

Facial Tumors Revealing Systemic Sarcoidosis: About a Case Report

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Received date: 08 Oct, 2023 |

Accepted date: 18 Oct, 2023 |

Published date: 23 Oct, 2023

Citation: Jahouh A, Barakat L, Echchilali K, Moudatir M and Kabli HE. (2023) Facial Tumors Revealing Systemic Sarcoidosis: About a Case Report. J Case Rep Med Hist 3(7): doi <https://doi.org/10.54289/JCRMH2300134>

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Abstract

Cutaneous involvement in sarcoidosis is highly variable, occurring in 9 to 37% of cases, and can be classified into specific and non-specific lesions. The specific lesions are essentially sarcoids with small and large nodules, plaques and annular lesions. However, the tumoral form is still very rare, and has only been reported in a handful of cases in the literature.

We report the case of a 33-year-old female patient diagnosed with systemic sarcoidosis, revealed by a 5-year history of atypical skin lesions on the face, the largest of which measured 7 cm long. Skin biopsy was consistent with sarcoidotic granuloma, and workup for other localizations revealed ocular, respiratory, salivary gland and joint involvement.

The originality of our observation lies in the extreme rarity of the tumoral form in cutaneous sarcoidosis.

Keywords: Cutaneous Sarcoidosis, Tumoral Lesions, Sarcoidotic Granuloma

Introduction

Sarcoidosis is a multisystem disease of unknown cause, characterized by the presence of granulomas in various organs [1]. Respiratory and lymph node involvement are the most frequent, noted in over 90% of cases [2]. Cutaneous manifestations are less frequent, reported in 9 to 37% of cases, and can be classified into two categories: specific lesions present histopathological signs of typical sarcoidosis granulomas; non-specific lesions develop following an inflammatory reaction pattern [3]. We report the case of a young female patient with systemic sarcoidosis, revealed by a very rare tumor-like skin lesion.

Case presentation

This is a 33-year-old female patient with a 5-year history of progressively progressive skin involvement, consisting of three facial tumor lesions, purplish-red in color and irregular in contour; the largest lesion measures 7 cm in long axis and is located on the forehead (**Figure 1**). The other lesions are located in the left zygomatic region (**Figure 1**) and the right mandibular region (**Figure 2**). Questioning revealed recurrent episodes of red eye, NYHA stage II dyspnoea, subjective dry mouth syndrome and inflammatory arthralgias without fever or deterioration in general condition. Complementary examinations revealed an elevated conversion enzyme level

of 89 mg/l, hypercalcemia of 110 mg/l, ANCA was negative, ophthalmological examination showed bilateral granulomatous anterior uveitis, and the chest CT scan was consistent with diffuse interstitial lung disease. Histologically, skin, bronchial and accessory salivary gland biopsies revealed chronic granulomatous inflammation of the

sarcoidosis type. The diagnosis of systemic sarcoidosis revealed by tumoral skin lesions was accepted. No ENT, cardiac, neurological or other manifestations were noted in the lesion assessment. Therapeutically, the patient received high-dose oral corticosteroids with tapering off for 2 years, and thalidomide with no improvement in skin involvement.



Figure 1: Tumoral lesion of the forehead measuring 7 cm long axis and lesion of the left zygomatic region measuring 3 cm



Figure 2: Purplish-red skin lesion 4 cm long on the major axis

Discussion

The cutaneous manifestations of sarcoidosis are highly polymorphic, and classically separated into specific, histologically granulomatous cutaneous lesions, and non-specific cutaneous lesions (mainly erythema nodosum) [4]. These lesions are important to recognize, as they may lead to the diagnosis of sarcoidosis, and prompt a lesion assessment to look for visceral localization. Specific lesions are generally infiltrated and painless, rarely involving the epidermis. On vitropressure, they have a distinctive yellowish "quince jelly" or lupoid coloration, with the presence of "candy cane" grains

[4].

These specific manifestations include nodular lesions such as sarcoids with small nodules, sarcoids with large nodules not exceeding 10 to 20 mm [5], dermohypodermal nodules (Darrier-Roussy sarcoids) [6], nodules on old scars [7]; the angiolupoid form [5]; patchy lesions, notably lupus pernio, which is a purplish placard of pasty or hard consistency, localized on the nose and extremities, simulating frostbite [8]; and finally, annular forms, which may suggest centrifugal erythema annulare and granuloma annulare [9].

The cutaneous tumoral form of sarcoidosis is very rare, even



exceptional, with only a handful of cases reported in the literature to date. These include a case of tumoral cutaneous sarcoidosis of the lumbosacral region [10], a cutaneous tumour on the chin measuring 3 cm [11], a case of pseudotumoral sarcoidosis with leonine facies [12], and a case of multiple tumoral and plaque-like lesions in a Chinese patient [13]. Our patient's cutaneous involvement is unusual in view of the tumoral nature of the lesions, as well as their localization and large size. The classic large nodules in cutaneous sarcoidosis usually occur on the extremities and measure less than 2 cm [10].

Conclusion

The cutaneous involvement of sarcoidosis is highly polymorphic, posing a problem of differential diagnosis and making sarcoidosis a great simulator. Classical manifestations are dominated by nodular and plaque-like forms, but tumoral forms are exceptional, hence the originality of our case report.

Declarations

Consent for publication: All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images.

Ethical approval: As international standard, written approval has been collected and preserved by the authors.

Availability of data and material: All data generated or analysed during this study are included in this published article.

Competing interests: Authors have declared that no competing interests exist

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