

## A Rare Association of Primary Orificial Tuberculosis with Cutis Verticis Gyrata

Ahmed Anas Zaougui<sup>1</sup>, Ouiame EL Jouari<sup>2\*</sup>, Rime Dassouly<sup>2</sup>, Ghita Senhaji<sup>2</sup>, Sara Elloudi<sup>2</sup>, Zakia Douhi<sup>2</sup>, Hanane Baybay<sup>2</sup>, Fatima Zahra Mernissi<sup>2</sup> and Moulay Hassan Farih<sup>1</sup>

<sup>1</sup>Department of Dermatology, University Hospital Hassan II Fez, Morocco

<sup>2</sup>Department of Urology, University Hospital Hassan II Fez, Morocco

\*Corresponding Author: Ouiame EL Jouari, Department of Urology, University Hospital Hassan II Fez, Morocco.

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### Abstract

Cutaneous tuberculosis is an uncommon form of extrapulmonary tuberculosis. Orificial tuberculosis comprise 0.1 - 1% of all clinical presentations of tuberculosis. Although, primary oral tuberculosis is extremely rare and more likely to occur in younger rather than older adults.

We present a particular case of primary orificial tuberculosis, revealed by a macrocheilia, occurring in very rarely reported immunocompetent adult. Moreover, we highlight an uncommon association with cutis verticis gyrata.

**Keywords:** Orifice; Tuberculosis; Scalp; Cerebral Gyri; PCR

### Introduction

Primary oral tuberculosis is extremely rare and more likely to occur in younger rather than older adults [1]. It is estimated that only 0.05% to 5% of total tuberculosis cases may present with oral manifestations, which may be either primary or, more often, secondary to pulmonary TB [2]. We aim to present a very particular case of primary orificial tuberculosis in an immunocompetent adult, unusually associated to cutis verticis gyrata.

### Case Presentation

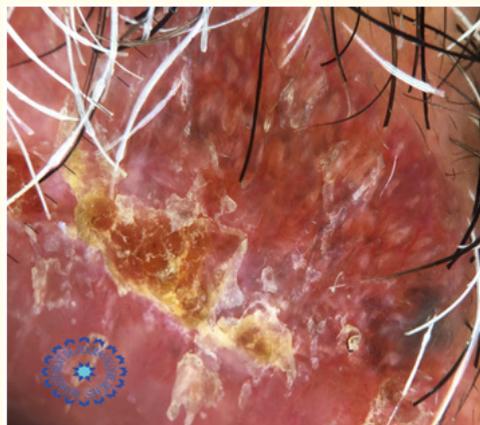
A 58-year-old Moroccan man presented with a 10 years' history of a gradual macrocheilia, complicated by the appearance of several ulcerations of the oral cavity. He also complained of multiple-thickened skin folds over the scalp, which began simultaneously along with the others skin lesions. The clinical examination revealed a pigmented macrocheilia, bilateral fissure pearls and multiple well-defined, perioral ulcerations, the largest of which was 2 cm, firm in consistency with fibrinous background and topped by yellowish and hemorrhagic crusts (Figure 1). Dermoscopy of the lesions showed an erythematous background, yellowish crusts and fusiform yellowish structures without any specific vascularization (Figure 2). A destruction of the nasal septum with a scarring notch of the left nostril, were noted (Figure 3). As well as, a hypertrophic scalp covered with furrows achieving a cerebral gyri appearance (Figure 4). The main differential diagnoses were granulomatosis with polyangiitis, tuberculosis, sarcoidosis and deep mycosis. The craniofacial CT had demonstrated a destruction of the nasal septum with an infiltration of the mucosa of the nasal fossae opposite without indirect signs of meningeal tuberculosis or lymph node involvement. Chest radiography was normal. Tuberculin intradermal reaction was positive. Laboratory tests including immunologic markers and viral serology were negative. A skin biopsy was performed. Histological examination revealed a granulomatous dermatitis suggestive of infectious tuberculosis etiology and no fungal organisms were seen. Finally, the diagnosis of orificial tuberculosis was confirmed by PCR. The patient was started on anti-tuberculous therapy (rifampicin 450 mg, isoniazid 300 mg, pyrazinamide 1000mg, ethambutol 800mg for 2 months and the 2 drugs (rifampicin and isoniazid) were continued for another 4 months. The evolution was favorable with a decline of 4 months.



**Figure 1:** several perioral ulcerations with fibrinous background, topped with yellowish and hemorrhagic crusts.



**Figure 2:** Destruction of the nasal septum with a scarring notch.



**Figure 3:** Dermoscopy showing erythematous background, yellowish crusts and fusiform yellowish structures without any specific vascularization.



**Figure 4:** Scalp is covered with furrows giving a cerebral gyri appearance.

## Discussion

Cutaneous tuberculosis (CT) is an uncommon form (2 - 10%) of extrapulmonary tuberculosis. Orificial tuberculosis (OT) is an extremely rare form of CT, comprising 2% of CT and 0.1-1% of all clinical presentations of tuberculosis [3]. OT may be either primary or, more often, secondary to pulmonary tuberculosis. Primary involvement of the oral cavity is exceedingly rare [2]. The mechanism of primary inoculation into the oral mucous membrane is not definitely established [1]. The intact mucous membrane presents a natural resistance to direct penetration by bacilli [2]. This resistance has been attributed to the thickness of the oral epithelium, the cleansing action of saliva, local pH and antibodies in saliva. Trauma, such as abrasion or erosion, or a persistent chronic inflammation disrupts this natural barrier and provides a route for the organism to penetrate the oral mucosa. Dentures, periodontal diseases, poor oral hygiene and consumption of unpasteurized milk have been proposed to be potential factors [2,3]. OT typically affects young and immunocompromised patients. The diagnosis in our patient was a challenge. He presents a rare primary form of this infection, without internal foci of tuberculosis, occurring in very rarely reported immunocompetent adult. The clinical manifestation of TCO is non-specific, it can present as ulceration, nodules, fissures, diffuse inflammatory lesions, vesicles, tuberculomas or granulomas [1]. Classically, it is characterized by rapidly ulcerating oedematous reddish or yellowish nodules in the oropharynx or the genitoanal tracts. The painful ulcers are frequently circular or irregular with indurated erythematous borders and necrotic bases [4,5]. The course of the disease is acute, possibly leading to extensive non-scarring ulcers, which may persist but occasionally heal spontaneously [5]. The most frequent localization sites were the tongue base and gingiva; lip involvement is very uncommon [3]. Through our observation, we report macrocheilia as a particular and an unusual clinical manifestation of OT. TB can mimic a variety of different conditions, including squamous cell carcinoma, traumatic and aphthous ulcer, syphilis, sarcoidosis, and deep mycotic infections [2]. The gold standard diagnosis of orificial TB relies on the isolation of *M. Tuberculosis* on culture. However, there is low sensitivity for isolating mycobacteria on culture, a process which may take many weeks. Histological examination of a biopsy typically shows a caseating granuloma in the deep dermis and acid-fast bacilli. Smear for demonstration of acid-fast bacilli is recognized as a useful investigation in skin lesions which, typically, have a high bacillary load [6]. The PCR assay for mycobacteria is excellent for detecting genomes of the bacilli [2]. The diagnosis in our patient was based on a constellation of clinical and histological findings. but we were tentative given the absence of an internal focus of tuberculosis which prompted us to carry out a PCR to confirm the diagnosis. Cutis verticis gyrata (CVG) also called “bulldog scalp or corrugated scalp”, is rare a benign proliferation and hypertrophy involving the scalp [7]. It is characterized by thickening of the scalp, which becomes raised to form ridges and furrows resembling the cerebral gyri, which cannot be flattened by traction or pressure [8]. The pathogenesis of this rare entity remains unclear. CVGs are classified into two main groups: primary and secondary. primary CVG refers to cases without obvious origin and with

no other associated abnormalities [7]. Secondary CVG occurs as a consequence of conditions that produce pathological changes in the scalp structure and has been linked to local inflammatory skin conditions (psoriasis, eczema, impetigo, tumors) and to systemic illnesses (amyloidosis, syphilis, myxedema, Ehlers-Danlos syndrome, acanthosis nigricans, and insulin resistance syndrome. The appearance of the skin folds in secondary CVG may be more asymmetrical and can appear at any age [7,8]. The diagnosis of CVG is based on clinical findings. complementary investigations (skin biopsies, blood tests, and radiology examinations) are recommended to rule out local or systemic underlying disorders [7]. To our knowledge, we describe the first case of tuberculosis as an etiology of secondary CVG. The relationship between tuberculosis and CVG is not clear. Although CVG may be disfiguring, the process is essentially benign and no intervention is required. Surgical repair may be implemented if desired, and the type of surgery depends on the size and location of the lesions, the underlying disorder, and the wishes of the patient [8].

### Conclusion

Macrocheilia and painful trailing perioral ulcerations may be suggestive signs of peri-orificial tuberculosis, even in immunocompetent in endemic areas. In patients with secondary cutis verticis gyrate, orificial tuberculosis should be suspected and detailed investigations should be performed.

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