Frontal fibrosing alopecia

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First described in 1994 by Kossard, frontal fibrosing alopecia (FFA) is a type of cicatricial alopecia. This condition is generally observed in post-menopausal females and recent studies suggest that it is increasing in occurrence.

These patients often seek treatment by hair restoration surgeons. Unfortunately, the medical treatments for this disorder are often disappointing, and while initial surgical treatment may seem promising, the results may turn out to be ineffective because the disease can reactivate after the transplant.

We feel that it is important for hair restoration surgeons to be able to recognize this disorder, carefully evaluate the patient, and perhaps suggest medical therapy or suitable referral before considering a surgical treatment. In this article, we offer an approach to FFA using a question and answer format that allows concise responses.

Q. What is cicatricial alopecia?

Cicatricial alopecias are scalp disorders that cause not only loss of hair follicles but also scarring of the scalp tissue. The process is an inflammatory one. Cicatricial alopecias are usually divided by the type of inflammatory cells involved, which are either lymphocytes or neutrophils, but at times the response is a mixture of cells. FFA is considered to be a lymphocytic cicatricial alopecia.

Q. Why is FFA considered to be a form of lichen planus?

Lichen planus is a skin disease of unknown origin. It is considered to be a papulosquamous skin disease. The skin lesions may have a characteristic skin marking referred to as Wickham's striae. Lesions often appear in the mucosal areas and on the wrists and ankles. When it affects hair follicles, the disorder is referred to as lichen planopilaris (LPP).

FFA appears to be a form of LPP. When examining a histologic sample of skin affected by FFA, there is a pattern of inflammation indicative of lichen planus. It is a band like area of inflammation with lymphocytes surrounding the infundibulum and isthmus regions of the hair follicles. There is an accompanying loss of sebaceous glands. It seems that vellus and intermediate hairs are more prone to destruction.

Another form of LPP is Graham-Little-Piccardi-Lasseur Syndrome. The cause of this entity is also unknown but there is a case reportedly occurring after Hepatitis B vaccine. The syndrome consists of three signs: scarring scalp hair loss that is patchy, non-scarring hair loss of the axillae and pubic area, and follicular papules that can be grouped or diffuse on the trunk and extremities that have a characteristic spinous scale. The scalp aspect of the disease is evident as typical LPP or as patchy alopecia. It is noted that the scalp eruption usually precedes the follicular papule eruption on the body, and this may occur at a much later time than the scalp involvement.

Q. Is FFA associated with other diseases?

FFA has been found to be associated with vitiligo, Sjogrens disease, and vulvar lichen sclerosus et atrophicus. Curiously, it has been noted to occur after surgical procedures such as rhytidectomy, rhinoplasty, and hair transplantation.

Q. Can FFA be found along with FPA?

Yes. At times the diagnosis of FFA may be overlooked because the clinician is more inclined to notice the appearance of female pattern alopecia (FPA).

Q. What is the clinical presentation of FFA?

Typically, the patient is a menopausal female who has noticed increased hair loss particularly in the frontal area. The course of hair loss may be quite slow or be rapid. The patient may complain of pruritus or pain in the affected area. FFA, however, is not just a disease of post-menopausal females. It can occur in pre-menopausal women and it has been seen in men as well.

Evaluation of the patient typically includes a routine medical history and review of systems. On physical examination, the patient may present with diffuse areas of alopecia, finite areas of alopecia, or areas that appear patchy. The clinician should examine the scalp and look for the presence of hair ostia. If the hair ostia are lacking then the possibility of a fibrotic process as is seen in scarring alopecias should be considered. The use of dermoscopy or video microscope may help to discern the loss of hair ostia.

Other clues include erythema, which may be perifollicular and a shiny appearance to the scalp. Perifollicular papules may be present. The scalp tissue may feel firmer to the touch. FFA usually involves the frontal tempero-parietal hairline area. It has a band-like appearance and often seems moth eaten and symmetric. A hair pull test is usually negative. It has also been noted that single isolated terminal hairs can be present at the hairline, which is referred to as the "Lonely Hair Sign".

The clinician should also look for scale, crusting, heme, and scratches. Examination of the facial skin may reveal papules that are often found with FFA. In addition, the clinician should inquire as to pubic hair loss and axillary hair loss. While this may accompany menopause, there seems to be an increased correlation with FFA. Loss of eyebrow hair is a common occurrence with FFA, so the clinician should examine the eyebrows. Additionally, the venous vasculature on the forehead area may be more prominent.

To further elucidate the diagnosis, it is prudent to obtain punch biopsies of the scalp. These should be 4mm in diameter and it is suggested that a specimen be used for horizontal sections as well as one for vertical sectioning. The biopsies should be taken from areas of apparent active disease. A review by a dermatopathologist is also suggested. The dermatopathologist may advise use of elastin stain and direct immunofluorescence to aid in confirming the diagnosis.

Additional lab work may be needed, particularly when the patient is female. These studies may include TIBC, Ferritin, TSH, T4, and hormonal assays.

Q. Can FFA go beyond the frontal scalp?

Yes. FFA can progress and extend to the crown area. In men, cases of FFA involving the sideburns have been reported.

Q. Is there a genetic basis to FFA?

There may be a spectrum of disease and there is evidence to suggest that in some instances there is a familial occurrence.

Q. Is there a correlation between pruritus/pain and nerve damage?

One study has shown that there is a decrease in epidermal nerve fiber density in FFA (Hordinsky, M., et al. Small fiber neuropathy in symptomatic lichen planopilaris and frontal fibrosing alopecia. *J of Investigative Dermatol.* 2013; 133(5):1403). The pain that patients experience may be related to loss of epidermal nerve fibers.

Q. What is the differential diagnosis for FFA?

The differential diagnosis includes central centrifugal cicatricial alopecia, Lupus erythematosus, and en coup de sabre, a form of localized scleroderma. Also included are frontotemporal androgenetic alopecia, which lacks an inflammatory component, traction alopecia, and ophiasis (a form of alopecia areata that has a crown-like distribution).

Q. What medical therapies are available for FFA?

At present, there does not appear to be any particular therapy that is proven to work consistently for FFA. It is noted that a common approach is to use a topical steroid with another modality. Some regimens for treatment include the following: finasteride; dutasteride; tacrolimus, topical steroids, minoxidil, methotrexate, hydroxychloroquin, minocycline and tacrolimus, cyclosporine, retinoids such as acitretin and isotretinoin, griseofulvin, oral prednisolone, mycophenolate mofetil, azathioprine, lymecycline, and gabapentin and clonidine for pain and pruritus.

At times, combinations of medications, such as dutasteride plus minoxidil, can be helpful. Another combination that has been reported to have some success is minoxidil and tacrolimus.

Q. If medications such as finasteride and dutasteride can help to treat FFA, is there a hormonal basis to the disease?

The fact that drugs such as finasteride and dutasteride can sometimes help in the treatment of FFA suggests that DHT may play a role in this condition. The evidence, however, is unclear. There may also be regional factors that may be important. Additionally, the skin in the forehead area may produce substances that contribute to the disease. Evidence for this is based on transplanting hairs into an affected area. The hairs may initially grow but usually do not survive for a long period of time.

Q. Can one tell if FFA is "burned out"?

It is difficult to ascertain if in fact FFA has subsided. Certainly, if one observes any areas of erythema not associated with another cause, the disease is probably still ongoing. If inflammation is not clinically evident, obtaining biopsies may be helpful; however, biopsies are only small samples of a larger region, and even if the histology does not indicate an active process, the disease may be present just a few millimeters away from the biopsy site(s). If there is no evidence of inflammation, no associated symptoms of pruritus or pain, and the area of alopecia has not extended, then the patient may consider the process stabilized.

Q. If FFA is felt to be "burned out," does this mean that the affected area can be transplanted?

The question requires a case-by-case approach. Most hair restoration physicians would be hesitant to perform the procedure even if the FFA seemed stable. In cases where transplants may be considered, the surgeon will often have waited one or two years after such "stabilization" before performing the surgery.

Sometimes, even if a hair transplant seems to do well initially, over time the hairs in the region can undergo destruction as the disease is reactivated. If considering transplanting in FFA, the patient should be appropriately informed as to the risks, and we suggest that a patient be put on maintenance therapy to try to avoid a recurrence of the inflammatory disease.

If undertaking a transplant procedure, it is a good idea to keep the cases small in terms of number of grafts. A session of 500 or so grafts at a time might allow the physician to see how the grafts will take without jeopardizing a large number of valuable donor hairs.

Q. Are there specific clues for diagnosis of FFA in males?

FFA is rarely observed in males. It may be that it is at times overlooked. To avoid missing the diagnosis, the clinician should be alert to symmetric hair loss in the frontal area and sideburns accompanied by a change in appearance of the skin involved. An important clue is loss of hair ostia. This is characteristically noted in other forms of cicatricial alopecia as well. Upon noting what appears to be loss of ostia, it is prudent to look with loupes or video microscope to confirm this finding. If there is loss of ostia, we would suggest biopsies of the involved areas to ascertain the diagnosis and determine what is occurring histologically. Similarly, erythema unrelated to clearly diagnosed seborrheic dermatitis, folliculitis, psoriasis, or other non-fibrosing disorders should prompt the physician to obtain biopsies to determine the cause of the inflammation.

Q. Should an area be biopsied if no inflammation is evident?

Even when there is no inflammation apparent to the eye, it is our opinion that biopsies should be obtained, particularly if the goal is to be able to consider transplanting the area affected. While having biopsies that show no significant inflammation does not absolutely assure the patient and clinician that inflammation is not occurring in the adjacent or distant areas, it may give some measure of "comfort" in pursuing the surgical course.

An important sign to note in the course of FFA is the distance of recession of the hairline from predetermined landmarks. While there may not be inflammation that is observed, if there are measurements of the height of the hairline from previous visits, the clinician can assess whether the disease is progressing by measuring the height of the hairline in successive visits. If the hairline has moved back, the disease is still active.

Q. Why is one or two years of quiescence of FFA considered an adequate period of time to wait before proceeding with surgical intervention?

There does not seem to be data to suggest that the one- or two-year period qualifies as a period of remission to allow surgery. The one- or two-year time delay seems to be an arbitrary one that clinicians have felt comfortable with. The lack of positive anecdotal reports from HT surgeons who have treated FFA seems to call into question such a consensus approach.

⇨ page 172

FFA

from page 171

Q. *If a one- or two-year period is accepted as the time before transplanting an apparently burned out FFA, what is the start date of the time period?*

Our personal opinion is that the start time is the day the patient is seen in our consultation with what seems to be burned out FFA. If the patient has been followed by an outside dermatologist who indicates that he or she feels that the patient's FFA has been in remission for a significant time, we will take that into consideration but will still re-evaluate the patient over a minimum of a one- to two-year span before possibly agreeing to perform a hair replacement procedure. In the case of FFA, it may be prudent to wait three or even five years before attempting a transplant as its course is insidious and active inflammation can be difficult to discern clinically.

Q. *In view of the case presented by Dr. Knudsen in the last issue of the Forum (2013; 23(4):117), what is the role of post procedure maintenance therapy?*

We believe that long-term therapy for FFA is prudent. At the minimum, we would use a mid potency topical steroid solution, topical immune modulator, minoxidil, and probably an alpha reductase blocker if the patient is a post-menopausal female or if the patient is male. We would consider using these medications even if no inflammation was visible.

Conclusion

FFA is a relatively common scalp disorder. Most hair restoration surgeons will inevitably come across patients with this malady. It is important to be able to recognize this disease entity and be able to provide accurate information to the patient, possible treatment options, and ways to treat the symptoms of pruritus and pain in the scalp.

If the surgeon is going to undertake transplantation in these patients, it is crucial to discuss the risks involved with the patient, particularly the possible reactivation of the disease and loss of transplanted hair.◆