Necrotizing Fasciitis of the Skull Base and Neck in a Patient with AIDS and Non-Hodgkin's Lymphoma

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ABSTRACT

Necrotizing fasciitis is a rapidly progressing, life-threatening soft tissue bacterial infection found more frequently in immunocompromised subjects and rarely in the head and neck. We report a rare case of a patient with acquired immunodeficiency syndrome (AIDS) and non-Hodgkin's lymphoma (NHL) who presented with a high fever and supraorbital cellulitis 1 week after undergoing chemotherapy. He received intravenous antibiotic therapy but soon developed dyspnea and trismus with rapid extension of the cellulitis to the face, ipsilateral infratemporal fossa (ITF), and bilateral neck. An awake tracheotomy was followed by surgical exploration and drainage and debridement of the supraorbital and ITF areas, parotid gland, and bilateral neck. He received intravenous antibiotic therapy and the surgical wound was regularly debrided for 10 days. Following a gradual recovery, the patient was discharged 2 weeks later. Early antibiotic therapy, wide surgical exploration, and a secured airway are the therapeutic mainstay for necrotizing fasciitis of the skull base and neck.

KEYWORDS: Necrotizing fasciitis, head and neck, AIDS, HIV, lymphoma, immunosuppression

Necrotizing fasciitis is an acute life-threatening soft tissue infection which is rarely encountered in the head and neck.^{1,2} Most of the cases of

necrotizing fasciitis of the head and neck involve previously healthy patients,² although patients with an underlying illness, mostly diabetes mellitus,

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immunosuppression, or chronic alcohol abuse, are more susceptible to develop this infection.²⁻⁴ Necrotizing fasciitis usually involves a mixture of aerobic and anaerobic bacteria⁵ (i.e., *Staphylococcus aureus*, *Escherichia coli*, group A Streptococci, *Peptostreptococcus*, *Prevotella* and *Porphyromonas*, *Bacteroides fragilis*, and *Clostridium*), and group A streptococci are particularly associated with a fulminant course.⁴ The disease usually starts with a focal infectious lesion, most commonly from an odontogenic or iatrogenic origin.¹⁻³ The infection then spreads along the superficial and deep fasciae, leading to inflammation of the subcutaneous tissue, muscles, and deep cervical organs. The clinical presentation of necrotizing fasciitis may resemble poorly demarcated cellulitis accompanied by severe swelling, pain, and tenderness. In the head and neck, this process may result in trismus, dysphagia, and rapidly evolving airway obstruction.

We report a rare case of an immunocompromised patient with acquired immunodeficiency syndrome (AIDS) and non-Hodgkin's lymphoma (NHL) who developed fulminant necrotizing fasciitis of the head (unilateral) and neck (bilateral) 1 week following chemotherapy. We describe the presentation, diagnosis, and long-term successful treatment of this patient.

CASE REPORT

A 63-year-old married man with a newly diagnosed NHL presented to the emergency room with a 36-hour history of a worsening right supraorbital swelling, severe localized pain, and difficulty in opening his eye. Six months earlier he had had a severe herpes zoster infection over his right chest. He had received his first chemotherapeutic treatment (cyclophosphamide, doxorubicin, vincristine, and prednisone—CHOP) 1 week before his current admission. He appeared severely ill and feverish. His temperature was 39.6° C. The physical examination revealed swelling, erythema, vesicular rash, and tenderness above the right eye and swell-

ing of the right parotid and submandibular glands. Upon his admission he had no trismus and examination of his upper aerodigestive tract appeared normal. His blood count showed 7700 leukocytes with 97% neutrophils and 200 lymphocytes per mm³. He was hospitalized in an internal medicine ward with the clinical diagnosis of cellulitis and herpes zoster of the supraorbital area, and received empirical treatment consisting of intravenous (IV) ceftriaxone, vancomycin, and metronidazole in addition to Acyclovir.

One hour following his admission, the patient was hemodynamically stable but started to appear dyspneic. Ten hours later he developed progressive trismus and swelling of his right temporal area and his right neck, from the mandible to the clavicle, extending to the contralateral lower neck. Fiberoptic examination revealed mild edema of the supraglottis without airway obstruction. The patient immediately underwent a computerized tomographic (CT) scan with contrast medium of the head, neck, and mediastinum. The scan showed diffuse and marked thickening, stranding, and ill-defined fluid collections involving the right scalp, face, infratemporal fossa (ITF), masseteric space, and neck, which was consistent with necrotizing fasciitis. Fascial and scalp involvement was most prominent in the periorbital region. Neck involvement was extensive and bilateral, more so on the right, with no involvement of the mediastinum (Fig. 1).

The patient was taken to the operating room immediately following the CT scan. Endoscopic intubation was attempted without success and an emergency tracheotomy was performed. The patient then underwent surgical exploration, which included a right hemicoronal incision with a Blair extension in continuity with a collar neck incision. The right temporal fascia showed necrotic foci and was extirpated and sent for culture and microscopic examination. The roof of the orbit and ITF were exposed, and the necrotic tissue above the eyebrow was debrided up to the level of the overlying skin. The superficial parotid gland was exposed and the area along the plane of the facial nerve was drained. Levels I to IV of the neck were explored bilaterally

Figure 1 Axial computerized tomographic scans with contrast medium of the head and neck. (A) Orbital level. (B) Laryngeal level. The scans demonstrate diffuse thickening, stranding, and ill-defined fluid collections involving multiple compartments of the head (right side) and neck (bilateral) consistent with necrotizing fasciitis. Note extensive involvement of the right periorbital tissue.

and severe swelling with fluid collection only, but no pus or necrotic tissue, was found. The wound was rinsed and left open for further drainage and debridement.

The patient remained intubated and was transferred from the operating room to the intensive care unit (ICU). On postoperative day 1, he developed a left lobar pneumonia and neutropenia (1000 neutrophils/microliter). His treatment consisted of debridement and rinsing of the surgical wound three times a day and IV antibiotics (ertapenem, ceftriaxone, vancomycin, and metronidazole), Acyclovir, and leukocyte-stimulating factors (G-CSF).

Pathological examination of the excised tissue showed necrotizing fascia with acute inflammatory infiltrate and hemorrhages (Fig. 2).

Blood cultures drawn prior to the operation were positive for *Streptococcus pyogenes* and *Staphylococcus lugdunensis*. Cultures from the surgical wound were positive for *Streptococcus pyogenes*. HIV serology tests taken during the hospitalization were positive (ELISA HIV 1,2 Ab's, Western blot, P24 Ag). A marked reduction in lymphocyte number and subtypes was noted (total lymphocyte count = 100; CD3 = 68; CD4 = 1 mm³ 0.9%; CD8 = 57; CD4/CD8 = 0.016; B cells CD20 = 0;

NK cells CD56 = 15). Serum viral load was 570,000 copies/mL (LCx HIV RNA quantitative assay). According to the AIDS surveillance case definition of 1993, the patient was categorized as category C3 and antiviral therapy was initiated.⁶

The patient's condition gradually improved, the wound was surgically closed, and he was discharged 10 days later. A CT scan was performed 1 month following discharge and showed no inflammation in the head or in the neck. The patient continued systemic therapy for AIDS and completed chemotherapy for NHL. He died 10 months later from complications of AIDS.

Figure 2 Hematoxylin and eosin-stained microsection of the right temporal fascia showing necrosis with marked neutrophilic infiltration (original magnification $\times 20$).

DISCUSSION

We report a rare case of fulminant necrotizing fasciitis of the head, skull base, and neck in a patient with end-stage HIV and NHL whose immunocompromised state was possibly aggravated by chemotherapy (CHOP administered 1 week earlier).

Bilateral rapidly progressive necrotizing fasciitis of the head and neck in a severely immunocompromised patient carries a high mortality rate.^{3,7} This evolving emergency state requires prompt recognition (both clinical and radiological), prompt implementation of pharmacological measures (broad spectrum IV antibiotic therapy based on blood and wound cultures), and emergent surgery. CT scanning is urgently needed to assist in the diagnosis and to determine the involved area (specifically, to exclude mediastinal involvement and to guide surgical strategy).⁸ In our case, the CT ruled out involvement of the mediastinum and of the right parapharyngeal and left submandibular spaces.

The time from diagnosis to surgery for our patient was less than 2 hours. Surgical management included wide exploration, drainage, and debridement. Daily debridement treatment of the wound is, in essence, the mainstay of postoperative treatment. The wound was left open for further treatment and was closed only when there was no contaminated secretion and when the tissue appeared vital and bled following debridement.

Necrotizing fasciitis occurs most frequently in previously healthy subjects, but there are a few predisposing conditions that can increase the risk for necrotizing fasciitis, including diabetes mellitus or other immunosuppressive conditions.⁹ We found no report relating NHL or chemotherapy to head and neck necrotizing fasciitis, and only two previous reports of cervical necrotizing fasciitis in a patient with HIV.² Chidzonga and his colleagues described the only two cases known to us of patients with AIDS and necrotizing fasciitis.¹⁰ These cases involved two African males presenting with a submandibular abscess from a dental origin, both

treated with antibiotics and wound debridement under local anesthesia. The first patient gradually developed an extensive gangrenous cervical infection within a 5-week period. No control of the wound was achieved and the patient was lost to follow-up.² The second patient was discharged 4½ months following his admission.¹⁰ Our regime included wide debridement under general anesthesia and the patient was discharged in good condition 14 days after the operation.

The source of infection in cervical necrotizing fasciitis is odontogenic in the majority of reported cases and pharyngeal or iatrogenic in the remaining ones. In our case, it is most likely that the orbital lesion, clinically assumed to be herpes ophthalmicus, was the port of entry for a subsequent bacterial superinfection that later developed into a fulminant necrotizing fasciitis. Nevertheless, in a patient with end-stage HIV (AIDS) and CD4 count < 50, any opportunistic pathogen could be the source of the infection.¹¹

Necrotizing fasciitis is an uncommon disease that rarely involves the head and neck. Prompt diagnosis, early instillation of broad spectrum antibiotic therapy, surgery, and postoperative debridement of the wound are essential for the patient's survival, even in advanced disease such as AIDS and NHL.

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