Darier Disease
Darier's disease is usually transmitted in an autosomal dominant pattern. It has been found to be due to
The oral mucosa is involved occasionally. In some cases of Darier's disease, keratotic papules that resemble those seen in acrokeratosis verruciformis of Hopf are found on the dorsa of the hands and feet.

Special clinical variants of keratosis follicularis and Darier's disease include:

- A hypertrophic type
- A vesiculobullous type
- A linear or fusiform type, usually limited to one side, with localized or widespread lesions that may be present at birth but in most cases have arisen in infancy, childhood, or adult life.

The question has been raised as to whether this type of lesion represents a linear epidermal nevus with acantholytic dyskeratosis rather than Darier's disease, and the designation acantholytic dyskeratotic epidermal nevus has been suggested. It is quite likely that some cases with scattered papular lesions of limited extent arising in adult life and diagnosed as acute, eruptive Darier's disease or acute adult-onset Darier-like dermatosis in reality represented transient acantholytic dermatosis. A recent review on the clinical aspects of this condition has been written.
Histopathology
The characteristic changes in Darier's disease include the following: (a) a peculiar...
The corps ronds occur in the upper stratum malpighii, particularly in the granular and horny layers; grains
pyknotic nucleus that is surrounded by a clear halo. By virtue of size and the conspicuous halo, corps ronds
Hyperkeratosis and papilomatosis may cause the formation of keratotic plugs, which often fill the pilosebaceous follicles. That Darier's disease is not exclusively a follicular disorder is also proved by the fact...
that areas devoid of follicles, such as palms, soles, and the oral mucosa, may be affected.

In hypertrophic lesions of Darier's disease, considerable acanthosis may occasionally be observed, either...

The vesiculobullous lesions, which occur in rare instances, differ from lacunae merely in size; they conta...
The keratotic papules that may occur on the dorsa of the hands and feet and that clinically resemble those seen in Darier's disease are a manifestation of the disease and not of acrokeratosis verruciformis. They are often supra basal clefts as well.

The lesions on the oral mucosa are analogous in appearance to those observed on the skin and thus show lacunae and dyskeratosis, although definite well-formed corps ronds generally are absent.
The occasional reports of patients having both Darier's disease and familial benign pemphigus or a transition from one of these two diseases to the other are discussed in the differential diagnosis of familial benign pemphigus.

Pathogenesis. Whereas histologically a distinction between Darier's disease and familial benign pemphigus is generally possible, with Darier's disease often showing characteristic intracellular acantholysis and dyskeratosis in others and only rarely showing both. In both diseases, however, acantholysis precedes dyskeratosis.
Acantholysis has been thought by some authors to be due to the loss of the intercellular contact layer within desmosomes, both in Darier's disease and in familial benign pemphigus. The two halves of the desmosomes then pull apart, after which the tonofilaments become detached from the attachment plaques and thus to acantholysis. It is likely that both processes take place simultaneously in both Darier's disease and familial benign pemphigus.
The cause for the acantholysis in Darier's disease and familial benign pemphigus is not yet definitely known. It is believed to be related to a mutation in the SERCA2 pump in the calcium signaling pathway that regulates cell to cell adhesion and differentiation of the epidermis.

In association with the loss of desmosomes, excessive amounts of tonofilaments form within the keratinocytes.
association with large keratohyaline granules, form large aggregates of homogenized dyskeratotic material.
In familial benign pemphigus, too, after loss of the desmosomes, excessive amounts of tonofilaments form...
On intralesional injection of tritiated thymidine, one group observed labeling of many acantholytic keratinocytes in familial benign pemphigus but not in Darier's disease, suggesting that in familial benign pemphigus, the epidermal cells participated in the renewal of the epidermis but did not do so in Darier's disease, probably because these cells were undergoing keratinization. However, another group could not confirm this observation.

Differential Diagnosis
Although acantholytic dyskeratosis in association with corps ronds is highly characteristic of Darier's disease, it is not always present. Occasionally, a few corps ronds are seen also in familial benign pemphigus.