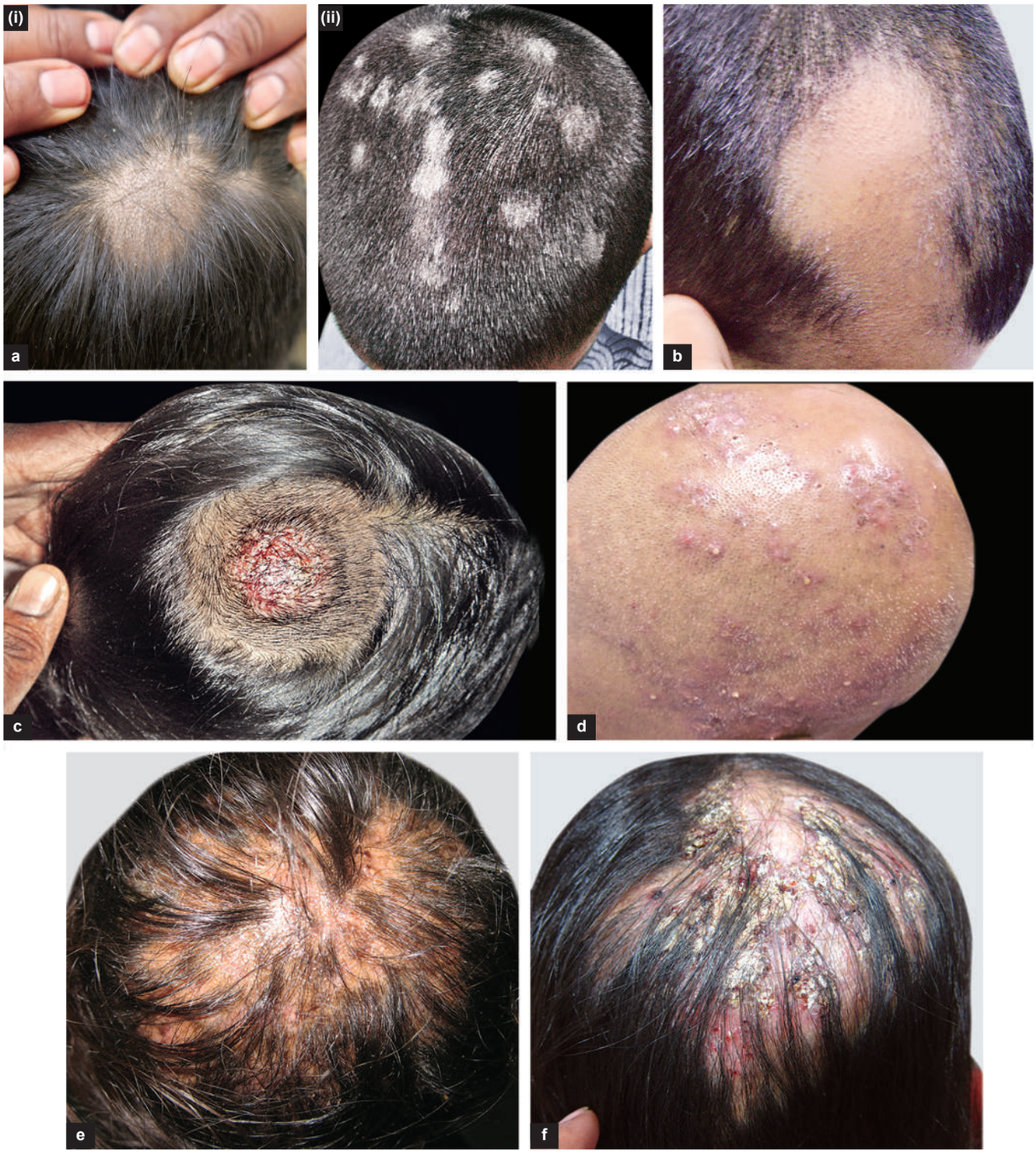
FOLLICULITIS DECALVANS

This is a rare chronic condition of the scalp characterized by painful, recurrent purulent follicular exudation, which can lead to a confluence of lesions with scarring alopecia (Fig. 1.7f).

It is now believed that this represents a chronic staphylococcal infection and the host immune response causes the hair loss.

Management

Long-term tetracycline treatment generally results in a sustained effect and this is usually preceded by a course of rifampin and clindamycin. Oral retinoids, oral and topical fusidic acid, oral zinc sulfate, photodynamic therapy (PDT) and topical tacrolimus have been reported as successful, and anti-TNF biologics have been used for refractory disease.



Figs 1.7a to f: (a) (i) Gray patch tinea capitis; (ii) A case of gray patch tinea capitis. There is patchy circular alopecia and fine scaling. Fine scales represent the fungal arthrospores; (b) "Black dot" tinea capitis, which was being treated as a case of alopecia areata; (c) Inflamed, boggy, tender areas of alopecia in a case of kerion; (d) Scalp folliculitis; (e) Scalp folliculitis with discrete areas of hair loss; (f) Folliculitis decalvans

HIRSUTISM

Hirsutism is defined as the presence of terminal hair in a male pattern, i.e. over androgen dependent sites in women (Fig. 1.8a). It should be differentiated from hypertrichosis that is increased hair growth in androgen independent sites (Fig. 1.8b). Hirsutism occurs due to an increase in circulating androgens secreted by the ovary or adrenal gland or due to an enhanced end organ sensitivity to androgens. It may be accompanied by seborrhea, acne and androgenetic alopecia. Such cases are referred to as "SAHA syndrome" (seborrhea, acne, hirsutism and alopecia). Another syndrome associated with hirsutism is the HAIR-AN syndrome that is characterized by hyperandrogenemia (HA), insulin resistance (IR) and acanthosis nigricans (AN).

Management

The first step is to determine the source of increased androgens by doing a testosterone (free and total), DHEAS (marker for adrenal source) and 4-androstenedione (marker for ovarian source). SHBG and prolactin levels should also evaluated.

Treatment options are various antiandrogens and androgen receptor antagonists. These are cyproterone acetate, spironolactone, flutamide and finasteride. These are given along with oral contraceptive pills (OCPs).

Topical eflornithine (available as 13.9% cream in India) is the only topical agent available for treatment of hirsutism. It is applied on the affected areas twice daily.

Alexandrite, Nd:YAG and diode lasers can be used for hair reduction.

PREMATURE CANITIES/PREMATURE GRAYING OF HAIR

It is defined as the graying that occurs before the age of 20 years in Whites, before 25 years in Asians and before 30 years in Africans.

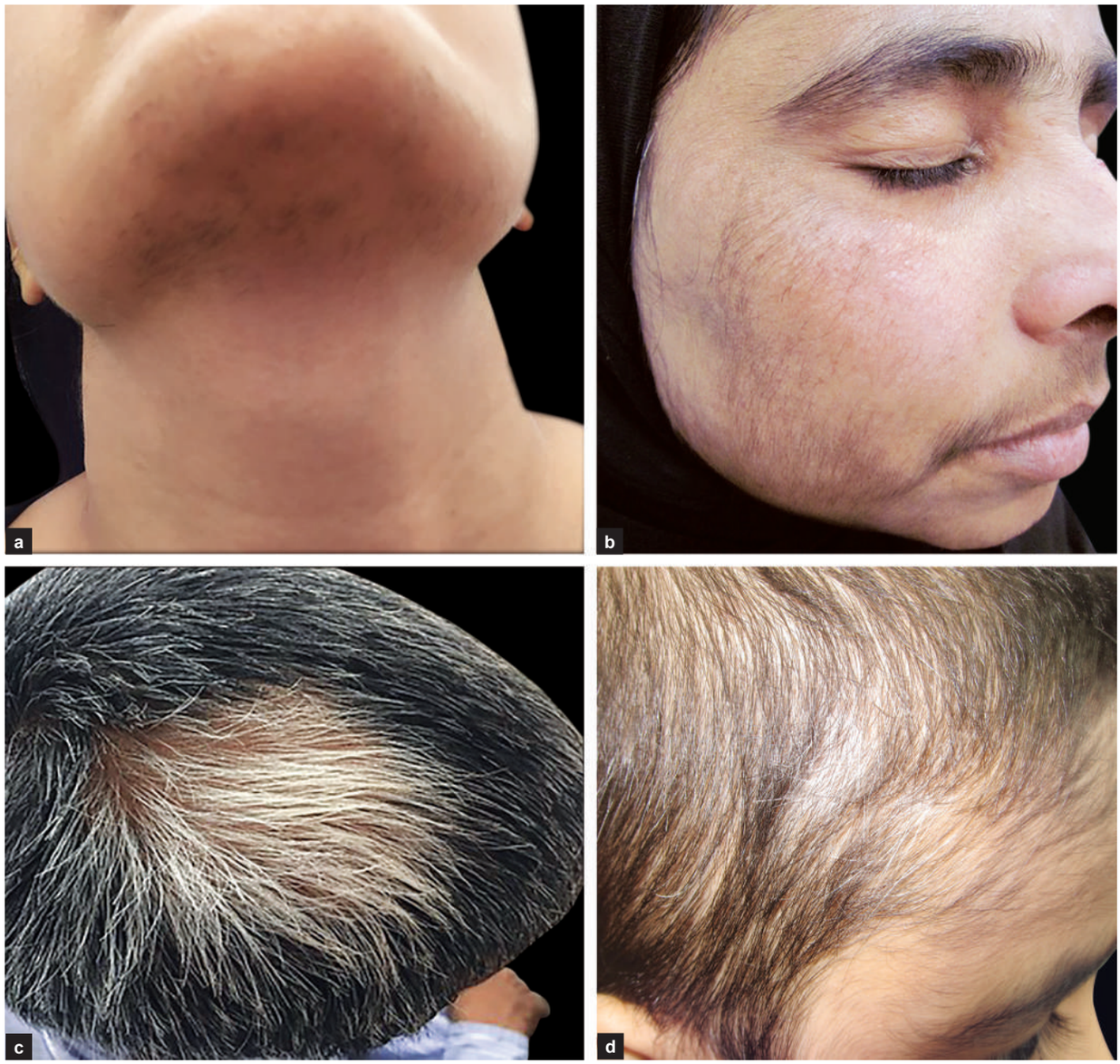
The cause is presumed to be reduced melanogenic activity as a result of fewer melanocytes and melanosomes, as well as a gradual loss of tyrosinase activity.

Its etiology is not completely understood. There is a genetic predisposition along with interplay of various environmental factors like stress, smoking, drugs (chloroquine, IFN-alpha, dithranol). Premature canities can occur in association with various autoimmune disorders like hypo/hyperthyroidism (Fig. 1.8c) and pernicious anemia and premature ageing syndromes. Sometimes it is seen overlying a lesion of vitiligo (Fig. 1.8d).

In males, the graying starts at the temples and sideburns and spreads to vertex. Occiput is affected the last. In women, graying starts around the hairline.

Management

Diagnosis is mainly clinical. Cases with very early onset should be investigated to rule out autoimmune disorders. There is no effective treatment. Patients are prescribed various nutritional supplements like biotin, zinc, copper and calcium pantothenate. However, there is no scientific evidence proving their efficacy.



Figs 1.8a to d: (a) Hirsutism in a female with PCOS; (b) Hypertrichosis secondary to prolonged steroid application over face; (c) Premature canities in a child with hyperthyroidism; (d) White hair over a patch of vitiligo

PITYRIASIS AMIANTACEA

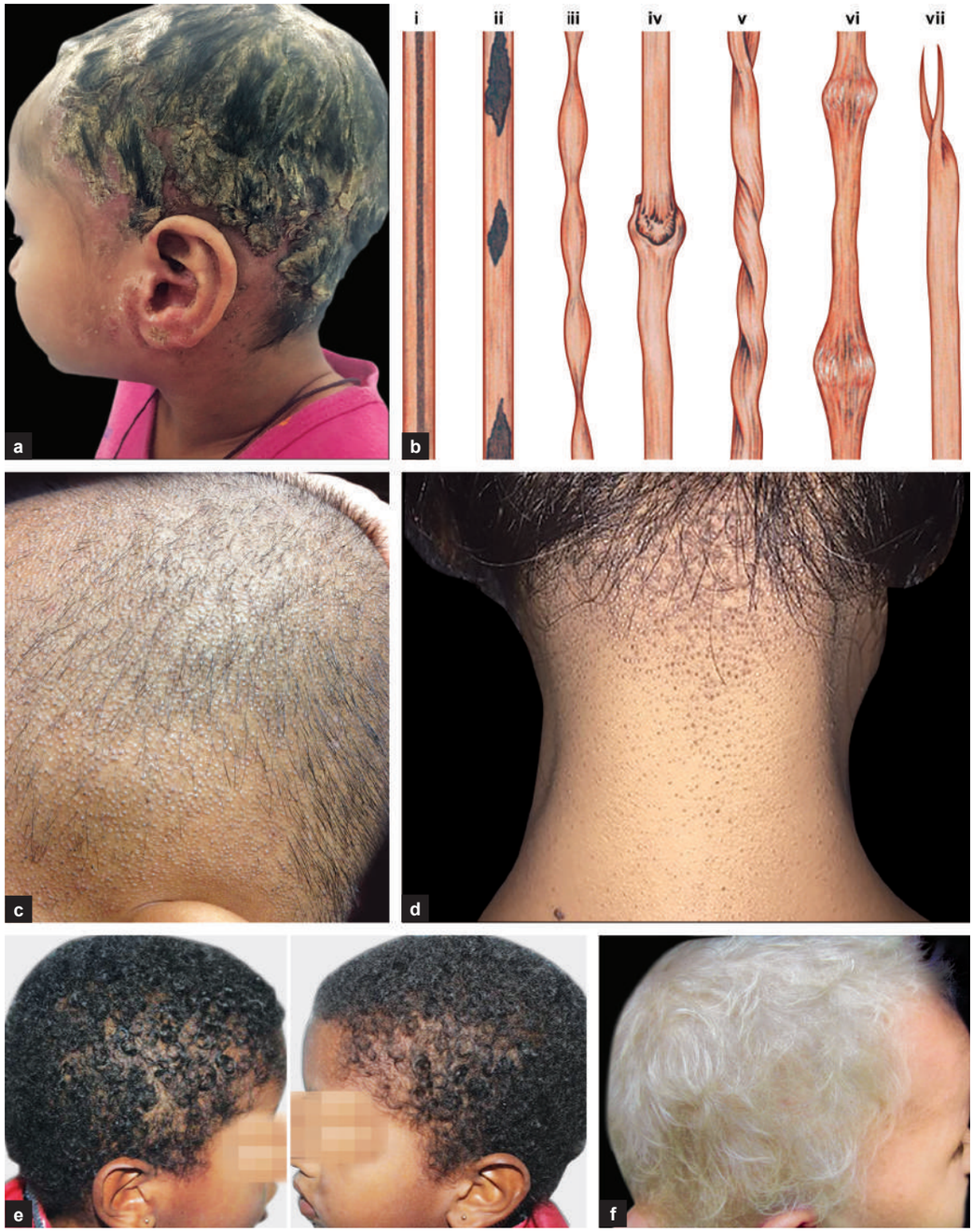
There is thick asbestos-like scaling over scalp. It is seen more commonly in children with pediculosis capitis, seborrheic dermatitis (Fig. 1.9a), psoriasis and tinea capitis. Shampoo with a selenium sulfide suspension or a tar- or steroid-containing shampoo for 2 weeks suffices in most cases.

HAIR SHAFT DISORDERS

Conditions with hair shaft defects are usually not amenable to treatment but can provide clues to other abnormalities. The diagnosis is based on light microscopy (LM) and trichoscopy. The disorders can be conveniently classified into those with increased fragility and those without increased fragility. Common disorders are depicted in Fig. 1.9b.

- **Monilethrix:** It is an autosomal dominant condition caused by defect in hair keratins. Affected patients give history of dry, lustreless, brittle hair that fail to grow (Fig. 1.9c). On examination, keratotic follicular papules can be seen (Fig. 1.9d). Severe cases have involvement of eyebrows and eyelashes. On examination of hair under a microscope, elliptical nodes can be seen at regular intervals giving rise to a beaded appearance. Oral retinoids and topical minoxidil may improve the alopecia.
- Other disorders include **woolly hair nevus** (Fig. 1.9e) and **loose** anagen hair syndrome/LAHS (Fig. 1.9f).

The majority of disorders have no definite treatment though in some an associated underlying systemic abnormality may be detected.

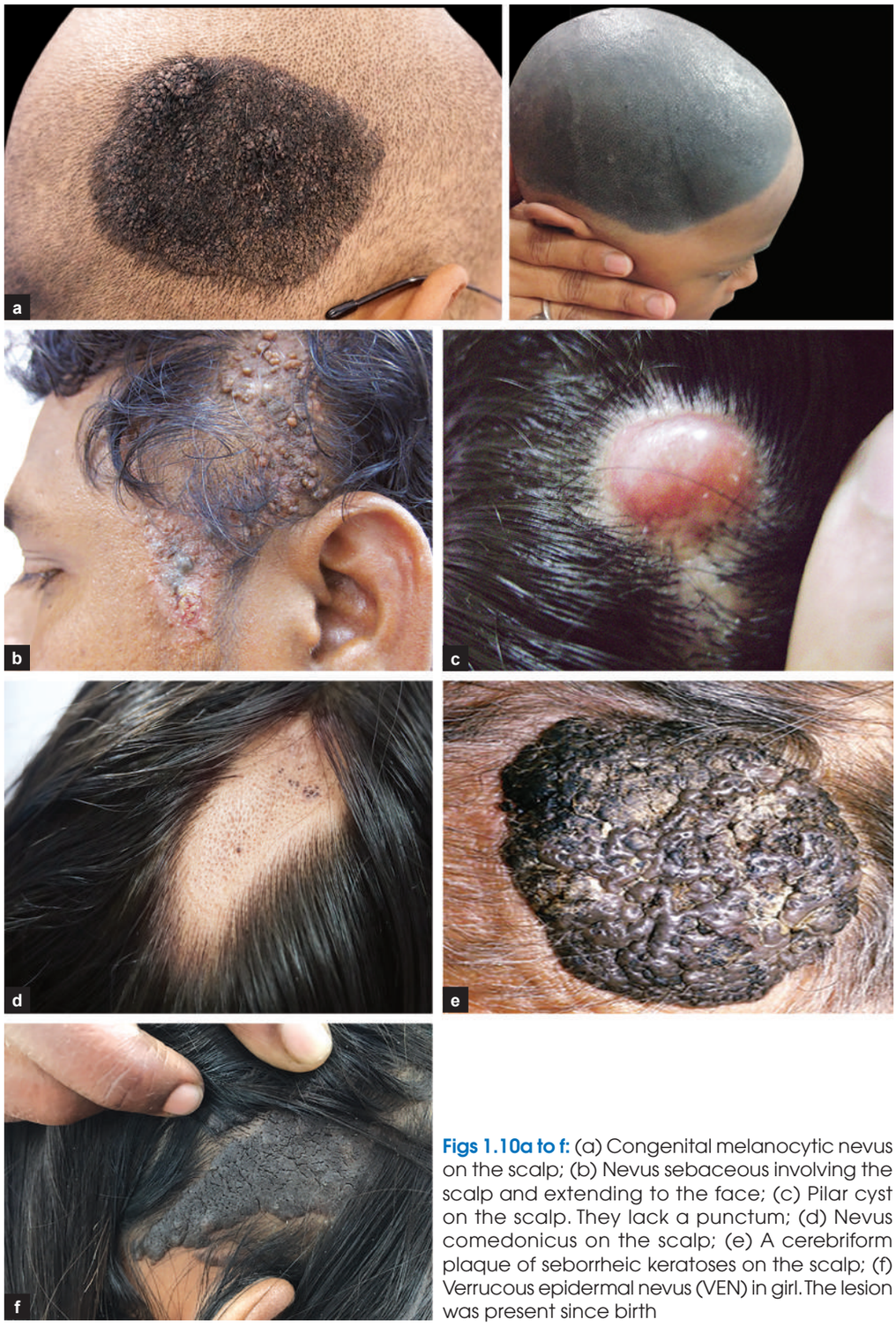


Figs 1.9a to f: (a) Pityriasis amiantacea in a child with seborrheic dermatitis; (b) A figurative depiction of the common hair shaft defects. (i) Normal hair, (ii) Pili annulati, (iii) Monilethrix, (iv) Trichorrhexis invaginata, (v) Pili torti, (vi) Trichorrhexis nodosa, (vii) Trichoptilosis; (c) Keratotic papules on scalp in monilethrix; (d) Keratotic papules over the neck and occiput; (e) Woolly hair: Tightly curled, short "Negroid" hair over the scalp. The hair length is decreased due to hair shaft brittleness; (f) An Indian patient with blonde hair, history of easy pluckability and a general "wind blown appearance" (LAHS)

TUMORS

The common tumors affecting the scalp can be divided into congenital and acquired. The congenital disorders may either be present at birth or may arise later on but have a genetic basis. Others include cysts and tumors arising out of the appendages. Malignant tumors are somewhat uncommon.

- **Congenital melanocytic nevi:** They can sometimes occur over scalp (Fig. 1.10a). They are usually of little concern in our population but can be prone to malignant change occasionally.
- **Naevus sebaceous:** This is present from birth or early childhood. It differs from a congenital melanocytic naevus in being yellowish with a flat, warty surface and hair loss (Fig. 1.10b). A basal cell carcinoma or other adnexal tumor may develop within it in middle age. If this happens, it is obvious, because there will be a lump within the naevus or a discharge from it. No treatment is necessary. It can be excised to reduce the area of hair loss. If a tumor develops or is suspected, an excision would be needed
- **Pilar (trichilemmal) cyst:** Pilar (or trichilemmal) cysts are derived from the external root sheath of hair follicles and occur predominantly on the scalp. They are inherited as an autosomal dominant trait, they appear between the ages of 15 and 30, and present to the doctor because the patient notices a lump when brushing or combing the hair. One or several subcutaneous nodules are present (Fig. 1.10c). They do not have a punctum and do not usually become inflamed.
- **Nevus comedonicus:** This is uncommonly seen on the scalp and is characterized by enlarged patulous follicles or comedones. Typical presentation is a pinkish, dome-shaped, smooth nodule with prominent telangiectasias (Fig. 1.10d). The tumor is firm or rubber-like on palpation.
- Other disorders that can rarely present on the scalp are **seborrheic keratosis** (Fig. 1.10e), **intradermal nevus and verruca vulgaris**, **verrucous epidermal nevus** (Fig 1.10f).



Figs 1.10a to f: (a) Congenital melanocytic nevus on the scalp; (b) Nevus sebaceous involving the scalp and extending to the face; (c) Pilar cyst on the scalp. They lack a punctum; (d) Nevus comedonicus on the scalp; (e) A cerebriform plaque of seborrheic keratoses on the scalp; (f) Verrucous epidermal nevus (VEN) in girl. The lesion was present since birth

ACNE KELOIDALIS NUCHAE

The condition is seen exclusively in men and begins as chronic follicultis with formation of firm, dome-shaped follicular papules over posterior neck and occipital scalp (Fig. 1.11a). Gradually, these become hard and keloidal and may coalesce to form plaques (Fig. 1.11b). There may be associated loss of hair due to scarring.

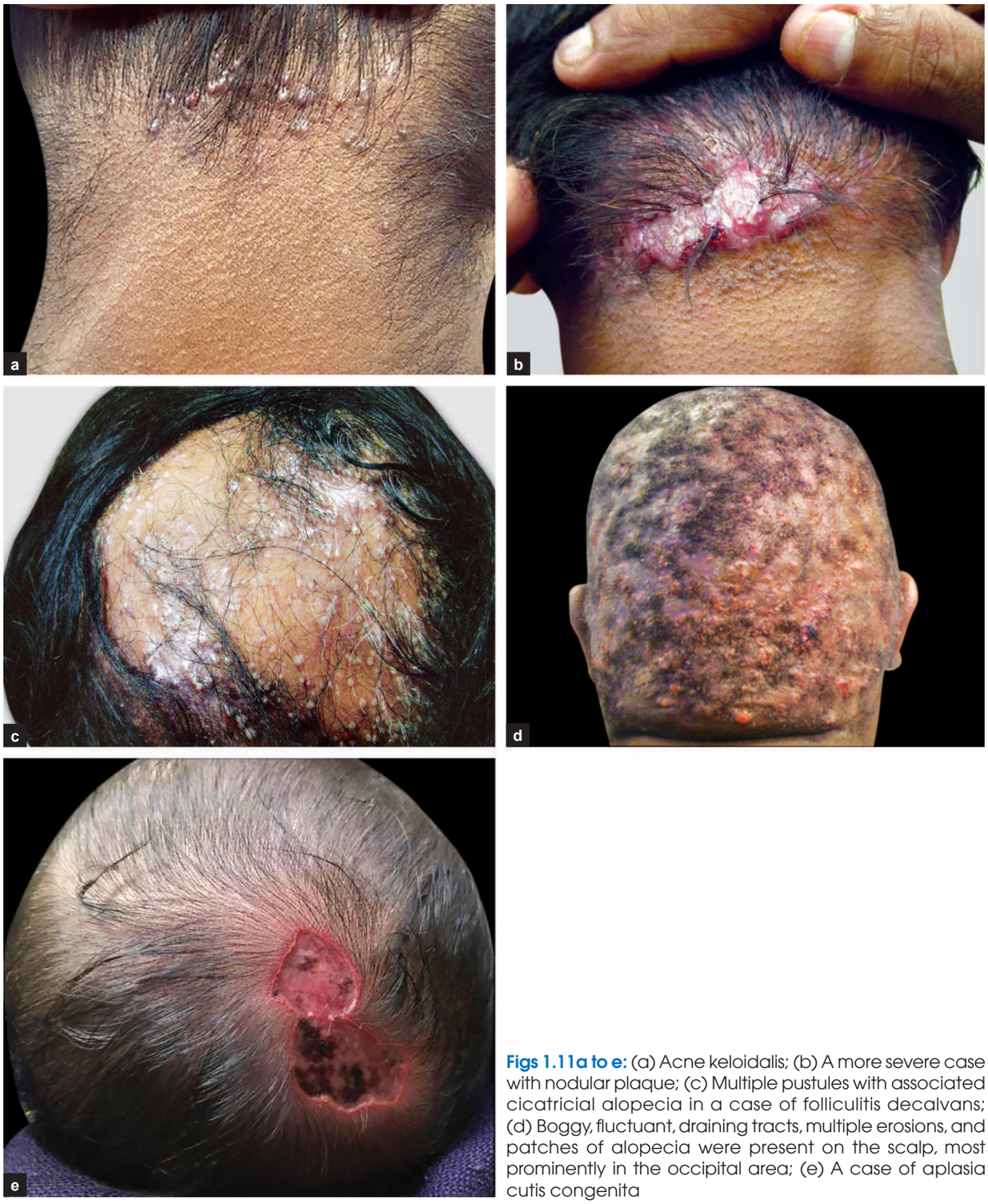
Management

Mechanical irritation of the area should be avoided. Patients should be advised not to wear headgear or shirts that irritate the posterior scalp. Options for treatment are topical tretinoin, mid to high potent topical steroid. Initial stages may respond to topical antibiotics whereas keloidal stage requires intralesional triamcinolone. Oral isotretinoin has been found to be effective in a few case reports.

A simple treatment option that can be employed involves injecting triamcinolone acetonide 5–10 mg/mL for the nodules and oral cycline antibiotics with a retinoid preparation at night.

MISCELLANEOUS DISORDERS

- **Linear morphea.**
- **Folliculitis decalvans:** Erythematous perifollicular pustules and scarring. Cause is unclear but may involve *S. aureus* (Fig. 1.11c).
- **Dissecting cellulitis of scalp:** Occurs most often in young black-skinned men. Firm, deep nodules become fluctuant, discharge malodorous pus, and develop interconnecting sinuses. May be associated with hidradenitis suppurativa and acne conglobata (Fig. 1.11d).
- **Aplasia cutis congenita (ACC):** There is congenital absence of skin with or without absence of underlying structures such as bone. It most commonly affects the scalp. Scalp ACC is divided into membranous and nonmembranous forms. Membranous ACC is notable at birth with a thin membrane often overlying a small nodule that can be filled with clear or serosanguineous fluid. This disorder warrants a specialist referral (Fig. 1.11e).



Figs 1.11a to e: (a) Acne keloidalis; (b) A more severe case with nodular plaque; (c) Multiple pustules with associated cicatricial alopecia in a case of folliculitis decalvans; (d) Boggy, fluctuant, draining tracts, multiple erosions, and patches of alopecia were present on the scalp, most prominently in the occipital area; (e) A case of aplasia cutis congenita