

Craniofacial Necrotising Fasciitis: A Case Report and Review of the Literature

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ABSTRACT

Objective: To report a rare case of craniofacial necrotising fasciitis and to review the aetiology, pathogenesis, diagnosis, complications, treatment and outcome.

Case Report: We report a 62-year old Nigerian female with craniofacial necrotising fasciitis of dental origin. A comprehensive review of the current data regarding craniofacial necrotising fasciitis was conducted using MEDLINE, PubMed, Scopus and Google Scholar.

Conclusion: Early diagnosis is of utmost importance. Quickly spreading erythema and extreme pain in the affected region serve as red flags. Extensive surgical debridement with antibiotic therapy remain the mainstay of treatment

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INTRODUCTION

The term necrotising fasciitis (NF) is used to describe a rapidly spreading infection involving the superficial fat, fascial layers with necrosis of skin. It is a disfiguring and fatal condition. It is characterised by its fulminating, devastating and rapid progressive course. Though there is rapid necrosis of the subcutaneous tissue and underlying fascia, there is characteristic sparing of muscle and bone (Wilson, 1952). Since NF was first reported by Wilson in 1952, a number of cases have been reported in American and European literature (Gillis & Gillis, 1992; Benavides et al., 2003; Yoder and Deep, 2004). Most often, the trunk, perineum and lower extremities are affected. Involvement of the face from NF is rare. Only 35 such cases have been reported in the visible literature since 1960. It is associated with a high mortality and can result in severe facial disfigurement and poses challenging reconstructive problems (Marrioni et al.,

2003). In 1997, Shindo et al. reported four new cases and presented a review of 35 previous true cases of NF of the face.

Head and neck are unusual site which is rarely

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affected. The NF in the head and neck region can present with three types: the cervical fasciitis (CNF), Craniocervical fasciitis (CCNF), and Craniofacial fasciitis (CFNF). The CFNF is the rarest form of NF in the head and neck region and has been described as the necrosis of the face, and sometime of the eyelids and the scalp (Obiechina et al., 2001; Ndukwe et al., 2002).

NF of the Head and neck usually results from odontogenic infection (Stoykewych et al., 1992; Whitesides et al., 2002; Umeda et al 2003), though

cases from peritonsillar abscess (Zilberstein et al., 2005), sinusitis (Raboso et al., 1998), salivary gland infections (Marioni et al., 2003), otologic (Gullung et al., 2014) and dermatologic abscesses (Fung et al., 2012) have been reported. Though it was considered a polymicrobial entity, most are usually due to β -haemolytic streptococci. It usually occurs in patients with systemic conditions such as diabetes mellitus, renal disease, cardiovascular disease, human immunodeficiency virus (HIV) infections and acquired immunodeficiency diseases (AIDs) and so on, but it can also occur occasionally in a non-immunocompromised individual. Complications that can arise if not treated early includes airway obstruction, jugular venous thrombosis, aspiration pneumonia, mediastinitis, descending necrotising fasciitis (DNF), ascending necrotising fasciitis (ANF), empyema, and lung abscess (Lawani & Kaplan, 1991; Shand et al., 2001; Serna et al., 2012). Prompt diagnosis, recognition of acuity, aggressive and serial surgical debridement and medical management contribute to improved survival. NF of the head and neck was reported to have a mortality rate of 7% to 20%, depending on the extent of neck involvement (Barie, 2004; McGurk, 2003).

The extreme rarity of CFNF result in poor characterisation of this entity in the literature. While several case reports and case series of CNF and CCNF are documented globally (Sepulveda & Saatro, 1998; Serna et al., 2012), few or none was reported on CFNF. To the best of our knowledge, it appears this is the first case of CFNF to be reported in Nigeria. The purpose of this paper is to report a case of CFNF in a Nigerian female in the hope that it will add to the literature of the few cases reported from Africa.

CASE REPORT

A 62-year-old Nigeria woman who was referred from a private hospital to our Oral and Maxillofacial surgery clinic with one-week history of facial swelling which was preceded by bilateral maxillary teeth ache with poorly controlled blood pressure.

Examination showed a pale, ill-looking patient with diffuse facial swelling involving the zygomatic, buccal and the ocular region bilaterally. There was slight, dark change in coloration, vesiculation and paraesthesia of the skin over the right zygomatic region (Figure 1). There was limitation in mouth opening (1.5cm), retained root of teeth number 24, 25, 26 and 27 and periodontitis of 16 and 17. Initial ophthalmologic team review showed poor light perception, sluggishly reacting right and fixed left pupils.



Figure 1: Patient at presentation

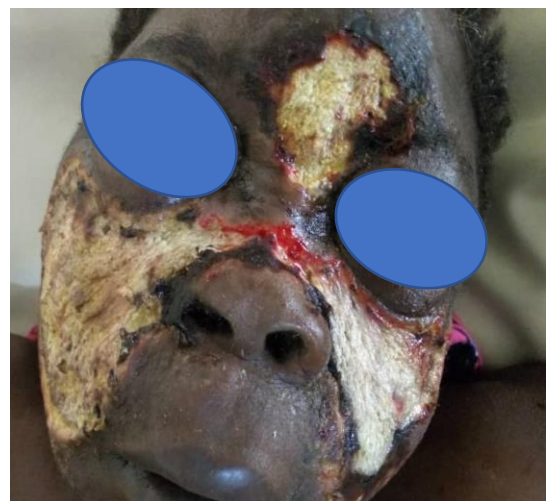


Figure 2: Patient after first attempt at debridement



Figure 3: Patient after second day of debridement



Figure 4: Patient with the nasogastric tube after one week of debridement

Investigations showed a pack cell volume (PCV) of 14%, white blood cell count was elevated, retroviral screening (RVS) was negative and serum renal parameters were normal. MCS results yielded *Raoultella ornithinolytica* formerly *Klebsiella oxytoca* or *Klebsiella ornithinolytica* and Coagulase negative *Staphylococcus*. Only the latter was sensitive to an antibiotic, imipenem. Fungi tissue culture as well as blood culture yielded no growth. Tissue biopsy showed mainly necrotic tissue. CT scan review showed iso-density of the maxillary, frontal and ethmoidal sinuses, intact globes bilaterally and no evidence of bony involvement. An initial assessment of fascial space infection secondary to odontogenic infection in a medically compromised patient was made.

Incision and drainage with extractions of the aforementioned teeth was done under local anaesthesia. Patient was placed on antibiotics and had transfusion of four units of blood, which brought the PCV to 25%. Blood pressure was controlled by the medical team with amlodipine and lisinopril. However, one week after incision and drainage, the dark change in colouration of the involved skin was noticed to have become deeper and spread further to the contralateral side and the frontal region, with increase in area and depth of anaesthesia. Ophthalmology team review at this time showed complete loss of vision in both eyes. A diagnosis of craniofacial necrotising fasciitis was made. Patient refused consent for aggressive debridement under general anaesthesia, as such, twice daily debridement was instituted (Figure 2 & 3), and this was done to the extent that the patient could tolerate with the aid of local anaesthesia. Nasogastric tube was passed for feeding, and ceftriaxone was changed to imipenem which the patient reacted to with seizure and was later

changed to meropenem (Figure 4). Further ophthalmology team assessment reported rupture of the left globe, and advise for immediate orbital exenteration under general anaesthesia. Patient decline consent to exenteration and requested for a discharge against medical advice.

Informed consent to use the clinical photographs for scientific presentation and publication was obtained from the patient by the specialist team managing the patients

DISUSSION

Aetiology: NF is mostly polymicrobial. While aerobic microorganisms especially group A β -hemolytic streptococci and coagulase-negative staphylococci (CoNS) are believed to be major role players, it is also known that strict anaerobes play very important role which portray a mixed or synergistic activity. (Umeda et al., 2003), In our case, *Raoultella ornithinolytica* and *Staphylococcus* were the only cultured organisms. *Raoultella ornithinolytica* is a gram-negative aerobic bacillus in the family *Enterobacteriaceae*. This species has been related to histamine-producing bacteria causing subsequent fish poisoning. *R. ornithinolytica* has also been isolated from dentin of infected root canals (Morais et al., 2009).

This is likely due to the fact that the patient has been on broad spectrum antibiotics before presentation. In NF of the head and neck region in immunocompromised patient, the most common cause is odontogenic (Chidzonga, 1996; Umeda et al., 2003). In our case, we found retained root of teeth number 24, 25, 26 and 27 and periodontitis of 16 and 17 which we later extracted. Our patient was also medically compromised with poorly controlled blood pressure and a PCV of 14% at presentation.

Pathogenesis: NF usually begins with local inflammation that spread rapidly along the fascial planes resulting in extensive tissue necrosis and skin changes over 24-48hours. For most cases of necrotizing fasciitis three factors are usually present: (1) Impairment of immune system (e.g., diabetes mellitus, malignancy, and alcoholism), (2) Compromise of the facial blood vessels; and (3) The presence of micro-organisms that are able to proliferate within this area. (Dunn 1997). Thrombosis of the cutaneous vessels leads to cutaneous ischemia, tissue gangrene, necrosis and sloughing. Presence of vesiculation, ecchymosis, crepitus, anaesthesia and necrosis are indicative of an advanced disease (Damstard, 2000). In our case, the patient presented with fast spreading facial swelling which appear cellulitic with subsequent change in skin coloration and loss of sensation down to the subcutaneous layer

which later became gangrenous and necrotic as a result of the thrombosis of the perforating dermal vessels. The rapidity of the spread involved the orbits with consequence loss of vision bilaterally and eventual left globe rupture within a few days.

Diagnosis: Diagnosis of necrotizing fasciitis in the head and neck region requires mainly clinical assessment. However, certain radiographic and laboratory investigations are also helpful. CT scan and MRI may reveal soft tissue gases and can be used in determining the extent of the infection and the anatomic structures involved. It can also help in identifying vascular thrombosis or vessel erosion. Laboratory investigations like MCS, blood culture and full blood count have also proven useful in aiding diagnosis, however treatment should be initiated while awaiting the results. MCS can also help (Umeda et al., 2003) in ruling out other disease condition e.g. erysipelas, deep neck space infections, deep fungal infection and gangrenous necrosis due to clostridium (Jemenez et al., 2004). In our case only *Raoultella ornithimolytica* and Coagulase negative Staphylococcus were cultured. Use of other diagnostic tools like tissue biopsy can also help in identifying disease free margins. Fungal culture is a useful tool in ruling out deep fungal infection. In our case the CT scan shows no involvement of the underline bone. Mycology as well as blood culture yielded no growth. The peculiarity of our case is the rarity of the pattern of spread which was restricted to the face and the frontal region, hence Craniofacial Necrotizing Fasciitis (CFNF), which to the best of our knowledge is the first to be reported in this region.

Complications: Necrotising fasciitis could have catastrophic consequences and could be life threatening hence requires emergency care (McGurk, 2003). In our case especially because of the peculiarity of the pattern of spread the patient develop blindness and rupture of the left globe. Also, there were neither ascending nor descending spread despite compromise of patient's medical condition rather a there was a localized complication of blindness. It can be suggested that anatomical factors play more role in an upward or downward spread of NF since this was never the scenario in the present case reported despite the medical conditions.

Treatment and outcome: Treatment of NF usually involves a combination of early and aggressive debridement, antibiotics and intensive supportive care, including modalities such as hyperbaric oxygen (Lalwani & Kaplan, 1991). Despite best management, report of mortality ranging from 25 to 40% has been documented (Barie, 2004). Intensive rehabilitation is

often necessary to return the patient to premorbid function once the acute infection is treated. In our case, our patient declined aggressive debridement under general anaesthesia which is the mainstay of treatment, as such we had to make do with twice daily debridement with the aid of local anaesthesia. This we could only do as far as the patient tolerated, and it became a limiting factor in our approach to treatment as it is obvious that we could not go as far as the infection has spread in our debridement. This we believe played a major part in the reported complication

CONCLUSION

The role anatomical factor played in the present case has demonstrated the importance of early recognition, prompt antimicrobial therapy and aggressive debridement in effective management of patient presenting with NF.

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