

Familial presenile sebaceous gland hyperplasia

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Abstract:

Familial presenile sebaceous gland hyperplasia is a rare autosomal dominant disease. Only few cases were reported in the literature ⁽¹⁻⁶⁾. Here in we are reporting an additional case.

Sebaceous gland hyperplasia is a common disorder in middle and old age males. Familial presenile sebaceous gland hyperplasia is a rare autosomal disease, which is first described by Dupre Bonafe and Lamon in 1980 ⁽¹⁾. Only few cases reported in the literature ⁽¹⁻⁶⁾. We are reporting an additional case of such a rare disease.

Case Report :

37 years Egyptian female presented with multiple asymptomatic skin lesions of the face which started at the age of 25 years and were progressively increasing in size and number and associated with excessive oily face. History of exacerbation and flare during each pregnancy. Patient gave history of treatment with multiple courses of tetracycline with no improvement.

Similar history was noted in her 80 years old father who had similar skin lesions started at the age of 30 years, the rest of her brothers and sisters were normal. Her blood group is A +ve.

Examination revealed innumerable 1-2 mm papules with umbilication at the center located on fore-

head, nose, cheeks, neck and upper chest. Some of the lesions were confluent forming plaques and sparing the periobital and perioral areas. Multiple deep furrows were seen on the cheeks and forehead. No acne lesions were seen (Fig.1, 2).

Biopsy of the skin showed sebaceous gland hyperplasia (Fig.3). Blood investigations including CBC, Cholesterol, Triglycerides and serum testosterone all were within normal limits.

Patient was started on Isotretinoin 1mg per kg (40 mg twice daily) for total period of 20 weeks.

Most lesions improved by the 4th week and almost 90% improvement at the end of the course (Fig.4). Investigations including complete blood count, liver enzymes serum cholesterol and triglycerides were done before, during and after the courses and all were within normal limits. The patient was re-examined at six months, one year and two-year post treatments and showed no relapse.

Discussion:

Familial presenile sebaceous gland hyperplasia is a recently described syndrome. It was first described in 1980 by Dupre et al ⁽¹⁾. Few reported cases had been described there after ⁽²⁻⁶⁾. The Dupre

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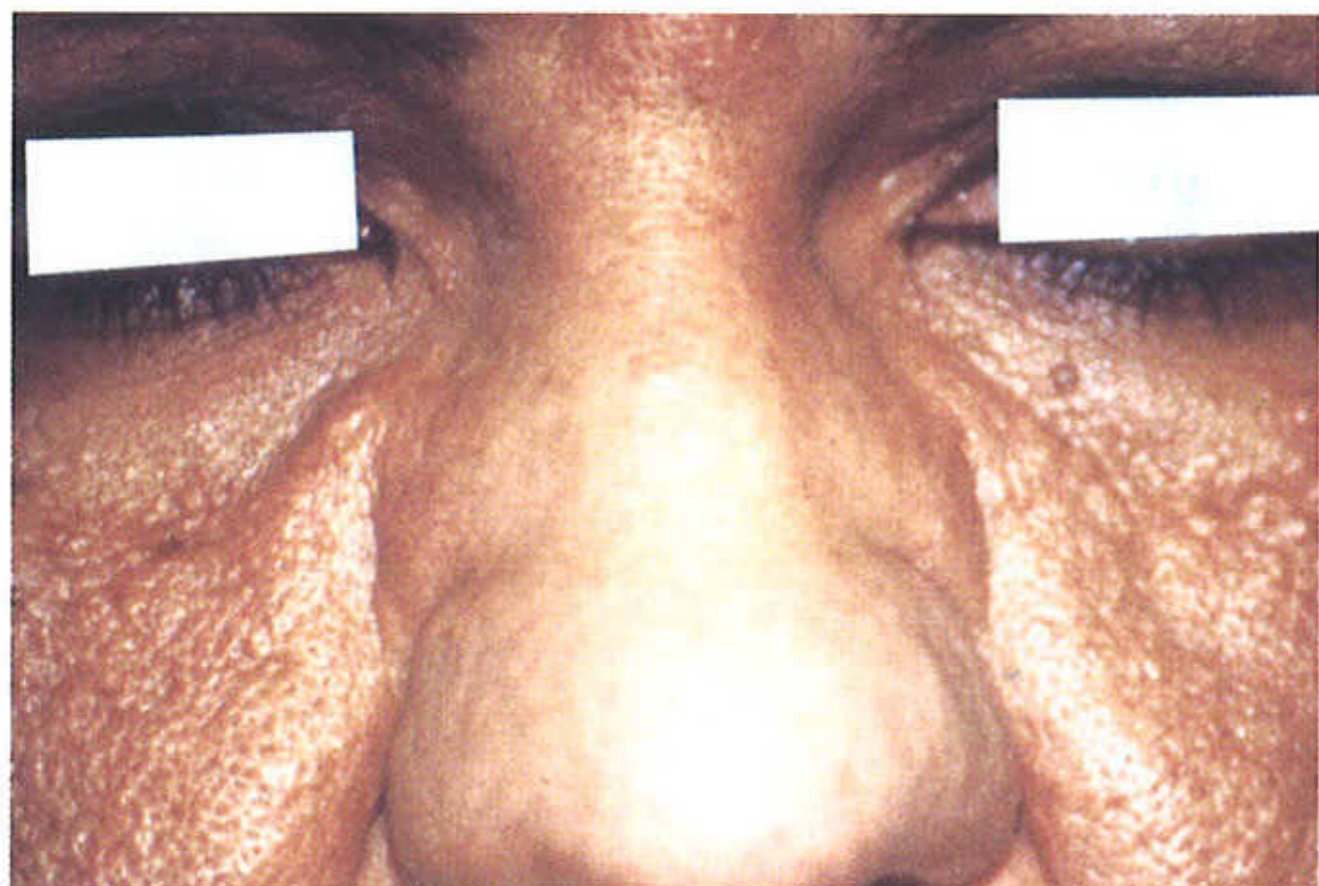
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Fig. (1)
 Patient before treatment showing multiple yellow papules.



*Fig (2) [a , b]
Closer view showing the umbilicated yellow papules.*

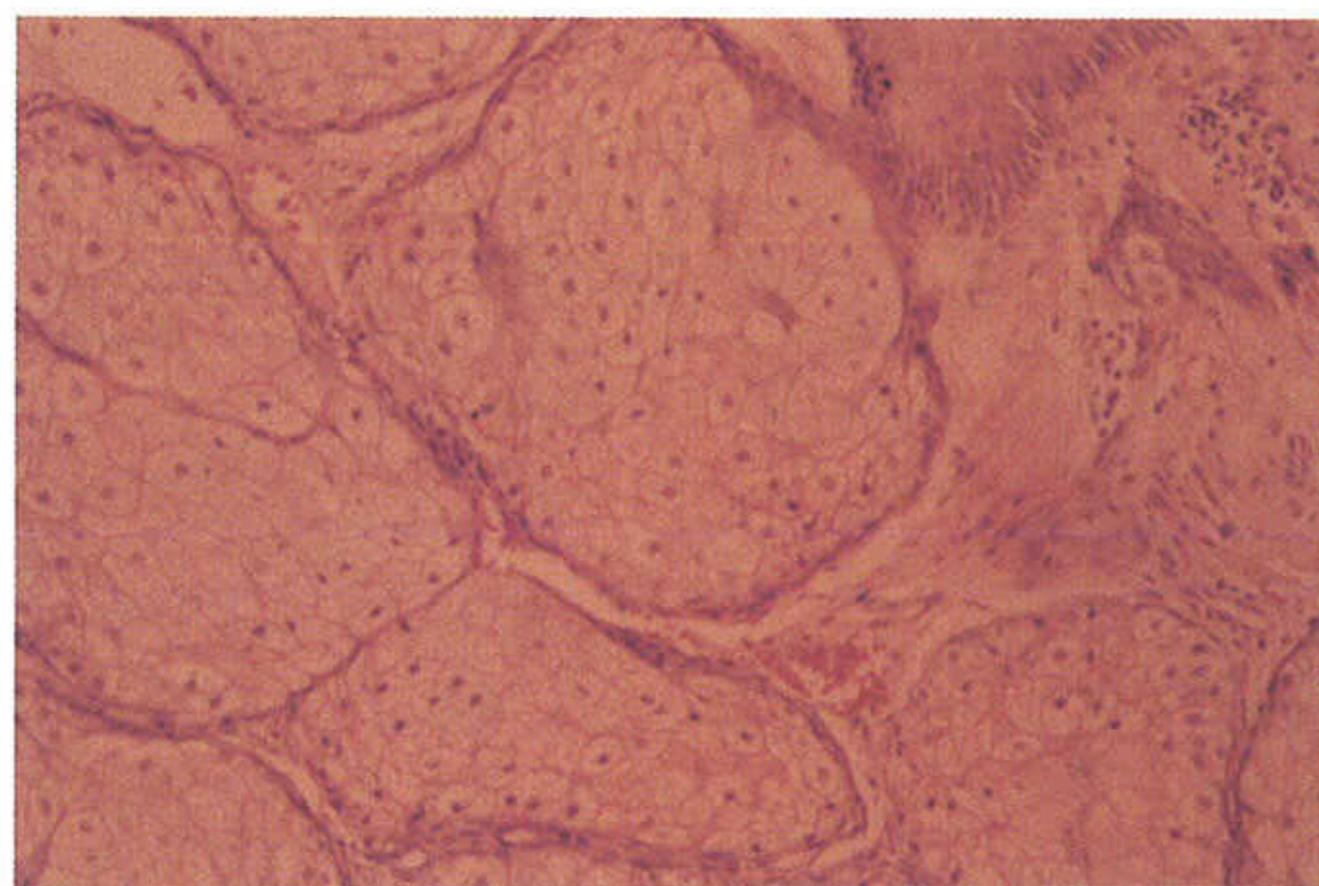
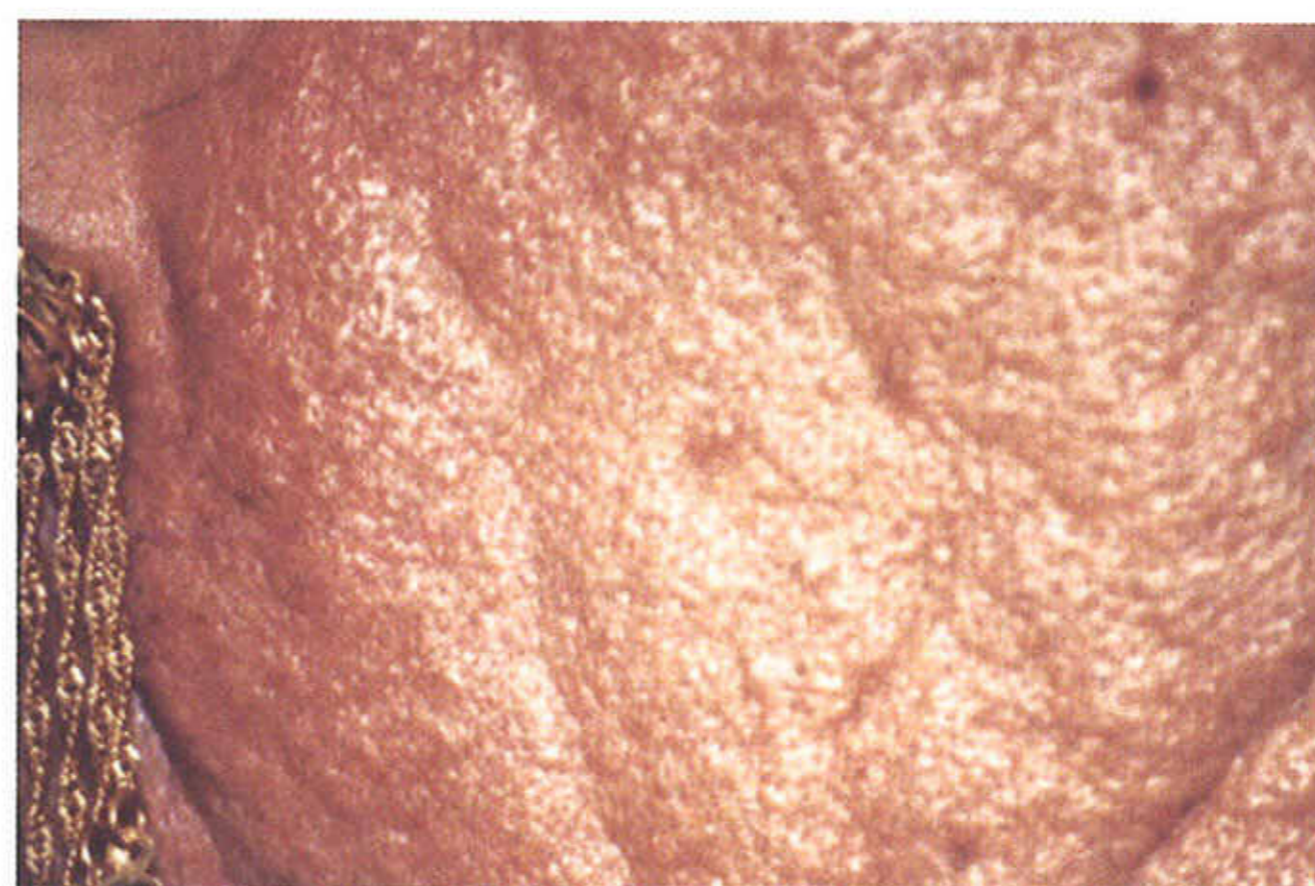


Fig. (3) Histopathology showing lobules of hypertrophic sebaceous glands around a central dilated duct (hematoxyline & Eosin original magnification x 86)



*Fig (2) [c]
Closer view showing the umbilicated yellow papules.*



Fig. (4) Patient post treatment showing marked improvement (Frontal view).

criteria for this rare entity includes:

- (1) confluence of lesions resulting in the formation of large plaques on the face but sparing of the periobital and perioral regions.
- (2) Highly functional glandular hyperplasia resulting in excessive sebaceous secretion
- (3) Absence of acne,
- (4) Lesions on the face but also on the neck and upper chest with a linear pattern
- (5) Unresponsiveness to conventional acne treatment
- (6) Histopathologic features similar to those of senile sebaceous hyperplasia
- (7) Appearance during puberty or just afterwards and a slow progressive nature^(1,6).

Our case fulfills the criteria of Dupre and showed marked improvement with isotretinoin.

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