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## Firm Yellowish Nodule on the Face of a 7-year-Old Child: What is Your Diagnosis?

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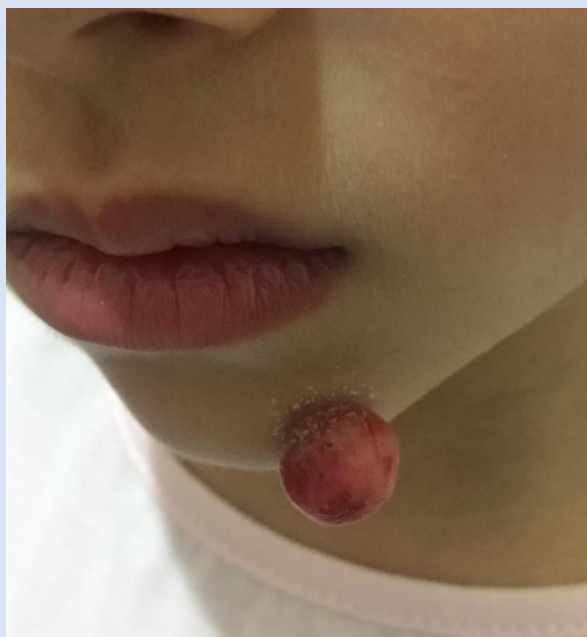
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Received Date: 18-04-2020; Accepted Date: 24-05-2020; Published Date: 31-05-2020

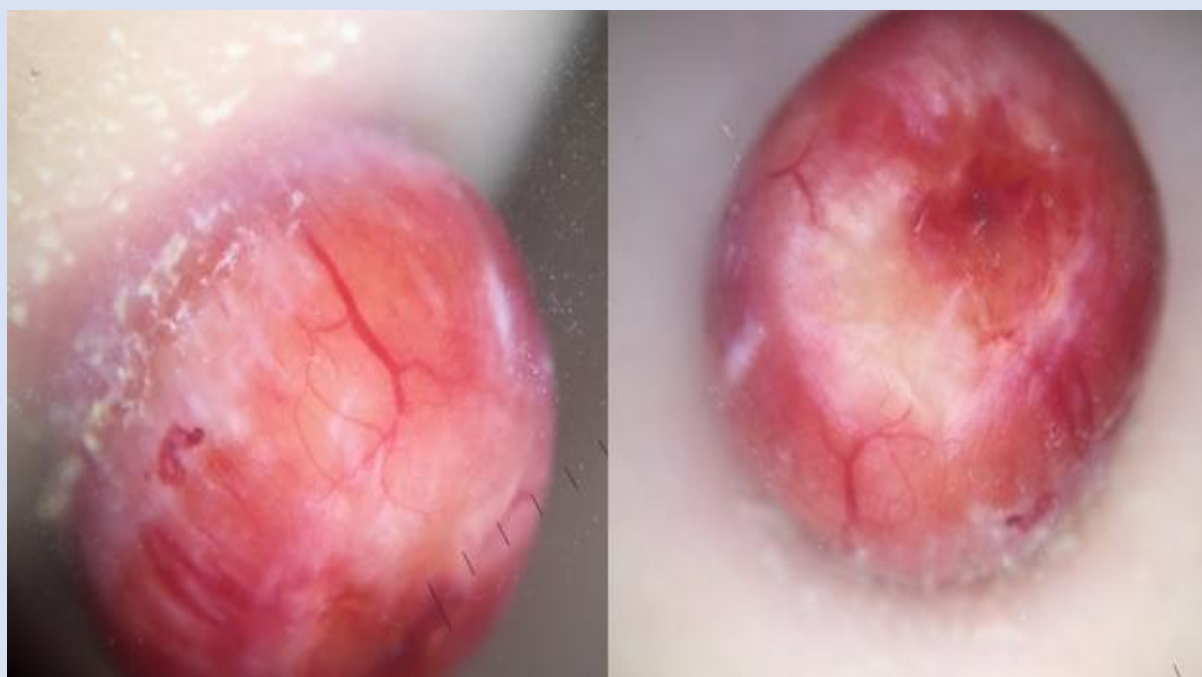
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### Introduction

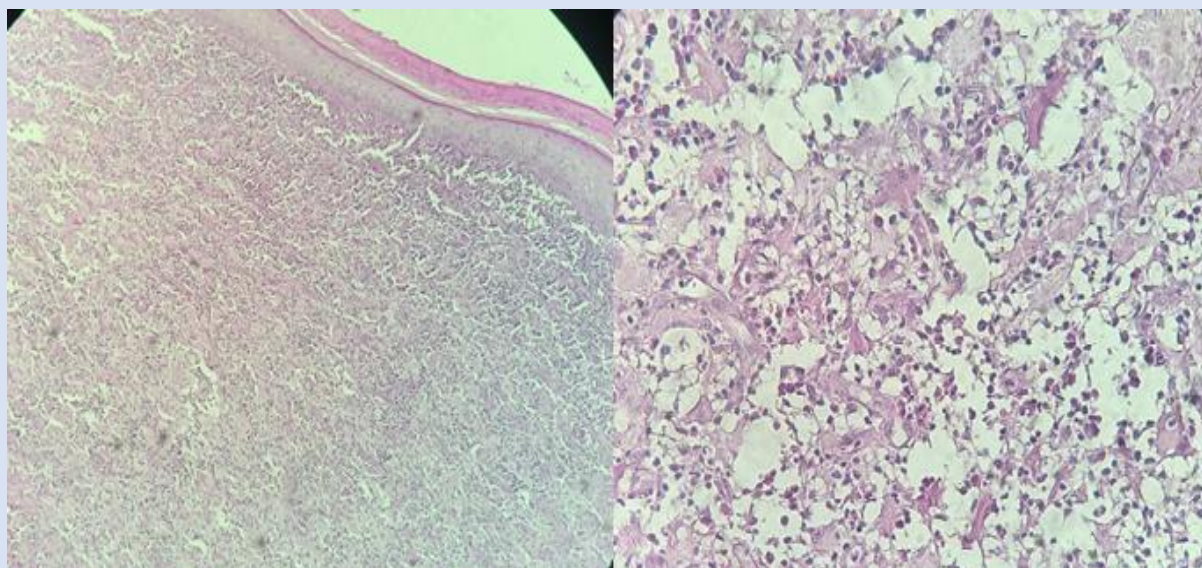
A 7 year old girl presented at our clinic with a 3 months history of cutaneous nodule on her face, which had gradually increased in size, non painless and pruritic. The dermatological examination revealed a 2 cm size firm erythematous nodule, yellowish in the center, on the mental region. There was no mucosal involvement. The general physical examination was normal, with no signs of lymph node enlargement. Dermoscopy showed yellowish homogeneous areas in the center of the lesion, whereas, in the outer part, was a milky-red area, traversed by large telangiectatic vessels on a red-orange background. Histology revealed a normal epidermis, in the dermis, a nappe of cellular populations, made up of large, macrophagic-looking cells, with image of phagocytosis of lymphoid cells, was noted. The rest of the dermis showed dense inflammatory infiltrates, composed of neutrophils, plasma cells, lymphocytes and histiocytes. A phenomenon of emperipolesis was clearly defined. Immunohistochemical staining revealed that the histiocytes were strongly positive for S100 protein, weakly positive for CD68 and negative for CD1a. Cell blood count, liver and kidney function test, chest x-ray and abdominal ultrasonography were all normal (Fig. 1-3).



**Figure 1:** Firm erythematosis nodule, yellowish in the center, of 2cm size, on the mental region.



**Figure 2:** Yellowish homogeneous areas in the center of the lesion, whereas, in the outer part, was a milky-red area, traversed by large telangiectatic vessels on a red-orange background.



**Figure 3:** A normal epidermis, in the dermis, a nappe of cellular populations, made up of large, macrophagic-looking cells, with image of phagocytosis of lymphoid cells, was noted.

The rest of the dermis showed dense inflammatory infiltrates, composed of neutrophils, plasma cells, lymphocytes and histiocytes. A phenomenon of emperipolesis clearly defined.

## What is Your Diagnosis?

Diagnosis: Purely cutaneous Rosai-Dorfman disease.

Rosai-Dorfman disease is a non-langerhans cell histiocytic proliferative disorder of unknown etiology. It is a rare disease with massive lymphadenopathy. Cutaneous form, which is limited to the skin, is extremely rare. The etiology of CRDD remains unknown with viral and immune causes hypothesized [1]. The clinical features of cutaneous Rosai-Dorfman disease are heterogeneous and can be just a single or, more commonly, multiple, papules, nodules or indurated plaques, of different sizes with no anatomical predilection localization [2]. In dermoscopy, milky-white ovoid structures with a cotton-like appearance over a light-red background, or yellow homogeneous areas surrounded by large telangiectatic vessels on a red-orange background, can be observed [3]. The histological examination is the key of diagnosis, in which we find, typically, a normal epidermis, but in the dermis, we find a diffuse infiltrate of histiocytes, accompanied by a background infiltrate of lymphocytes and plasma cells. The phenomenon of emperipolesis, which represents the presence of intact lymphocytes within histiocytes, is frequently observed. Rosai-Dorfman disease stains positively for S100 protein and CD68, but negatively for CD1a [4]. The treatment of this entity is difficult, surgical excision of the lesions has been helpful in certain cases, which is the case for our patient. Others options, as cryotherapy or local radiation or medical treatment as corticoide, dapson, are

thalidomide, isotretinoin or imatinib have also been proposed with good improvement of a number of patients, while the response in others patients remained poor [5].

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