Noma: A Rare Abnormal Presentation in a Cameroonian Child

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Abstract

Noma is a disease which causes a progressive and mutilating destruction of tissue. The localisation is frequently facial, meanwhile other sites do exist although rare. We present, the case of a 13 months old female child, admitted in the pediatric unit for a foul-smelling ulcerative wound on the neck with weight loss in the past 3 weeks. The diagnosis of noma with an oro-cervical localisation, was made on the basis of the ulcerative necrotic nature of the wound with a poor buccal hygiene, the age, and the nutritional status. She was treated with large spectrum antibiotics, surgical debridement, and nutritional rehabilitation. The outcome was favourable with regression of the lesions.

Keywords: Noma; Malnutrition; Antibiotic treatment

Introduction

Noma (also called cancrum oris) is a gangrenous necrotizing disease, which causes a progressive and mutilating destruction of the facial tissue [1-3]. This pathology was first described by Hippocrates since the 5th century and has remained spread worldwide. It is the consequence of a complex reaction between multifactorial immuno-depression of the host and neglected intra-buccal infections [1]. With improvement of living conditions and the advent of antibiotics, it has greatly regressed since the 19th century [2]. It persists in developing countries, where it is strongly linked to extreme poverty and its consequences, such as malnutrition and limited access to health care [2-6]. It affects mostly children under 6 years of age, with no sex preference, and carries a mortality rate of about 90% in the acute phase, if not well managed [7]. It often has an oro-gingival on-set and later extends to the face [8]. We present here the case of an oro-cervical noma, which is an abnormal localisation, in a 13 months old Cameroonian child.

Case report

A 13 months old female child was hospitalised in the Pediatric unit of the Yaounde Gynaeco-Obstetric and Paediatric Hospital (YGOPH) in 10th January 2017 for a wound on the neck with weight loss, since more than 3 weeks.

It all started with weight loss and little ulcerations in the mouth which later evolved to a painful facial oedema and progressively extended to the neck. She was given oral antibiotics (Sulfamethoxazole-Trimethoprim and Cloxacillin) at home for 3 days, without any improvement. A few days later, a point-like opening was noted on the right side of the neck, which later got swollen and discharged pus. The evolution was marked by a centrifugal extension of this lesion which became more painful and extruding. The appearance of a dark spot at the centre of this lesion motivating consultation in our hospital.

In the past history, the child was born from an HIV infected mother on treatment and weighed 3500g. She had received antiretroviral prophylaxis for mother-to-child prevention of HIV transmission from birth to 2 months 1/2. We also noted that family food-stuffs and breastmilk substitutes were introduced in her diet at the age of 2 months. She was not well vaccinated for her age (she had not receive the measles vaccine at 9 months), and lived in a 2-bedroom house with 14 people.
On admission, the child had an altered general state with visible signs of malnutrition (soft, brownish hair, protrusion of the ribs, and muscle wasting). She was febrile with a rectal temperature of 38.5°C. Examination of the neck revealed a large, deep, ulcerative wound at the right lateral side, painful, and 10cm in the largest diameter with a hardened necrotic centre figure 1, 2. The weight was 5.15kg, height 72cm, and a weight- for - height Z score less than -3. The mid upper arm circumference was 110 mm and there was no edema. The diagnosis of an oro-cervical noma on severe acute malnutrition specifically marasmus was made. The differential diagnosis was a Burkitt’s lymphoma, a cutaneous leishmaniosis, burulli ulcer or syphilis (Figure 1).

Burkitt’s lymphoma was clinically ruled out because of the ulcer - necrotic nature of the lesion, localization, associated malnutrition and absence of enlarged lymph nodes; whereas burulli ulcer occurs mainly on the limbs and in endemic areas, and a syphilis sore was unlikely considering the age of the child and localization of the lesion. Cutaneous leishmaniosis was also unlikely because of its mode of onset (its starts with a papule which enlarges to form a nodule, and tiny vesicles form on the nodule and will later ulcerate to form an ulcer which is often painless but very pruritic), and also for the fact that it occurs in endemic areas.

A complete blood count showed 15 000 white blood cells/mm³ with 65% neutrophils, normocytic normochromic anaemia with a haemoglobin of 7.3 g/dl. The HIV serology test was negative, and the C reactive protein was 48mg/L. Blood cultures and culture of pus samples collected from the wound were all sterile, probably due to prior antibiotic administration before admission. This patient was placed on broad spectrum intravenous antibiotics (ceftriaxone, metronidazole, and gentamicin). Nutritional management was done with therapeutic milk products (F75 and F100) during hospitalization. Necrosectomy was done, and daily local wound cleaning and dressing was also done with 0.9% saline solution.

On the 2nd day of hospitalisation, we observed minute collections of pus in the subcutaneous tissues of the right jaw and were drained (Figure 2).

A more extensive necrosectomy was done on the 3rd day of hospitalisation and this greatly relieved the patient as she regained appetite. On the 7th day, we noted sprouting of the wound, with gradual closing-up of the edges by the 12th day (Figures 3 and 4).

This patient was discharged on the 14th day with the wound almost entirely healed, and the nutritional status was much improved.

**Discussion**

Noma is a mutilating, necrotizing disease of the face [1,2,3]. The initial primary lesion is usually acute gingivitis, which quickly becomes ulcerative-necrotic with progressive extension from the mouth towards the skin, and classically extends progressively from the mouth to the rest of the face [3,9,10]. Although most classifications limit to facial lesions [11], other authors have observed that tissue destruction could sometimes be of insidious-onset, minimal, or deep and more extensive [10,11]. Besides, the spontaneous occurrence of a cutaneous cervical opening before the gangrenous stage in our patient strongly suggested fistulization from an initial neglected gingivitis. This could explain the oro-cervical presentation in this child. Examination of the oral cavity showed inflamed gums and caried teeth.

In 90% of cases, noma affects children of between 2 and 6 years of age who live in poor-resource settings, [1,3,4,12]. It is a multifactorial disorder resulting from a complex interaction between the host’s immuno-depression, malnutrition and intra oral lesions [5,6,12,13]. Severe acute malnutrition was among the main risk factors in our patient. Malnutrition could also act directly, leading to a cellular immune deficit from late lymphocyte maturation [6,10], and causes fragility of the mucosa which will no longer perform well its role as a barrier within the framework of innate immunity [14]. It can also cause debilitating infections which might aggravate it through a vicious cycle [1,6]. In our patient other risk factors could be noted, such as incomplete immunizations, being born from an immunocompromised mother although the child was not infected. Poverty, promiscuity with unhygienic lifestyles, lack of access to health care services, and poor body and oral hygiene could have also exposed the patient to such situations [6,12].

The implication of anaerobic pathogenic bacteria in this pathology is obvious with the typical foul odour, and the spectacular improvement of the lesions after administration of broad spectrum antibiotics, in adequate doses and for an adequate time. Bacteria involved include fusobacterium which produces dermatotoxins and haemolysins responsible for the destructive lesions of the disease [6,13], and Prevotella implicated in parodontal diseases [10,15].

Sterile cultures are frequent and could be due to prior antibiotic administration, poor sample collection and conservation, or even the culture methods used [16]. It is therefore not unusual for a particular pathogen not to be isolated from cultures of the wound, especially if the patient had already received antibiotic treatment, as was the case with our patient.

The aim of management is to sterilize the site of infection and limit the extension of tissue damage. Antibiotherapy is indispensable to curb the infectious cycle. The association of β-lactamines and metronidazole for anaerobic germs are most efficient for treatment [17]. Our patient received triple antibiotherapy, comprising ceftriaxone, gentamycin and metronidazole, associated with surgical debridement and nutritional rehabilitation.

Surgical debridement favors rapid deterion and healing of the wound, and improves the patient’s comfort [10]. Nutritional rehabilitation and treatment of infections increases the chances of fast healing and recovery [5,10]. Without treatment, the mortality rate rises to as high as 90% and this is strongly related to acute complications such as septicemia, and dehydration, and management of co-morbidities [7]. When the ulcerative-necrotizing gingivitis appear, evolution to gangrene will depend on the underlying disease over a few hours or days (8,10).

Prior antibiotic administration can prolong the onset of the gangrene [10,11] Even at the stage of gangrene, good antibiotic treatment with adequate management of other co-morbidities can help in minimizing tissue loss and shortening the stay in hospital.

Unaesthetic retractile scarring are part of the possible eventual sequelae of noma [1,4,10]. These have a negative impact on the psychologic aspect of the patient and render social integration difficult [10]. The wounds of our patient at the end of hospitalization were healed, but the initial large size and cervical localization (with skin

folds) increased the risk of retractile scarring. In our patient follow-up was planned to be regular, so that any problems with scarring could be surgically repaired.

Conclusion

Noma is a debilitating disorder, affecting mostly young children in poor resource settings. The cause is multifactorial with a complex vicious circle of malnutrition, immune deficiency and mouth infections. The case described was a neglected cervical necrotising lesion, not as the classical noma which has an endo-buccal infection with orofacial extension. In the absence of treatment, the mortality rate could be as high as 90%, and survivors might have severe facial aesthetic problems. It could be prevented by improving the living standards, ensuring good oral hygiene and the prevention of severe infections through routine vaccinations.

Bibliography


