Noma Masquerading as Squamous Cell Carcinoma - A Case Report in an AIDS Patient

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Abstract

Rationale: Cancrum oris, also known as noma, is a rare and rapidly progressing gangrenous infection affecting the oral cavity, commonly seen in malnourished children. We discuss the clinical presentation, diagnostic dilemma and management in a 54-year-old male human immunodeficiency virus-positive patient with oral ulceration clinically resembling squamous cell carcinoma (SCC). **Patient Concerns:** The patient had severe oral ulceration with pain and difficulty eating food. **Diagnosis:** Histopathological examination was misleading as it revealed features akin to early invasion of SCC. Immunohistochemistry findings were not in favour of SCC. **Treatment:** Antiretroviral therapy was started owing to the decreased CD4 cell count. **Outcomes:** Lesions began to show signs of healing on follow-up. **Take-away Lessons:** This case aims to highlight the unique challenges of diagnosing and emphasises the importance of considering opportunistic infections in immunocompromised patients presenting with oral ulcerations to prevent misdiagnosis and maltreatment.

Keywords: Acute immunodeficiency syndrome, antiretroviral therapy, cancrum oris, noma, squamous cell carcinoma

Introduction

Noma, a rare life-threatening necrotising infection, primarily affects malnourished children. The aetiology of noma is multifactorial and often associated with a combination of contributing factors such as malnutrition, immunodeficiency, poor oral hygiene, bacterial infection, viral coinfections, socioeconomic status and immunogenetic factors.[1] In acquired immunodeficiency syndrome (AIDS) patients, the risk of noma is heightened due to compromised immunity.[2] Cancrum oris can be especially difficult to diagnose in such patients accurately due to its clinical resemblance to squamous cell carcinoma (SCC) in the form of oral ulcerations. Early diagnosis and appropriate management are essential to improve patient outcomes.

Case Report

A 54-year-old male patient sought medical attention with a complaint of severe oral pain, ulceration in mouth, facial swelling and difficulty in eating and speaking for the past one month. The patient reported a history of episodes of fever and redness, irritation and swelling in relation to the right eye. In addition, the patient also reported experiencing rashes and pustules over the skin, most commonly on the upper arms for two weeks. The patient was unaware of any significant medical history. Clinical examination revealed extensive ulceration in the right buccal mucosa. The lesion was rapidly progressive, with irregular borders, central necrosis and surrounding erythema [Figure 1]. The skin opposite the lesion did not show any abnormality with no symptoms of paraesthesia. On ocular examination, visual acuity was 20/20 in both eyes; however, redness of conjunctiva was present along with pustules in the lower eyelid of the right eye [Figure 2]. Head-to-toe examination revealed the presence of pruritic rashes in shoulder, trunk, upper arms and lower limbs. Submandibular lymph nodes were normal in size and consistency.

Routine laboratory investigations were advised along with serological tests and the patient was diagnosed as human

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immunodeficiency virus (HIV) positive. His CD4 cell count was 142 cells/μL which indicated the condition of AIDS. Given the clinical presentation and the patient's immunocompromised status, the differential diagnosis included cancrum oris, SCC, fungal infection, tuberculosis and acute myeloid leukaemia. Ultrasonography was done which revealed lymph nodes normal in size and consistency. A biopsy was carried out with all necessary precautions and wound care and debridement were done. The patient was immediately started on broad-spectrum intravenous antibiotics, antiretroviral therapy (ART) (dolutegravir, lamivudine and tenofovir, disoproxil fumarate) and nutritional support. Locally applied drugs were avoided.

Histopathological examination revealed features of SCC with extensive necrosis along with atypical squamous cells showing dysplastic features such as basal cell hyperplasia, individual cell keratinisation, nuclear pleomorphism and altered nuclear–cytoplasmic ratio. Hence, these findings initially raised suspicion of SCC. However, the presence of abundant acute and chronic inflammatory cells, vascular congestion and vascular thrombosis warranted consideration of infectious aetiology. To confirm the diagnosis, immunohistochemistry was performed, and it showed nests of squamous cells with intact p63 and p40 and ki67 in basal lining cells. Immunohistochemical markers were not in favour of SCC which confirmed the diagnosis of cancrum oris [Figure 3].

Follow-up after one week revealed lesions of acute necrotising ulcerative gingivitis in the hard palate and gingiva of the patient. On further follow-up after two weeks, the oral lesion showed a remarkable response to treatment with a decrease in inflammation and gradual re-epithelisation, which confirmed noma infection [Figure 4]. In addition, the patient showed significant improvement in skin and ocular lesions and overall general health.

Discussion

A comprehensive evaluation and multidisciplinary approach are crucial to differentiate between malignancy, cancrum oris

Figure 1: Intraoral appearance showing ulceration in the right buccal mucosa

Figure 2: Ocular appearance showing redness of conjunctiva and pustule in relation to the lower eyelid

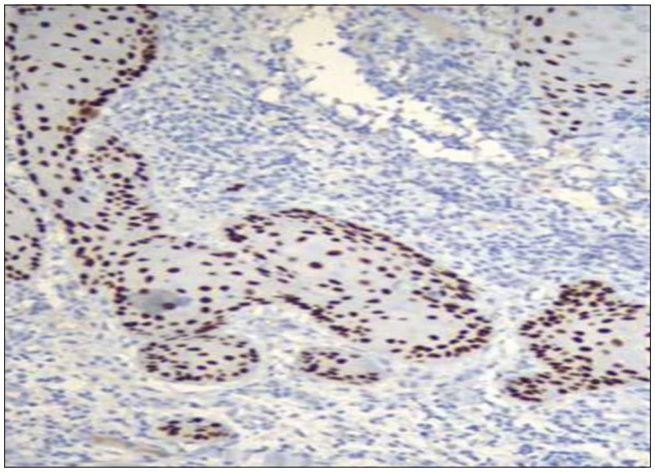

Figure 3: Immunohistochemistry showing markers pathognomonic of noma

Figure 4: Follow-up showing healing of the lesion

and opportunistic infections. Immunodeficiency associated with AIDS patients reduces the body's ability to defend against bacterial infections, allowing opportunistic pathogens to proliferate. Moreover, poor oral hygiene and lack of access to dental care can lead to the accumulation of harmful bacteria in the mouth contributing to the initiation and progression of infection in immunocompromised individuals.[2,3]

Some studies suggest that bacterial co-infection (as with fusobacterium necrophorum, prevotella intermedia, porphyromonas gingivitis and other anaerobic bacteria) and viral co-infections (with herpes simplex virus and cytomegalovirus) predisposes individuals to cancrum oris, especially in those with immunocompromised systems.[4] Diagnosis of noma is based on a combination of clinical presentation, medical history and specific diagnostic investigations. Gathering patients' medical history is vital in diagnosing this condition as immunosuppressed and malnourished individuals are more susceptible to this infection. Differential diagnoses include oral SCCs, gangrenous stomatitis, fungal infections, tuberculosis, acute myeloid leukaemia and autoimmune diseases.[5-7]

Histopathological examination is a crucial step in differentiating SCC from cancrum oris. Microscopic examination of cancrum oris usually reveals characteristic findings of necrosis, inflammatory cell infiltrates and vascular congestion. However, in this case, microscopy revealed features resembling SCCs, so additional tests like immunohistochemistry have to be carried out to confirm noma and to rule out malignancy. In the present case, Mantoux test and fungal culture were done to rule out tuberculosis and opportunistic fungal infections.[5-7]

The treatment modalities should aim in controlling infection, promoting wound healing, managing underlying immunosuppression and addressing nutritional deficiencies. The treatment strategies include a combination of broad-spectrum antibiotics, ART, surgical debridement, nutritional support, pain management, psychological support and management of complications like sepsis.[8-10] ART is the cornerstone of treatment for HIV patients. ART typically involves the use of at least three antiretroviral drugs from different classes to target various stages of the HIV cycle. ART helps in reducing viral load, improving CD4 cell count and restoring immune function. In our present case, highly active antiretroviral therapy (HAART) was followed for the patient and the patient showed significant improvement within three weeks.[8-10] Thus, timely diagnosis and early initiation of appropriate treatment are necessary to control the infection and prevent complications associated with it and ensure a favourable outcome.

Conclusion

Noma in AIDS patient can clinically and histopathologically mimic SCC, necessitating a thorough evaluation and definitive diagnostic tests. The mortality rate of noma can be as high as 90% where the infection is not treated promptly and effectively. Early recognition, prompt initiation of treatment and a coordinate approach involving different specialities are essential to improve outcomes and enhance the quality of life for AIDS patients affected by this aggressive infection. Proper adherence to ART and measures to address malnutrition and immunodeficiency can potentially prevent the occurrence of noma in vulnerable populations.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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