ASSOCIATION OF FRONTAL FIBROSING ALOPECIA AND TRIPLE X KARYOTYPE

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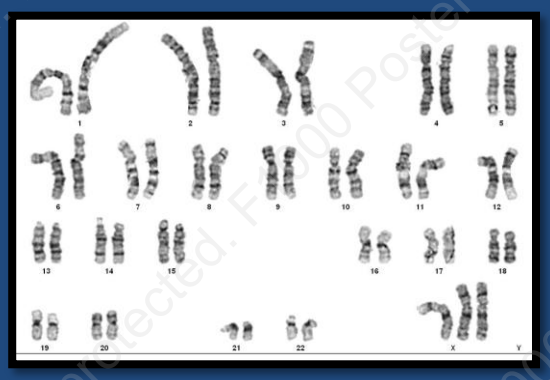
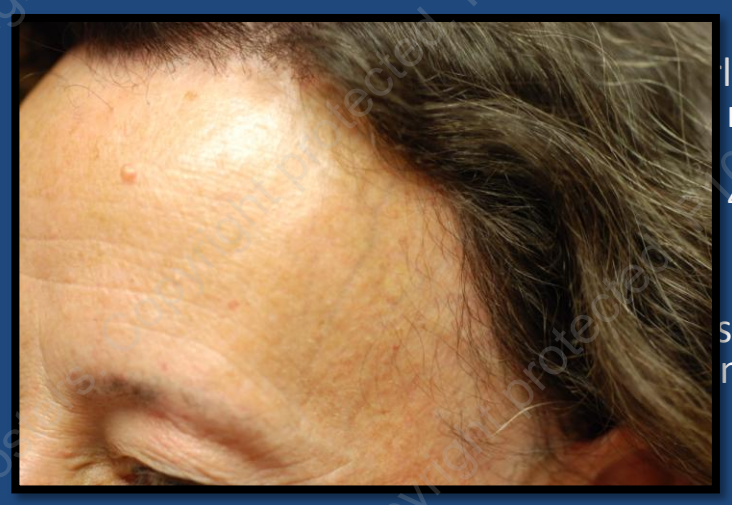
Frontal Fibrosing Alopecia (FFA) , first described by Kossard in 1994[1], is a primary lymphocytic cicatricial alopecia characterized by a progressive frontotemporal hairline recession with in some cases, a progressive loss of eyebrows . The disease affects most often postmenopausal women but can also be observed in men and premenopausal women.
We report the case of FFA associates with a 47 XXX karyotype.



CASE REPORT



A 57-year-old woman visited us in June 2010 with a frontotemporal
line recession and a scalp atrophy appearance. Personnal
revealed fertility problems, linked to an early menopause,
1990, which highlighted a triple X syndrome in its mosaic
47 XXX).

The patient has been treated for many years for hypothyroidism in the context of autoimmune thyroiditis. During the same period, a
stitutive hormonal therapy (natural estrogen and progestative) has
n taken.

DISCUSSION

To date, no assocation FFA/XXX has been reported to our knowledge. FFA occurs most of the time in menopause (mean age : 63.8 in one series[2]) . Similary to one of the cases reported by Tan and al[3], our patient presented a premature menopause, which explains the relatively early onset of the FFA. These observations effectively suggest that hormonal changes linked to menopause, contribute to the occurance of the disease. Clinical signs of triple X are rather discreet, particulary in cases of mosaicism. In these cases, a majority goes undiagnosed. In other cases, one can observe a tall stature, psychiatric disorders, infertility, academic difficulties or delayed motor and speech development[4]. The incidence of the syndrome is 1 in 1000 females. More interesting is the existence of the association triple X and autoimmune diseases (Sjögren syndrome[5], and thyroiditis[6]). Moreover these conditions have been described in association with FFA which is also considered to be an autoimmune reaction. Coexistence of autoimmune processes with triple X syndrome points to the role of the X chromosome in the maintenance of immunocompetence. This chromosome has been reported to provide a survival advantage in the case of pathogenic conditions but thus can also enhance the female susceptibility to auto-immunity[7]. Association triple X and FFA, sustains the hypothesis that hormonal changes may have an inducting role in the genesis of this cicatricial alopecia. Furthermore, these observations give us insight into a possible link between hormonal climate and immunity. Therefore, a karyotype should be carried out in FFA cases with clinical signs evocating a triple X syndrome.

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