Acute alopecia totalis

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SUMMARY

From a group of 1,189 AA patients seen in our dermatology unit, thirteen (3 males, 10 females) experienced hair shedding that started profusely and diffusely over the entire scalp. They were under observation for about 5 years, histopathology and trichograms being performed in all instances.
The mean age of the patients was 26.7 years. It took only 2.3 months on average from the onset of hair shedding to total denudation of the scalp. The trichogram at the time of diffuse shedding showed that about 80% had dystrophic roots and the remaining 20% had telogen roots. Histopathological findings and exclamation mark hairs were compatible with alopecia areata.
Regrowth of hair was noted 3.2 month after the onset of hair shedding and recovery observed in 4.8 months. All patients were treated by methylprednisolone pulse therapy. During the follow-up period, 53 months on average after recovery, 8 of the 13 patients (61.5%) showed normal scalp hair without recurrence, in 4 patients the recovery was cosmetically acceptable in spite of focal recurrences and only 1 patient showed a severe relapse after recovery. Considering all of the above findings, this group of the patients should be delineated by the term acute alopecia totalis.

KEY WORDS

alopecia, diffuse, acute, total

Introduction

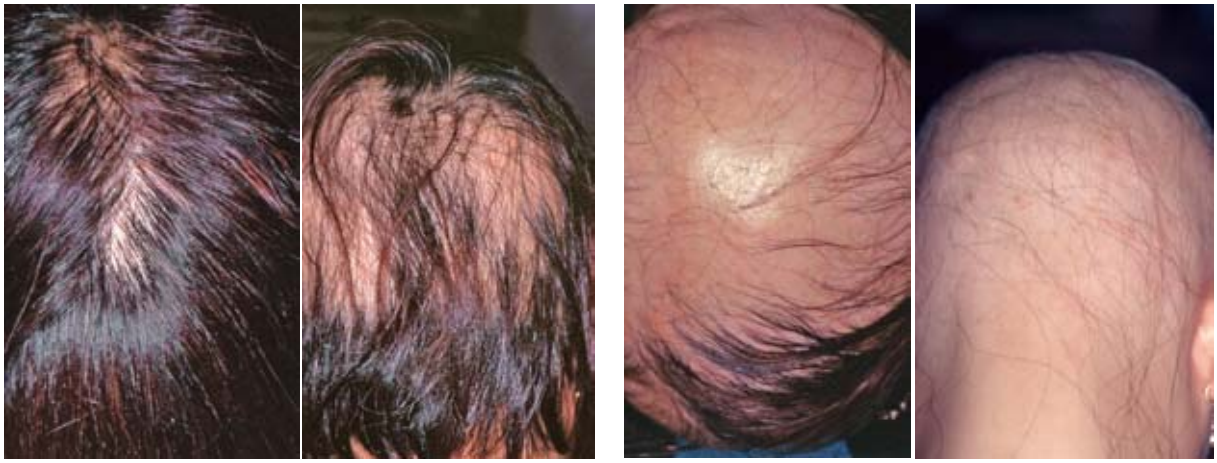
Diffuse alopecia areata (AA) is well known, but less well characterized is its acute form (1-4). Sato-Kawamura and associates (5) called it *acute diffuse and total alopecia of the female scalp (ADTAFS)*, considering it to be a new subtype of diffuse alopecia areata. They characterized 9 cases of ADTAFS as follows: occurrence limited to females, tissue eosinophilia and short clinical course. We have evaluated patients with AA whose hair shedding was the same as that in ADTAFS and further characterized this type of AA with acute diffuse hair shedding leading to total scalp denudation. We found it in men as well as women.

Patients and methods

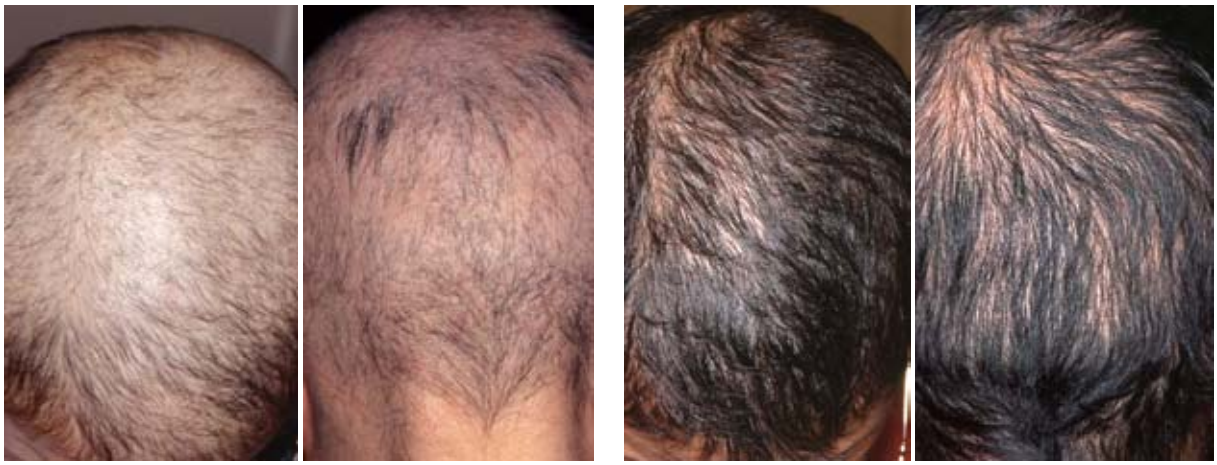
Among the 1.189 patients with AA who visited us, from January 1989 to December 2002, we identified

Figure 1. Trichogram of the hair roots on the first examination, the hairs were shedding diffusely and profusely. No intact anagen hair root was seen.

13 cases of acute diffuse hair loss leading to rapid total denudation. We checked the period of time from the onset of the alopecia to totally denudation, to the emergence of new hair, and to the recovery of the scalp hairs with follow-up observation. Recovery was defined by terminal hairs being thicker than 60 microns in diameter and longer than 1 cm in length covering more than 80% area of the scalp. Scalp biopsy specimens, 60 vertical sections for each biopsy, were examined in 12 patients. Clinical histories with physical and laboratory examination were done to check if there were any possible contributing diseases to the acute hair loss such as toxic drugs or illnesses. Family history revealed no cases of alopecia areata in their families.



A. 1 month B. 2 months



C. 4 months D. 5 months

Figure 2. Clinical pictures of case 10, frontal and occipital views.
A. 1 month from the onset of hair shedding. Although the hair falling was diffuse, pillow-rubbing made it more prominent. B. At 2 months, almost alopecia totalis. C. At 4 months, emerging new hairs. D. At 5 months, the recovery of terminal hairs is progressive.

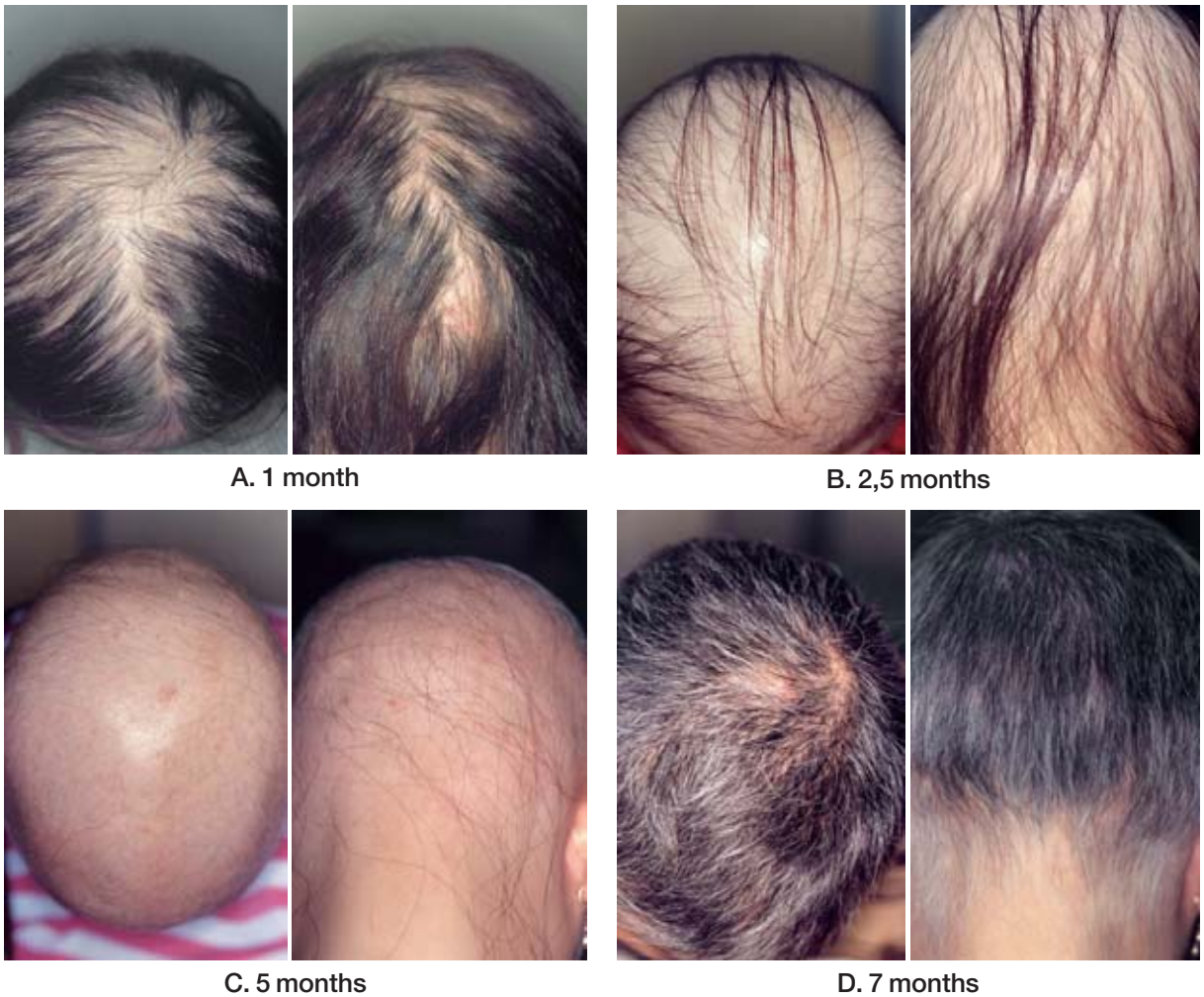
Results

The mean age of patients was 26.7 years. Eight were in first to third decade of their age. There were 3 males and 10 females. The time from the initial onset of excessive hair shedding to the first visit was 1.0 month on average. At the time of the first visit many of the patients' hair looked almost normal at a glance, but a simple hair pull test revealed that the hair was easily plucked out without resistance. Although several patients showed more prominent hair thinning areas with diffuse shedding over the scalp, there were no smooth circumscribed bald patches of alopecia areata.

The trichogram invariably showed that approximately 80 % of the examined hairs had dystrophic roots. The rest of them were club hairs (Figure 1). It took only 2.3 months on average after the onset of the hair shedding to produce a totally denuded bald scalp. Newly emerging hairs with tapered tips were recognized in 3.2 months on average from the initial onset. The recovery of scalp (terminal hairs thicker than 60 microns and longer than 1 cm over more than 80% of scalp) appeared in 4.8 months on average from the onset.

The scalp biopsy specimens showed a peribulbar lymphocytic infiltration in 11 (91.7 %) patients, with included eosinophils in 6 patients. Trichomalacia were found in 7 patients. The exclamation mark hairs were seen in all cases in the period of hair shedding; 9 patients showed the exclamation mark hairs at the first visit and the remaining 4 during the following 4-6 weeks. At the time of the first visit, 3 patients described a pruritic scalp from onset; one patient also experienced painful sensations. Nail abnormalities including pitting nails were not found.

The diagnosis of AA was substantiated by histopatho-



A. 1 month B. 2,5 months

C. 5 months D. 7 months

Figure 3. Clinical pictures of case 11, frontal and occipital views
A. 1 month from the onset of diffuse shedding. B. At 2.5 months. C. At 5 months. D. At 7 months. A and B may be called diffuse type alopecia areata, but the trichogram revealed no intact anagen roots at the time. After the stage of alopeca totalis, the full recovery followed.

Table 1. Clinical course of the 13 cases of acute alopecia totalis

	Sex	Age	Duration from onset of hair loss (months)				Duration of follow-up after recovery	Result
			First visit	Becoming alopecia totalis	Emergence of new hair with tapered tip	Recovery of scalp hair (Terminal hairs longer than 1cm over 80% of scalp)		
1	M	56	0.3	2.5	3.5	6.0	23.3	Excellent
2	M	17	1.0	1.75	3.0	5.0	68	Excellent
3	M	19	1.0	2.0	3.0	5.0	168	Good
4	F	13	1.0	2.0	3.0	5.0	16	Poor
5	F	16	0.3	1.0	4.0	6.0	72.3	Excellent
6	F	52	2.0	4.0	4.0	5.0	20	Excellent
7	F	10	2.0	2.5	2.5	3.5	29.5	Good
8	F	5	0.3	2.0	2.5	5.0	53.5	Excellent
9	F	31	2.0	3.0	3.0	4.0	56	Excellent
10	F	47	0.75	2.5	3.5	4.0	2.75	Excellent
11	F	22	1.0	3.0	4.0	6.0	11	Good
12	F	19	0.5	1.0	2.0	4.0	155.5	Good
13	F	40	0.25	2.0	3.25	4.0	14.25	Excellent
Mean		26.7	1.0	2.3	3.2	4.8	53	
Cases of Sato-Kawamura (5)		34	2.	3.1	2.4	6.0	(-)	

logical findings and the presence of exclamation mark hairs. All the patients were otherwise in good health and there was no history of potentially incriminating medications or of severe emotional stress. One patient had coexistent pulmonary tuberculosis and another had atopic dermatitis. Complete blood counts and blood and urine chemistry tests were within normal limits.

Treatment

All cases were treated with a total of three doses of intravenous methylprednisolone pulse therapy, 12.5 - 25mg/kg body weight, either on three consecutive days or once weekly, during a three week period. This regimen was introduced in its early stage when hairs were acutely shedding. For the intravenous injection methylprednisolone sodium succinate was mixed with 5% dextrose water 180ml, 90ml in children, and given over 60 minutes. The patients were not admitted to the hospital for the injection. After finishing the pulse administration, triamcinolone oral tablet 12 mg per every second day was given for a month. Follow-up periods were 53 months on average after the recovery of scalp hairs. The result through the follow-up period was defined as follows: excellent if complete recovery without recurrence; good if focal recurrence with keeping enough hairs for hair styling; poor if a severe recurrence was observed.

During a mean follow-up period of 53 months (minimum 11 months, maximum 168 months), the recovery of scalp hairs was as follows: 8 cases excellent, 4 good, 1 poor. The clinical characteristics of these cases are summarized in Table 1; clinical photographs of two are presented (Figures 2 and 3).

Discussion

Alopecia areata (AA), most probably an autoimmune condition, presents a difficult clinical challenge. Recognizing the clinical subtypes and their characteristic patterns can be helpful for management because the prognosis varies considerably (6, 7). AA is commonly classified into the following subtypes by the extent of the involvement: round or oval patches of hair loss (*alopecia areata circumscripta*), loss of all terminal scalp hair (*alopecia totalis),* and loss of all scalp and body hair (*alopecia universalis).* The alopecic scalp can be further subdivided into one of the followings by the pattern of hair loss: patchy, reticular, ophiasis, diffuse, and total. The appearance and clinical course of the each subtype are well known. For example, the patchy type shows one or several circumscribed smooth bald

patches of hair loss and is easier to treat than the reticular or ophiasis type, whereas alopecia universalis with complete loss of all body hairs is not responsive to treatment. The diffuse variant is not so well characterized in its clinical manifestation and course.

In general AA is most common between the second to fourth decades of life (1-4,8). The ages of the presented cases and those of Sato-Kawamura were essentially in the same range. Both sets also showed the histopathological hallmark of alopecia areata: peribulbar lymphocytic infiltration, frequently including eosinophils, and exclamation mark hairs. But, contrary to all other subtypes of AA, the course of the disease, from the onset to the recovery, was relatively short. Recovery of hairy scalp occurred within 6 months from the initial hair shedding in all our cases, which were treated with systemic pulse methylprednisolone.

The term diffuse AA is used because there are some remaining scattered hairs on the scalp at the time of examination. But the remaining hairs have either dystrophic or telogen roots as shown in the trichograms (Figure 1). These hairs appear uninvolved only in their external appearance, their roots being already affected. That is why a totally denuded surface soon appeared. The diagnosis of diffuse AA was made by observing loosely attached scalp hairs. Thus, we favor the term *acute alopecia totalis*, as it better suits such patients. It implies loss of all scalp terminal hairs (alopecia totalis) due to an acute process within a period of a couple of months.

The correct diagnosis is suggested by medical history, the presence of exclamation mark hairs, changes in trichogram and the rate of excessive hair loss. An exclamation mark hair, or a characteristic broken hair with club root which is known to appear in early active stage of alopecia areata, is not seen at the beginning stage of hair shedding. It needs several weeks to form (9-11). In all our patients the exclamation mark hairs were seen, although not in four of them at the first visit, but rather only after 4-6 weeks. The duration from the initial hair shedding to the first visit in these four was only 0.4 months on the average. The differential diagnosis includes acute telogen effluvium, anagen effluvium after anticancer drugs, and androgenetic alopecia (3).

Conclusion

We favor dividing alopecia totalis into two types*: the hair loss with almost simultaneous involvement of all terminal hair follicles of the scalp* as discussed here, and *the chronic persistent type,* which is characterized by a progressive worsening of the alopecia areata.

Our cases were treated with high dose systemic corticosteroid pulse therapy, as we consider the first type to be an acute and fulminating inflammatory phase of alopecia areata. The corticosteroid pulse therapy is regarded to be effective in rapidly evolving AA but not in severe longstanding AA (11-13), but we could not be sure if early high dose corticosteroide had changed the course of the disease, which otherwise might have persisted. Usually in the treatment of AA, it is common to see recurrences of the alopecia with the cessation of corticosteroids, while in the majority of the patients presented here, hair regrowth continued for about 4 years, if the high corticosteroid dose was given in the very early stage.

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