Hypertrichosis and hirsutism

Amanda M M Oakley MBChB FRACP DipHealInf

ABSTRACT

Excessive hair growth due to hypertrichosis should be distinguished from hirsutism, which occurs in an adult male pattern. Localised hypertrichosis is most often a component of a naevus. Generalised hypertrichosis may be genetic or caused by drugs or systemic illness. Hirsutism is frequently familial. It is often associated with seborrhoea, acne and sometimes pattern alopecia. In most cases circulating androgen hormones are normal as the hair growth is caused by increased end-organ sensitivity. Severe hirsutism due to excessive circulating androgens of ovarian, adrenal or exogenous origin may also be associated with virilisation. Hirsutism affecting the lateral face and areola is usually of ovarian origin and central hirsutism is more likely to be adrenal. Hirsutism may be managed medically with antiandrogens including the oral contraceptive pill, spironolactone and cyproterone acetate. Physical methods of hair removal include shaving, plucking, waxing, depilatory creams, electrolysis and laser epilation.

Keywords

Hirsutism, hypertrichosis, hair removal

*Clinical Associate Professor **Amanda Oakley** is Clinical Director of the Department of Dermatology of Health Waikato and in private practice in Hamilton. She has a particular interest in teledermatology and online medical education, preparing online courses in dermatology for the University of Auckland and managing the New Zealand Dermatological Society's successful website at http://www.dermnetnz.org. She has broad clinical interests.*

*

Introduction

The term hirsutism should only be applied to women with excessive growth of terminal hair in a male pattern. It is related to hormonal factors. A study in London reported hirsutism in 1–2% of women.[1] Hirsutism should be distinguished from hypertrichosis, a more general term used for excessive hair growth.

Hypertrichosis

Hypertrichosis can refer to fetal type of hair (lanugo), vellus hair or terminal hair. It may be generalised or localised, congenital or acquired.

Excessive lanugo hair is very rare and may be genetic in origin or a paraneoplastic phenomenon.

Congenital diffuse hypertrichosis can be a manifestation of an underlying genetic syndrome, fetal alcohol syndrome or due to maternal drug ingestion. However varying degrees are seen in otherwise healthy children of both sexes. Typically there is increased hair on face, trunk and limbs and a positive family history.

Congenital localised hypertrichosis may be isolated or seen in association with a melanocytic naevus. A lumbosacral 'faun tail' is usually a marker for spina bifida oculta or diastematomyelia.

Localised hypertrichosis presenting later in childhood may be a feature of Becker's naevus, a flat area of pigmentation on the shoulders that is more common in boys. Hypertrichosis may also arise because of repeated injury, friction or inflammation. It is occasionally a feature of the lichen simplex form of eczema but potent topical steroids may also provoke hair growth in treated areas. Topical minoxidil is used therapeutically because of the same property.

Hypertrichosis is sometimes seen in association with neurological disorders, anorexia nervosa, hypothyroidism and acquired immunodeficiency syndrome. Acquired hypertrichosis on sun-exposed areas is a feature of porphyria cutanea tarda, in association with hyperpigmentation, blisters and skin fragility.

Generalised hypertrichosis may be induced by a number of drugs, when it is usually reversible. These include glucocorticoids, minoxidil, anticonvulsants and ciclosporin.

Treatment of hypertrichosis is limited to shaving, plucking, depilatory creams containing barium sulfate or calcium thioglycolate, electrolysis and hair removal with lasers.

Hirsutism

Hirsutism is not present before the secondary sexual characteristics develop at puberty. Men grow hair on the face (moustache, beard), chest, shoulders, back, arms, thighs, pubic

and lower abdominal area and buttocks. This distribution of hair in women means that there is an increased secretion of androgens by the ovaries or the adrenal glands, exogenous anabolic steroids, or increased end-organ sensitivity. Dihydrotestosterone (DHT) is the intracellular androgen directly responsible for hirsutism via type 2, 5-alpha reductase.

Pituitary tumours may result in hirsutism due to production of ACTH or prolactin. Excessive adrenal cortisol can also be a cause of hirsutism. Congenital or late-onset adrenal hyperplasia is associated with increased dehydroepiandrosterone (DHEA) or DHEA sulfate, and pituitary production of excessive ACTH. Excessive ovarian androgens (progesterone, 4-androstenedione and testosterone) may be due to polycystic ovarian syndrome or rarely, tumours. Other tumours (carcinoid tumor, choriocarcinoma, and metastatic lung carcinoma) may also produce ectopic androgens. Hepatic disease can result in reduced circulating sex hormone binding globulin (SHBG) so that there is more free testosterone and greater conversion to DHT.

Constitutional hirsutism is the term used in affected women that have no detectable endocrine abnormality. It is thought to be due to increased end-organ response to normal plasma levels of androgens and is often familial.

Clinical features of hirsutism

Hirsutism is often associated with other manifestations of androgen activity i.e. oily skin (seborrhoea), acne, and sometimes, female pattern alopecia. The effects are generally noticeable between the ages of 16 and 20 and may be associated with obesity. Later and abrupt onset is more likely to be due to a tumour.

The illustrations provided by the Ferriman and Gallwey hirsutism chart may be used to score severity in each location (upper lip, chin, breasts, abdomen, arms, thighs, upper back, lower back). The higher the score, the greater the chance there is an organic cause for the hirsutism. The patient should be fully examined for signs of excessive androgenaemia or other endocrine disorder.

An increase in plasma free testosterone of ovarian origin most often results in hirsutism affecting the lateral face and breasts.

Hirsutism is present in 90% women affected by polycystic ovary syndrome. This is characterised by infertility, secondary amenorrhea or menstrual alterations and, in 50%, obesity. Many have acne. POCS is associated with decreased follicle stimulating hormone (FSH), increased luteinising hormone (LH), oestrone and testosterone, and sometimes, increased prolactin. There are no signs of genital virilisation in Type 1, autosomal dominant inherited POCS. Up to 5% of hirsute women have Type 2 POCS, with more marked hyperandrogenaemia, signs of virilisation, androgenetic alopecia, insulin resistance and acanthosis nigricans.

Lateral hirsutism presenting in an older woman and associated with virilisation is suggestive of an ovarian tumour.

Adrenal sources of androgen tend to result in hirsutism that is prominent on the central trunk (neck to pubis). Menstrual cycles tend to be longer than 30 days and affected women are often slender. High levels of adrenal androgen may result in male pattern androgenetic alopecia and virilisation. Early onset of pubarche and hirsutism suggest 21-hydroxylase deficiency, the commonest cause of congenital adrenal hyperplasia.

Cushing's syndrome, due to increased plasma cortisol of pituitary, adrenal or exogenous origin, is often accompanied by an increased production of androgens thus causing hirsutism and virilisation as well as typical fat distribution, hypertension, striae and purpura.

Hyperprolactinaemia due to pituitary adenoma may result in infertility, oligomenorrhoea (70%), galactorrhoea (50%) and both lateral and central hirsutism. Some patients also present with seborrhea and genital virilisation. Hyperprolactinaemia may also be due to drugs.

When the results of hormone tests are normal, the hirsutism is often confined to the face and there are no other androgenetic problems. Contraceptives containing progestagens or other drugs are responsible for some cases.

Investigations

Free androgen index (testosterone x 1000/SHBG), androstenedione, DHEA-S and prolactin are the minimum tests required to identify excessive circulating androgens and

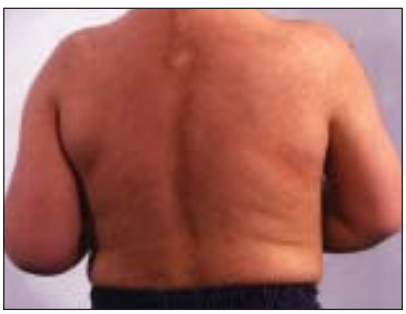

Figure 1. Congenital hypertrichosis

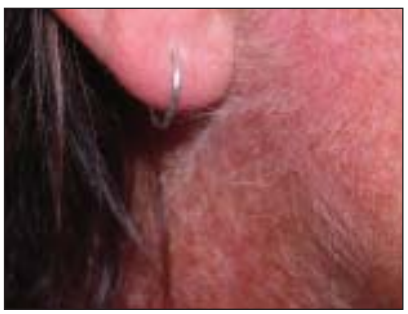

Figure 2. Hypertrichosis induced by ciclosporin

Figure 3. Facial hirsutism in patient with PCOS

help define their origin. If there is a significant abnormality, the patient may be referred to an appropriate service for further investigation. This may be dermatology, gynaecology or endocrinology depending on the patient's presentation.

Medical management[2]

In iatrogenic cases, the responsible drug may be discontinued. In patients presenting with an adrenal source of androgen, adrenal suppression may be achieved using dexamethasone at an initial dose of 0.5 mg every night for three months and then alternate nights for three months. Alternatively, prednisone may be prescribed at a dose of 7.5 mg daily for two months, reduced to 5 mg daily (for two months) and then 2.5 mg daily until six months of treatment are completed.

Ovarian suppression includes the use of combined oral contraceptives containing ethinyl oestradiol, and a progestagen, especially cyproterone acetate (Diane-35, Estelle 35). The oestrogen increases SHBG. The progesterone inhibits the secretion of FSH and LH. Both modify binding of DHT to its receptor.

Antiandrogens are contraindicated in pregnancy as they may cause feminisation of a male fetus. They may be combined with an oral contraceptive agent.

Spironolactone (Aldactone, Spirotone) is an aldosterone antagonist that also has antiandrogen activity. The dose ranges from 50–200mg/day and should be prescribed for at least six months. It is contraindicated in pregnancy. Side effects include irregular periods, loss of libido, increase in breast size, hyperkalaemia, headache and gastrointestinal complaints.

Specialist management for those with more severe hirsutism often employs a higher dose of cyproterone (50–100mg/day; Pacific Cyprotrerone, Androcur). This may be prescribed for days one to 10 of the menstrual cycle, combined with the ethinyl oestrodiol days one to 21. Postmenopausal women can take cyproterone without oestrogen on days one to 21, with a seven-day break. Drug Tariff funding requires Specialist prescription. Contraindications include pregnancy, liver disease and thromboembolic disease. Side effects include loss of libido, depression, fatigue, mastalgia, hypertension and weight gain.

Other drugs reported to be effective in the management of severe hyperandrogenism include flutamide, finasteride and gonadotropin-releasing hormone (GnRH) agonists. Bromocriptine may be used for hyperprolactinaemia.

There are at present no effective topical treatments for hirsutism available in New Zealand. Topical eflornithine cream (Vaniqa) twice daily has evidence to support its use and is available in Australia and elsewhere.

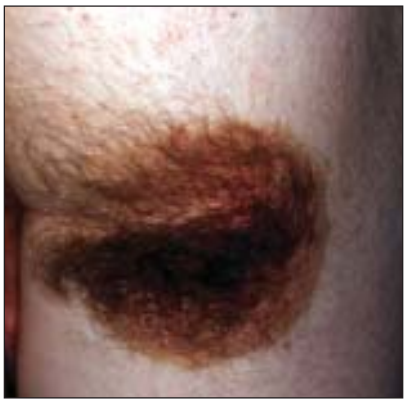

Figure 4. Localised hypertrichosis in congenital melanocytic naevus

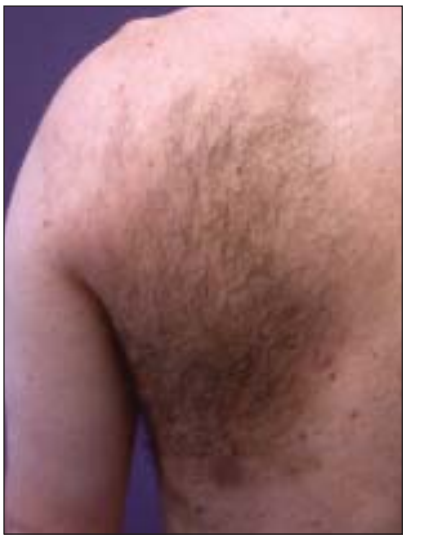

Figure 5. Localised hypertrichosis in Becker naevus

Physical treatments used by hirsute women

Hair removal products are very popular personal care products; a search on Amazon.com listed 1042 items. Depilate means to remove hair; epilate means to remove hair and hair root.

Hair bleaching may be carried out with 6% hydrogen peroxide or with a 20% ammonia solution.

Shaving and trimming are straightforward and inexpensive. However, women don't like to shave their faces because of the inevitable stubble requiring frequent attention.

Depilatory creams mostly contain 2–4% calcium thioglycolate (as in perming solution). Sugar and lemon juice may also be used. A thick layer

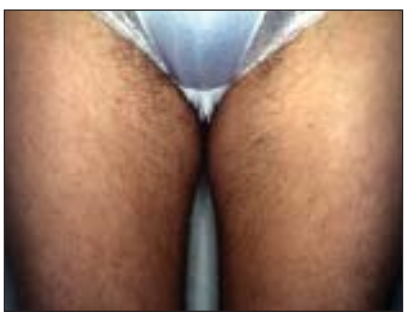

Figure 6. Hirsutism of thighs in patient with PCOS

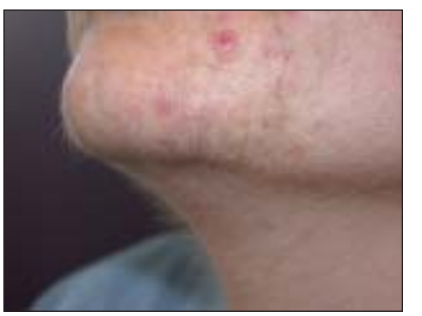

Figure 7. Hirsutism and acne in patient with normal hormone profile

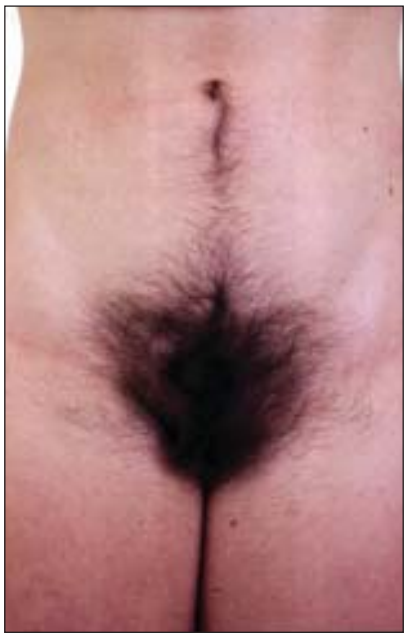

Figure 8. Male-pattern escutcheon due to adrenal androgens

is applied for 15 to 30 minutes then wiped off. Depilatory creams may cause irritant contact dermatitis and are poorly tolerated on facial skin. They need to be repeated every week or two.

Plucking with tweezers or an epilator may be used to remove hair in the line of its growth. Disrupting the hair root may result in an inflammatory papule, epidermal cyst or scarring.

Waxing needs to be repeated every six weeks. The warm wax hardens on the skin and as it is stripped off, the hairs are pulled out with it from the roots. Thermal burns, folliculitis and contact irritant or allergic dermatitis are potential problems. Waxing is contraindicated when taking oral isotretinoin.

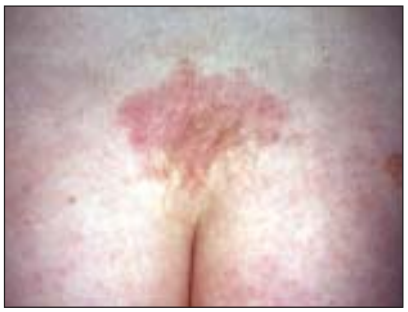

Figure 9. 'Faun tail', i.e. localised hypertrichosis, in association with a capillary vascular malformation and underlying spina bifida occulta.

Electroepilation may employ electocoagulation or thermolysis to destroy anagen hairs. It should be carried out by an appropriately qualified and experienced technician. It is uncomfortable, tedious and must be repeated frequently over long periods of time to be permanently effective. Inflammatory lesions, hyperpigmentation and scarring are the main risks.

Laser and light source epilation is best used to remove dark hair in untanned and naturally fair skinned individuals. Laser hair removal is less painful, quicker and more effective than electrolysis and can be used for large areas. Permanent hair reduction may occur after multiple treatments, but is less likely if the hair is fair in colour. Alexandrite, Ruby, Nd:YAG and diode lasers, and intense pulsed light (IPL) devices use selective photothermolysis, which targets the chromophore, melanin. Lasers and IPL are not effective for non-pigmented hair and can result in temporary hyperpigmentation, and temporary or permanent hypopigmentation of adjacent skin. Adverse effects also include Koebnerisation of skin conditions such as psoriasis, and rarely, scarring. Pre-cooling treated skin can reduce these complications. Regrowth is significantly slower than following conventional methods of hair removal.

Weight loss undoubtedly improves hirsutism in some hyperandrogenic women.

Conclusion

The majority of women complaining of excessive hair do not have excessive circulating androgens and should be advised to use physical methods to disguise and remove the hair. If they are hirsute, rather than presenting with non-hormonal hypertrichosis, the oral contraceptive pill and/or spironolactone may be of benefit. Patients with severe hirsutism are more likely to have other androgenetic symptoms due to an organic cause. They should be investigated and managed by an appropriate specialist.

References

1. Dawber R, Sinclair R. Hirsuties. Clin Dermatol. 2001; 19:189–99.
2. Camacho-Martinez F. Hypertrichosis and Hirsutism. In: Bolognia J, Jorizzo J, Rapini R, editors. Dermatology. CV Mosby; 2003. p. 1051–1060.

The discipline of general practice is founded in the long term relationship between doctor and patient, which nurtures mutual respect and trust. Patients can trust doctors only if they feel heard and understood, and this can happen only if doctors, in turn, trust patients to give a truthful account. If general practitioners feel themselves to be under pressure and begin to regard every request for a sick certificate as suspicious, trust is very rapidly undermined and, once gone, is difficult to rebuild. Increasingly general practitioners feel themselves to be the functionaries of an oppressive economic system and obliged to control access to the benefit system, a position that conflicts with a responsibility to act as the individual patient's advocate.

Nilsson B. Patients, doctors, and sickness benefit. BMJ 2003;327:1057.