A Comment on Frontal Fibrosing Alopecia (Axel Munthe’s Syndrome)

Ralph M Trüeb

Address for correspondence:
Prof. Ralph M Trüeb,
Center for Dermatology and Hair Diseases Professor Trüeb,
Bahnhofplatz 1A, CH-8304 Wallisellen, Switzerland.
E-mail: rtltrueeb@derma-haarcenter.ch

ABSTRACT

Frontal fibrosing alopecia (FFA) represents a peculiar condition with a quasi-symmetrical, marginal alopecia along the frontal and temporal hairline with scarring. Steven Kossard is credited with the original description of the condition in 1994. Since its first description, FFA has become increasingly common while its etiology has remained obscure. While FFA has been related to lichen planopilaris, in fact, the pattern of clinical disease presentation might be more specific for the condition than the underlying inflammatory autoimmune reaction. It has been speculated as to whether FFA existed before Kossard’s original report since artistic depictions dating from the 15th to 16th century often show a high frontotemporal hairline. Rather, these represent fashionable forms of frontal pseudo-alopecia than FFA. Nevertheless, there is compelling evidence that FFA existed well before 1994 from the Swedish Physician Axel Munthe’s (1857–1948) account of the distinctive features of the condition in his book of memoirs “The Story of San Michele.” Therefore, Axel Munthe is to be acknowledged the first description of FFA in 1929.

Key words: Axel Munthe, fashionable forms of frontal pseudo-alopecia, frontal fibrosing alopecia, Mamsell Agata

“What you keep for yourself, you lose. What you give away, you keep forever.”

Axel Munthe; The Story of San Michele

Frontal fibrosing alopecia (FFA) represents a peculiar condition with a quasi-symmetrical, marginal alopecia along the frontal and temporal hairline with scarring. Affected women typically present with the complaint of asymptomatic, progressive recession of the frontal hairline and loss of eyebrows. Steven Kossard is credited with the original description of the condition in 1994, when he reported six postmenopausal women with progressive frontal hairline recession, that was associated with perifollicular erythema within the marginal hairline, producing an FFA extending to the temporal and parietal hair margins. Scalp biopsy specimens revealed histologic features that were indistinguishable from those seen in lichen planopilaris, in the absence of associated lesions of lichen planus. Eventually, Kossard interpreted this type of alopecia as a frontal variant of lichen planopilaris on the basis of extended immunohistochemical studies.

Originally considered to be an uncommon condition, the number of cases of FFA has exploded exponentially worldwide while its etiology has remained obscure. The condition has meanwhile also been described in premenopausal women and in men,[1-7] and most recently in children,[8] though with a significantly lesser frequency. Moreover, it has been recognized to represent a more generalized rather than localized process of inflammatory scarring alopecia, with extension beyond the frontotemporal hairline to include the parieto-occipital hairline, involve peculiar facial papules as evidence of facial vellus hair involvement,[9] and loss of peripheral body hair.[10,11] More recently, also lichen planus-type nail involvement has been reported,[12] again pointing to a close relationship of FFA to lichen planus.

A recent questionnaire-based study suggested a possible association between FFA and the use of facial skin care products.[13] This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

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How to cite this article: Trüeb RM. A comment on frontal fibrosing alopecia (Axel munthe’s syndrome). Int J Trichol 2016;8:203-5.
products, particularly sunscreens,[13] but the causality of this relationship remains to be confirmed. Familial cases of FFA[14‑16] point to the possible contribution of hereditary factors, probably related to androgenetic alopecia.

Finally, cutaneous lupus erythematosus has been reported to be capable of presenting as FFA,[17‑19] suggesting that the pattern of clinical disease presentation might be more specific for the condition than the underlying inflammatory autoimmune reaction. Moreover, it has been discussed to what extent again a background of androgenetic alopecia may contribute to this particular clinical presentation of the disease,[17] with possible implications for treatment.

As yet, there is no cure for FFA, though a number of treatments have been proposed to control symptoms or halt disease progression:[3‑7,20‑23] where androgenetic alopecia represents a comorbid condition, topical minoxidil, and oral dutasteride may be of benefit, in addition to topical and/or systemic anti-inflammatory treatment modalities (topical or intralesional corticosteroids, topical calcineurin inhibitors, and oral doxycycline, hydroxychloroquine, or mycophenolate mofetil, respectively). Eyebrow regrowth in patients with FFA could be achieved with intralesional triamcinolone acetonide.[24]

It has been speculated as to whether FFA existed before Kossard’s original report since artistic depictions dating right from the 15th into the 16th century often show a high frontotemporal hairline. Nevertheless, these rather represent fashionable or cultural forms of frontal pseudo‑alopecia[25‑26] than a true pathologic condition, since the hair was then shaved around the hairline or plucked at the temples and the napes of the neck, and the condition later disappeared again from the arts. Besides the well-known portrait of Eleonora Gonzaga, Duchess of Urbino (1493–1570),[27] typical examples include King Henry V of England (1387–1422) [Figure 1a], and Elizabeth Woodville (1437–1492), Queen consort of England and spouse of King Edward IV of England from 1464 until his death in 1483 [Figure 1b].

Nonetheless, there is more compelling evidence that FFA existed before 1994 from celebrated Physician Axel Munthe’s (1857–1948) description of his housekeeper Mamsell Agata in his autobiographical account. “The Story of San Michele” (originally published in English language in 1929): “To describe what she looked like is beyond me. She had thin golden locks arranged in a sort of early Victorian fashion – Rosalie said it was a wig but I do not know. An exceptionally high and narrow forehead, no eyebrows.”[28]

Swedish‑born Axel Munthe went to medical school in France and then opened a medical practice in Paris. He later assisted in the 1884 cholera epidemic in Naples. In 1887, he managed to purchase the ruined chapel of San Michele in Anacapri, Italy, and subsequently spent much of his life on the Island of Capri building his celebrated Villa at the site [Figure 2]. Munthe also had a fashionable medical practice in Rome on the Piazza di Spagna to help finance for construction. The “Story of San Michele” consists of a series of overlapping vignettes, loosely but not consistently in chronological order, recollecting periods of his eventful life. He related with a number of celebrities of his times, all of whom figure in the book, including: Eminent French Neurologist Jean‑Martin Charcot (1825–1893), also called “the Napoléon of the neuroses,” sophisticated French Chemist and Microbiologist Louis Pasteur (1822–1895), renowned for his achievements in rabies vaccine research.

Figure 1: Fashionable forms of frontal pseudo‑alopecia: (a) King Henry V of England and (b) Elizabeth Woodville, Royal Collection, Windsor.

Figure 2: In the atrium of the Villa San Michele at Anacapri: The author (far right) together with hair and nail experts Jerry Shapiro, Eckart Haneke, and Bertrand Richert‑Baran (second from left to right).
and the development of pasteurization, and French Naturalist Writer Guy de Maupassant (1850–1893), who is remembered as a master of the short story. Munthe also associated with the underprivileged of society, including Italian immigrants in Paris and plague victims in Naples, as well as rural people such as the islanders of Capri, and the Nordic Laplanders. Axel Munthe’s writing is light-hearted, being mainly memoirs drawn from real-life experience. He primarily wrote about people and their idiosyncrasies, portraying the foibles of both the rich and privileged and the poor and disadvantaged. Munthe died at Stockholm Palace, Sweden, aged 91, after serving as appointed physician to the Swedish royal family. In particular, he attended the Crown Princess Victoria of Baden (1862–1930) as personal physician and continued to do so when she became Queen consort of Sweden until her demise.

The reliability of the literary source and the distinctive features of FFA as described by Axel Munthe in the “Story of San Michele” are compelling to the extent that one should acknowledge that the condition did exist well before 1994, and propose to eponymize it “Axel Munthe’s syndrome” in recognition of the legacy of the personal reminiscences and medical observations recorded in his timeless book for posteriory.

Financial support and sponsorship

Travel costs of the author to the Capri Hair and Nail and Anti-Aging Practical Course by appointment of Course President Antonella Tosti were supported by Galderma, Italy.

Conflicts of interest

There are no conflicts of interest.

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