in patients with Parkinson disease treated with this drug, but there is no evidence that its use increases the risk of melanoma or its progression.¹

In conclusion, we would like to draw attention to the importance of using a high level of sun protection and of referring these patients to a dermatologist if any pigmented lesion develops.

Conflicts of Interest

The authors declare no conflicts of interest.

References


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Unilateral Focal Dermal Hypoplasia

Hipoplasia dérmica focal unilateral

To the Editor:

Focal dermal hypoplasia (FDH) is a rare genodermatosis characterized by specific skin manifestations such as hyperpigmentation and hypopigmentation, atrophy and telangiectasia with cribriform or linear distribution, following the Blaschko lines.

The disease is X-linked dominant due to the predominance in women and the frequent history of aborted and stillborn male fetuses. It is thought that the few cases reported in men are due to somatic mosaicism, mutations in one sister chromatid, or new mutations.¹

In the initial stages of embryogenesis, 1 of the 2 X chromosomes in each somatic cell becomes inactivated and forms the sex chromatin (lyonization). This phenomenon is random and permanent and gives rise to 2 functionally different cell populations (functional mosaicism).²

In X-linked dominant diseases, affected women present different clinical manifestations because lyonization may give rise to 3 patterns of functional mosaicism: following the Blaschko lines, in a lateralization pattern, or in a checkerboard pattern.³

FDH is an X-linked dominant disease that is fatal in males and in which lyonization usually manifests along the Blaschko lines.

We report the case of a neonate with skin manifestations limited to one side of the body, who 2 years later, developed minimal lesions on the contralateral side.

Our case is probably due to the lateralization pattern of lyonization. We found only 3 similar cases of unilateral FDH in a review of the literature.⁴⁻⁶

We describe the case of a 20-month-old girl who, since birth, had presented atrophic scarring, with telangiectasia, hyperpigmentation, and hypopigmentation, affecting the left axilla, the left side of the torso, and the lower left limb, following the Blaschko lines (Figure 1). The patient presented a cribriform atrophic plaque on the left side of the nasal pyramid (Figure 2).

There was no relevant family history and the mother had no history of abortion.

Figure 1 Atrophic linear lesions following the Blaschko lines, affecting the left side of the body.
Since birth, the patient had presented bilateral obstruction of the tear ducts, which had been treated with dacryocystectomy. She suffered no musculoskeletal disorders, visceral involvement, or abnormal psychomotor development. Skin biopsy of the atrophic linear lesions revealed a normal epidermis with adipose tissue in the upper dermis, which was slightly atrophic (Figure 3).

Ultrasound scans of the brain and abdomen and karyotype analysis revealed no abnormalities.

FDH was diagnosed based on clinical and histologic characteristics.

After 2 years of follow-up, the patient developed minimal linear cribriform lesions on the right arm.

FDH or Goltz syndrome is a rare genetic disease that affects tissues derived from ectoderm and mesoderm germ cells.

Several mutations have recently been reported in the PORCN gene; this gene is located on chromosome Xp11.23 and codes for an enzyme that is essential for the normal development of embryonic tissues.7

Skin manifestations, particularly poikilodermatous lesions, that follow the Blaschko lines are the most common findings. Another frequent finding is fibroepithelial papilloma, which usually affects the perianal, vulvar, and perineal regions but may also affect the oral mucosa and respiratory tract, and the fatty hernias, which present in more than half of individuals and are due to adipose tissue in the dermis.

The nails may be absent or present atrophy or dystrophy. The hair is usually sparse and fragile and 30% of patients develop alopecia.

Extracutaneous manifestations vary widely and include musculoskeletal abnormalities (syndactyly, missing digits, scoliosis, and corporal asymmetry), facial dysmorphia (facial asymmetry, prominent chin, and narrow nasal bridge), oral abnormalities (enamel defects, dysplastic teeth, and missing teeth), and abnormalities of the gastrointestinal and urinary tracts.

Most patients with FDH present longitudinal striations in the epiphysis of the long bones (striated osteopathy), which is considered a distinctive sign of this disease. Some patients present osteoporosis, fibrous dysplasia, and cystic lesions of the bones.8-10

The most common ocular findings include coloboma, microphthalmia, strabismus, nystagmus, and subluxation of the lens; other findings that are not uncommon include defects of the lachrymal apparatus, such as chronic inflammation and tear duct cysts.11

It is not rare for patients with FDH to present differing degrees of mental retardation.

Most cases have a normal karyotype and are sporadic, as occurred with our case.

Although this girl did not suffer visceral involvement, the skin manifestations were sufficiently characteristic to allow a diagnosis of FDH to be made.

We believe that this case is unusual because the patient presented intense skin lesions that were initially restricted to one side of the body; this may be explained by a phenomenon of selective inactivation of the X chromosome or by postzygotic mutation.

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References


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Atrophic Lichen Planus Annularis: Presentation of 3 Cases

Liquen plano anular y atrófico: presentación de 3 casos

To the Editor:

Atrophic lichen planus annularis is a variant of lichen planus that is rarely reported in the literature. It is characterized clinically by annular papules with elevated margins and a central atrophic area; these lesions may be associated with the typical lesions of lichen planus. Histologically, the borders of the lesions present the typical findings of lichen planus, together with progressive thinning of the epidermis, and the center reveals reduction and fragmentation of the elastic fibres of the dermis.

Due to the rare nature of the disease, it has not been possible to establish a typical location for the lesions.

We report 3 cases of patients with atrophic lichen planus annularis with no other accompanying lesions; the lesions were located in the lumbar region and showed similar responses to the prescribed treatment.

The first case was a 47-year-old man with no relevant personal history. During a checkup for melanocytic nevus using epiluminescence, several skin lesions were detected that had been absent in previous annual checkups.

The second case was a 50-year-old woman with a history of diabetes mellitus on treatment with insulin, hypertension, and hysterectomy due to endometrial cancer; the patient was seen at our department for assessment of lesions that had appeared 3 months earlier (Figure 1A).

The third case was a 15-year-old boy with no relevant medical or surgical history, who visited our department for assessment of skin lesions that had been present for an unknown length of time.

In all 3 cases, the lesions were asymptomatic and located in the lumbar region in the midline. There was no characteristic grouping pattern of the lesions, except in the third case, in which they had a linear distribution. The lesions consisted of circular papules with a raised violaceous margin, which was slightly scaly in the second case, and an atrophic central area.

No lesions were found elsewhere on the skin or on the mucosa or nails.

Additional examinations included routine laboratory tests in the first case; these tests showed no abnormalities in the