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Case Report

A Case of Alopecia Areata in a Patient with Turner Syndrome

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Abstract

The Authors report a case of alopecia areata totalis in a woman with Turner syndrome.

Introduction

Alopecia areata (AA) is an inflammatory non-scarring form of hair loss, which may involve all the follicular units of the body. It is a common disease, with an incidence of 1-2% [1].

AA may affect people of both sex and all age, even if it is more commonly described in under 30-years old patients [2]. Often, it is associated with different inflammatory or autoimmune diseases, like atopic eczema, Hashimoto's thyroiditis, Graves' disease, Celiac disease, vitiligo, psoriasis and others [1].

AA is an autoimmune disease, characterised by a T-cell mediated immune response that targets hair follicles. The clinical manifestations are numerous, ranging from mild lesions, characterised by round or oval patches of hair loss, to the total hair loss, such as in the universal form [3].

Case report

A 28-year-old woman affected by AA, showed up to our Clinic presenting a widespread non-scarring alopecia, which was localised in the scalp and face area. The skin was white-pink in colour and normal-trophic. Black dots and exclamations points were no detected. Only a few vellus-like hairs were present in the midline of the scalp (Fig.1).The patient did not report any subjective symptoms and enjoyed apparent good health.

She reported the appearance of the disease about 26 months ago. Initially, the hair loss was limited to the left temporal side, and consisted in two round little (diameter less than 2 cm), well defined, areas. After few weeks, the hair loss rapidly spread, involving the entire scalp. By five months, the patient also observed the loss of her eyelashes and eyebrows.

The woman told us to be affected by Turner syndrome, Hashimoto's thyroiditis and celiac disease. Her medical treatment consisted of oral levothyroxine

and oestrogen replacement. No other diseases had been reported. She had no familiarity for AA or other autoimmune diseases.

The patient reported previous treatment of alopecia with oral supplements and topical minoxidil 2% 1 ml twice a day for six months. Due to the lack of clinical improvement, she was treated with low dose of topical corticosteroids (hydrocortisone once a day) for three weeks and PUVA therapy, which has been performed twice a week for a total of 20 sessions, both without beneficial results.

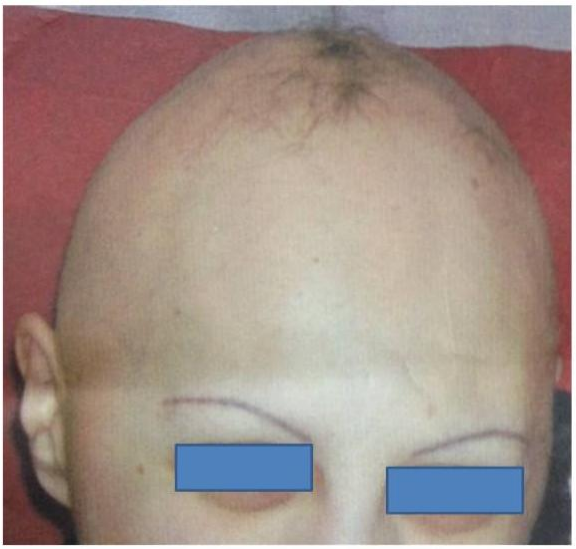

Figure 1: Alopecia areata in a woman with Turner syndrome

During the clinical evaluation, the woman showed characteristic features of Turner syndrome, such as short stature (150 cm), short and squat neck, poor breast development and stubby little hands. She had ears and lower eyelids bigger than normal, enophthalmos, reduced upper lip and a small chin. On the other hand, no other skin lesions were observed. Nails were smaller than normal. A routine laboratory test, including thyroid function tests, were normal.

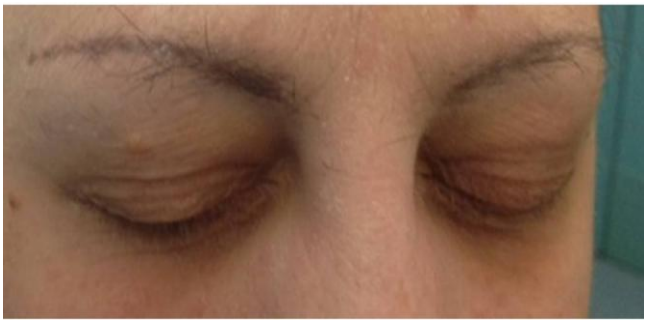

Figure 2: Eyelashes and eyebrows re-growth after cyclosporine A therapy

Due to the long duration of the hair disease, we applied intralesional triamcitolone acetonide once a week for two months. Surprisingly, even if localised, we observed a re-growth of the hair. Since hair growth responded to corticosteroids, we switched to oral methylprednisolone 32 mg/die for ten days, followed by 16 mg/die for ten days and, eventually 8 mg/die for another ten days. The clinical response was quite good: the patient showed new hair in the frontal area of the scalp.

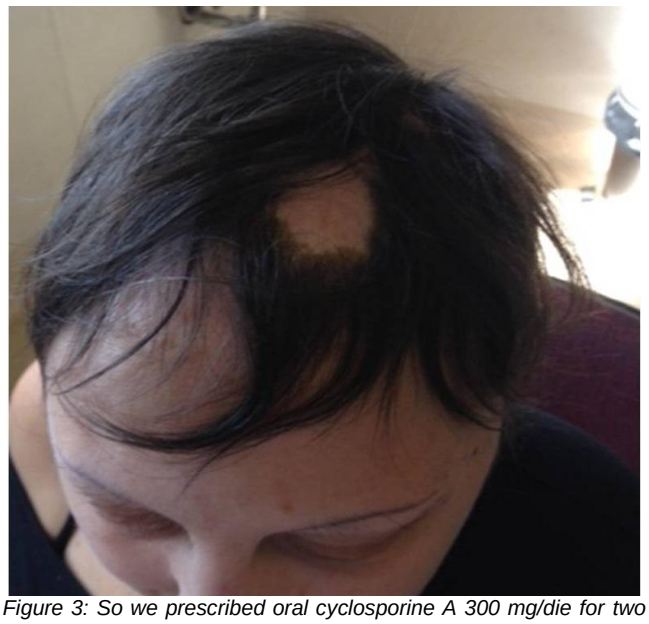

Figure 3: So we prescribed oral cyclosporine A 300 mg/die for two months, and intralesional triamcitolone acetonide once every two weeks. Due to the good clinical response to the therapy and the possible side effects of corticosteroids, after two months, we decided to prescribe the only cyclosporine A 200 mg/die

After one month, we observed the diffuse hair re-growth on the scalp. New eyelashes and eyebrows were also observed (Fig.2).

We decided to reduce progressively the cyclosporine therapy and to stop it. Because of the appearance of new hair loss patches (Fig.3), the patient re-started the treatment with cyclosporine A at a dosage of 200 mg/die. The clinical response was rapid and excellent (Fig.4).

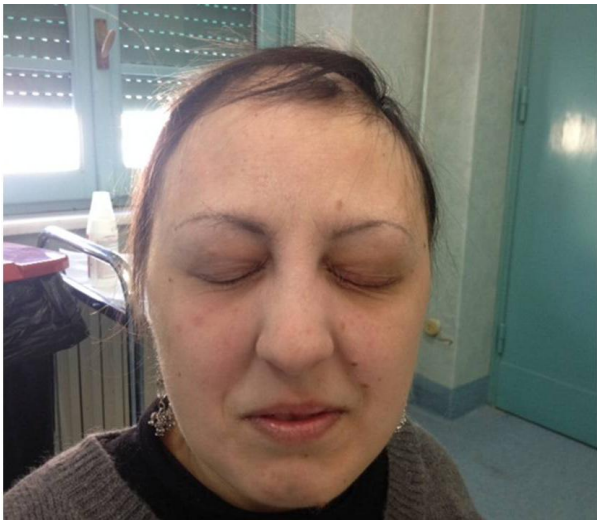

Figure 4: Our patient with complete hair re-growth

During the treatment we constantly evaluated the patient's clinical conditions, monitoring her blood pressure and routine blood test. No side effects or complications had been observed.

Discussion

Turner syndrome (or Ullrich-Turner syndrome) is one of the most frequent chromosomal abnormalities, which results from a sex-chromosomal anomaly characterised by a presence of one normal X chromosome and a missing or structurally abnormal second sex chromosome. It is a rare disease affecting 1:2500 live born girls [4].

Table 1: Characteristic clinical features of Turner syndrome

Skin and adnexa	pterygium; ↑ moles; vitiligo; ↑ growth of body hair (forearm); low-set hair; ↓ hair density; congenital lymphedema of the hands and feet; over the curvature of the nails
Mouth	palate with a pointed arch form ("Gothic" palate); ↓ development of the jaw; mouth as a "carp."
Ears	low implantation; ↓ development of the ear's board; large ears; others malformations; neurosensorial defects
Eyes	alteration in the position and shape of the eyelid (hypertelorism and epicanthal folds); strabismus; dyschromatopsia
Neck	short and squat
Skeletal system	short fingers; widened distal phalanges; ↓ length of the fourth metacarpal bone (XR Archibald sign); cubitus valgus; medial condyle of the tibia agenesia (XR Kosowicz sign); delayed bone maturation (first three years of life, after ten years)
Chest	enlarged; pectus excavatum; hypoplasia and the excessive distance between the mammary areola
Urinary system	horseshoe kidney; renal cysts; unilateral renal agenesis; pelvis and ureters alterations
Cardiovascular system	bicuspid aortic valve; aortic coarctation; aortic valve disease; anomalous venous return of the pulmonary veins; mitral valve prolapse; hypertension; conduction defects
Blood	abnormalities of coagulation factors
Metabolism	abnormal lipid profile (cholesterol, triglycerides) and glucose
Nervose system	defects in visual-spatial and visual-perceptual skills; ↓ motor function (unable to walk before 15 months); ↓ non-verbal memory; ↓ attention
Psychological	disorders in emotional

The Turner syndrome phenotype includes female gender, short stature, primary ovarian failure and some characteristic physical features (tab.1) [5]. Patients with Turner syndrome have an increased incidence of autoimmune disorders (AID), such as Hashimoto's thyroiditis, Grave's disease, celiac disease, inflammatory bowel disease, and diabetes mellitus [6-8].

Even if dermatologic autoimmune diseases (e.g. psoriasis, vitiligo, halo nevi) are well-known in Turner patients (9, 10), only a few cases of associated AA have been reported so far [11-13].

AA is a chronic inflammatory autoimmune disease, characterised by non-scarring hair loss on the scalp or any hair-bearing area of the body. Clinically it may represent in variable patterns, such as patchy, diffuse, reticulate, linear, or opiates-type. Depending on the severity of hair loss, AA may also be classified as localised (few patches of hair loss), subtotal (diffuse alopecia of the scalp), total (complete loss of scalp hair) or universal (complete loss of body hair) [14].

Table 2: Therapeutic options for alopecia areata

Classical treatments	Corticosteroids (topical, intralesional, systemic)
	PUVA-therapy (topical or systemic 8-Methoxypsoralen + UVA)
	Topical immunotherapy (diphenylcycloproperone/DPCP, squaric
	acid dibutyl ester/SADBE)
	Antralin cream or ointment
Aneddotic treatments	Antidepressant
	Hypnotherapy
	Psychological support
	Topical minoxidil
	Topical triiodothyronine
	Garlic gel
	Azelaic acid
	Topical onion juice
	Imiquimod
	Botulinum toxin
	Photodynamic therapy
	Topical phenol
	Excimer lasers
Promising treatments	Azathioprine
	Methotrexate
	Cyclosporine A (topical, systemic)
	Tacrolimus
	Pimecrolimus
	Alitretinoin
	Biological therapies
	Platelet-rich plasma
	UVA1 (340-400 nm)

The course of the disease is highly variable: it may spontaneously regress, be stable or progress to a severe form. Even if numerous treatment options are now available for AA (tab.2), no definitive therapy exists [15, 16].

In our case, the patient showed an excellent clinical response to cyclosporine A treatment, initially combined with intralesional corticosteroids.

In conclusion, the authors have presented this case study to record the possible association of AA with Turner syndrome, and therapeutic validity of cyclosporine in stimulating hair growth where other therapies had previously failed.

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