LETTERS TO THE EDITOR

The cutaneous disease of saint ubald of gubbio (1084–1160)

Editor

Ubald was a Catholic Church Saint (1084–1160), born in Gubbio (Umbria, Italy) and died after 2 years of painful illness; his mummy is now guarded in a glass sarcophagus at Sant'Ubald Cathedral in Gubbio. Informations about Ubald are derived by his two main biographers: Iordanus (Giordano) of Città di Castello[1] and Theobaldus (Teobaldo), his successor in the episcopal seat.[2]

We found interesting the description of the disease that struck him. Iordanus reported (Chapter 19.2–19.5, translating from Latin): 'Already in advanced age, with the body marked by incredible penitences, really exhausted from excessive suffering, because once he had broken an arm and twice a leg, he was affected by a disease who covered entirely his body of tiny pustules, "minutissimis vulneribus," like a second Job. From these wounds got out a copious filthy liquid, his underwear was changed five times a day... the replaced underwear, soon just cooled, became rigid like a dried leather. He had to stay in bed for a long time in painful position between two not flanked benches, he could rest his head on one of these, and the feet on the other, in the middle was the body; he only could have a bar, which joined the benches like a bridge, and he sustained himself to this bar with the hands and knees... about his body, it was remained only skin and bones: his flesh, almost completely removed by the skin, was likely deep burned. Each cloth or object that touched his body, it was like a torment for him.'

From this old description, we can deduce that a blistering disease affected the Saint at 74 years old. The symptoms and the way of developing of the disease may suggest a Bullous Pemphigoid (BP), an autoimmune blistering disease most common among elderly people.[3,4] BP is an autoimmune skin disease, which usually consists of tense pruriginous blisters on erythematous skin. It is a debilitating condition even described as fire under skin. If untreated, large parts of the body may become disepithelized, with subsequent loss of fluids and a clinical aspect that resemble a deep burned body.

Years of a chronic, progressive disease caused a loss of muscular tissue and heavy loss of fluids which could explain, after death, together with a favourable condition of temperature and humidity, the excellent preservation of the body (Figs 1 and 2).

The term 'tiny pustules', is a traduction from the old manuscripts, so the meaning of this term in the ancient literature may not be exactly the same as actually used. Pruritus is not mentioned, but we know how in the ancient literature this symptom was rarely reported; moreover, in the past pruritus was easily related to contagious or psychiatric diseases, so, even if present, this symptom could never been reported in a description of a saint, being a major focus made on the suffering of last periods of his life.

Figure 1 The satisfactory preservation state of the Saint Ubald mummy, which is now guarded in the city of Gubbio, country of Umbria, Italy, as it was appreciated in the last recognition (courtesy of M. Pierotti).

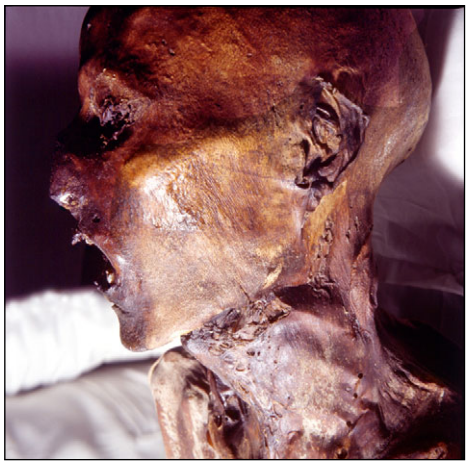

Figure 2 A close view of the face and neck (courtesy of M. Pierotti).

This scanty description was made about 800 years ago, so it is difficult to picture the special bed that Saint Ubald needed, but it is clear that it was designed to prevent the resting of the body at maximum degree, in order to relieve the pain or burning of the illness, like we can see in deep burned patients or in condition characterized by a wide disepithelization, such as chronic untreated bullous diseases.

Moreover, Saint Ubald reported three limb fractures in the last part of his life, so he was probably affected by hypovitaminosis D, which was recently found correlated with BP.[5]

We report probably one of the oldest descriptions of blistering disease where we assume the occurrence of Bullous Pemphigoid.

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Giant orf tumour in an immunocompromised patient

Editor

Herein, we report the efficacy and the safety of open-spray cryotherapy in an immunocompromised woman presenting with tumour-like orf lesion.

A 58-year-old woman was referred with a 4-month history of an undiagnosed, painful, ulcerated, tumour-like lesion on the last phalanx of the fifth digit of her left hand. Six months earlier, the woman had received a cadaveric kidney transplant for an indication of focal segmental glomerulosclerosis. Since then, the patient had been receiving immunosuppressive therapy with a combination of tacrolimus (8 mg/day), mycophenolate mofetil (1 g/day) and prednisone (5 mg/day). The ulcerated digital lesion was first diagnosed as a whitlow and was surgically excised. Histological analysis revealed an ulcerated pyogenic granuloma. The lesion recurred a few days later and had grown (accompanied by increasing pain) to five times its initial volume after 1 month (measuring 13 cm of circumference in the greatest dimension, Fig. 1a). A physical examination was otherwise unremarkable, and an X-ray of the hand was normal. Histological examination of a novel skin biopsy evidenced florid granulation tissue with eosinophilic intracytoplasmic viral inclusions in the upper layers of the epidermis (Fig. 2). A PCR assay confirmed the diagnosis of human orf disease. A month-long course of valaciclovir (3 g/day) and imiquimod (once a day for 5 days a week) and a twofold reduction in the dose of immunosuppressive drugs (tacrolimus 4 mg/day and mycophenolate mofetil 500 mg/day) had no effect. Thus, additional, weekly open-spray cryotherapy (one cycle: 200 s) was initiated (with local anaesthesia). Complete regression was obtained after seven sessions, with no recurrence in the following nine months (Fig. 1b).

Orf is a parapoxvirus infection acquired through zoonotic transmission.[1] Orf is common among persons with contact with sheep and goats, including farmers and veterinarians. It usually manifests as an erythematous papule, predominantly located on the hand, spontaneously healing within 4–6 weeks. No specific treatment is required. This infection is being reported with increasing frequency, particularly in immunocompromised patients from whom orf could take atypical forms such as tumour-like lesion that does not regress spontaneously.

Only a few cases of orf in immunocompromised patient have been described. Thus, a uniformly effective treatment for these patients has not been yet identified. The therapeutic option that is first considered is often a surgical excision, although this

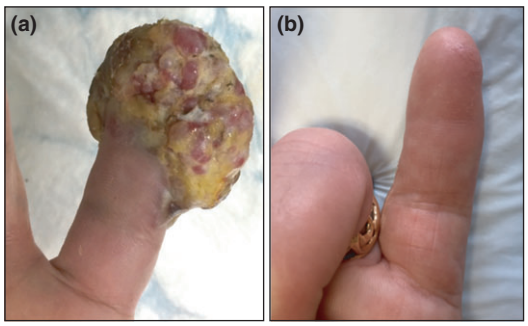

Figure 1 Clinical presentation of the orf disease before and after cryotherapy. (a) Initial presentation of the digital giant orf lesion. (b) Complete resolution of the giant orf tumour after seven sessions of cryotherapy.