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EXTENSIVE CERVICAL ACTINOMYCOSIS : A CASE REPORT

KEY WORDS: Actinomyces, Actinomyces israelii, Cervical

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ABSTRACT

Actinomyces today is a rare disease affecting 1 in 3 lakhs people (0.0003%). It is a chronic disease caused by a group of gram positive, anaerobic bacteria, namely *Actinomyces israelii*, that normally colonise mouth, colon and urogenital tract. Infections involving cervicofacial area is the most common clinical presentation. The diagnosis is made by histopathological identification of organism in tissue biopsy. Although medical therapy alone is frequently sufficient for cure, combined medical-surgical therapy may be required in some cases. An awareness of full spectrum of disease, prompting clinical suspicion, can expedite the diagnosis and treatment. Due to its rarity and its propensity to mimic many other disease, there is a chance of missing its diagnosis and proper treatment, making it a challenge. We have described a case of 40 year old male having cervical Actinomyces with extensive distribution in the neck the larynx.

INTRODUCTION

Actinomyces is an indolent, chronic bacterial infection caused by a gram positive, non acid fast, anaerobic to microaerophilic bacteria primarily of the genus *Actinomyces*. These bacteria normally colonise oral cavity, colon or urogenital system. As described by Russo, most common micro-organism causing infection are *Actinomyces israelii*, *A. naeslundii*, *A. gerencseriae*, *A. odontolyticus*, *A. viscosus* and *A. meyeri*¹.

First described in 19th century and common in pre-antibiotic era, Actinomyces has diminished in incidence in recent times. Actinomyces has no geographic boundaries and occurs throughout life with peak incidence in middle age and males having three fold higher incidence than females¹. As per Acevedo et al (2008), no person to person transmission has yet been documented².

Cope classified Actinomyces infection into three distinct clinical forms³:

- Cervicofacial (50%)
- Pulmonothoracic (30%)
- Abdominopelvic (20%).

Cervicofacial actinomyces (lumpy jaw) is the most common clinical presentation.

Rarely, it may involve central nervous system, musculoskeletal system or present as a disseminated disease.

The Actinomyces species usually grows in soil and often affects farmers and gardeners. It enters human body by inhalation and remains harmlessly in oral cavity and upper respiratory tract unless tissue lining is damaged. Oral trauma, dental extraction, dental caries, periodontal diseases can act as predisposing factors to the disease.

Contiguous spread : Since the bacteria is non virulent, the critical step in the development of Actinomyces is the disruption of mucosal barrier and presence of devitalised tissue through which it spreads contiguously in a slow, progressive manner, ignoring tissue planes; thereby mimicking malignancy¹.

Polymicrobial infection : Brook asserted that the infection is

polymicrobial where the companion bacteria act as copathogens by inhibiting host defences and reducing local oxygen tensions⁴.

Granulomatous and Suppurative inflammation : The host response is manifested as intense suppurative and granulomatous inflammation¹.

With an early diagnosis and prompt treatment a cure rate of 90% can be expected with reduced mortality(0-28%) and morbidity². However the diagnosis of this 'masquerader' remains a challenge.

CASE REPORT

A 40 year old male patient, a chronic tobacco chewer for 20 years and a farmer by occupation, presented with complaints of midline neck swelling since 12 year with multiple skin ulceration over neck since 7 years and change of voice since 1 year. Patient gave a past history of being operated twice for neck swelling twelve years and ten years back without documented evidence. Seven years back patient has presented again with neck swelling and multiple skin ulceration for which he took frequent medication from private hospitals with intermittent relief.

On clinical examination neck swelling was seen with multiple non healing ulcerations having whitish discharge (Figure 1). Oral cavity examination revealed multiple carious and tobacco stained teeth. Patient underwent endoscopic examination which showed soft irregular lesion on epiglottis and small red lesion on vocal cords.

Patient was advised CT neck with contrast which was suggestive of nodular lesion in submental, bilateral submandibular, pre laryngeal, pre hyoid, epiglottis, bilateral ary-epiglottic folds and supraglottic larynx. Narrowing of supraglottic airspace was also noted (Figure 2).

His blood parameters were within normal limits. The discharge was collected for culture and sensitivity which showed heavy growth of coagulase negative *Staphylococcus Aureus* and tissue biopsy was sent for histopathological examination which revealed "*Actinomyces israelii*" (Figure 3).

Patient was then followed and treated with injectable penicillin (injection PPF 4 lakhs IU intramuscularly every 3 weekly) after negative test dose along with oral penicillin (800mg per day) which

showed remarkable results in his recovery. He was briefed about the need of long term treatment and was followed up every 2 months. A year after the therapy the patient has shown considerable improvement with significant reduction in size of the swelling and healing of ulcerations. There is no evidence of recurrent lesion since last one year.

DISCUSSION

Cervical Actinomycosis is an uncommon but fascinating infection of the head and neck. It has been called by Russo 'the most misdiagnosed disease', and it has been said that no disease is so often missed by experienced clinicians¹. During the 1970s, an annual incidence of 1 case per 300 000 was found in Cleveland area². During same period, Desai et al reviewed 40 cases of mycetoma infection in India, in which there were 2 cases of *Actinomycosis israelii* infection of the cervicofacial region³. Recent reports are however rare. A review of literature, as described in Volante et al study, revealed 48 cases of cervicofacial Actinomycosis reported over the last 25 years³. Improved dental hygiene and the widespread use of antibiotic treatment have contributed to a decrease in this incidence.

As seen in the present case study, the disease is more commonly seen in middle aged men, usually endemic in farmers, gardeners or stable workers in whom the causative organisms, *Actinomyces* sp. (most commonly *A. israelii*), are often present as commensals in oral cavity, until the tissue-lining becomes damaged by injury or disease. The bacteria can then penetrate deeper into the body and produce disease with the 'copathogens'. Sometimes, the antecedent history of oral trauma or pathology may not be elicited. No underlying disease or immunosuppression is found in most cases². Classically, Cervical Actinomycosis is characterised in the initial stages by soft- tissue swelling of the perimandibular area. Direct spread into the adjacent tissues occurs over time along with formation of abscesses, fibrosis and woody induration of tissues, and draining sinuses that discharge purulent material. However, according to some authors, this classic formation appearance of woody swelling with sinus tracts draining purulent material is observed in approximately 40% of cases, and, when present, may be helpful in the differential diagnosis³. Symptoms like pain is rare however slight fever may be associated in around 50 % patients³.

The classic clinical presentation that should prompt consideration of this unique infection are¹:

- the combination of chronicity, progression across tissue boundaries and mass like features.
- the development of sinus tract which may spontaneously resolve and recur
- the remission and exacerbation of symptoms occurring in parallel sequence with initiation and cessation of antibiotic administration

In the present case, when our patient presented with recurrent neck swelling with non healing skin ulcers and change of voice, our differential diagnosis included tuberculosis, chronic abscess, other chronic granulomatous lesions, fungal infection or even malignancy. Since Actinomycosis infection is rare, insidious, chronic, has nonspecific symptoms and imitates more common conditions such as malignancy and TB, the diagnosis was a challenge.

The diagnosis of Actinomycosis is suspected clinically and is confirmed on microscopy, culture and imaging. The diagnosis may be established only by a positive culture, however, *Actinomyces* growth is very difficult even on appropriate anaerobic media. Thus, microbiological identification of this organism is often impossible. The macroscopic presence of the classic granules with a yellow sulfur like appearance, termed as 'sulfur granules', in tissue specimens or drainage may be of some help when making diagnosis even if these features are not pathognomic, since nocardiosis may also present with sulfur granules³. However, according to Lancella et al study, the characteristic sulphur granules, in the specimens, are present in only 35-55% of cases and in these cases, the diagnosis is confirmative².

The histopathological examination of the tissue biopsy is the cornerstone for definitive diagnosis of the disease. The microscopic examination reveals a typical finding of an outer zone of granulation and a central zone of necrosis which contains

multiple basophilic granules, that represent lobulated micro-colonies of *Actinomyces*³. Less invasive diagnostic techniques, fine-needle aspiration (FNAC) is not very useful as it shows only inflammatory cells. The radiological investigations does not provide a specific diagnosis but can help us to asses the pattern and spread of the disease⁴.

Actinomycosis requires prolonged treatment with high doses of antimicrobials and Penicillin is the drug of choice for prolonged treatment. Although therapy must be individualised , the intravenous administration of **18 - 24 million units** of penicillin daily for **2-6 weeks**, followed by oral therapy with penicillin or amoxycillin for a total duration of **6- 12 months** is considered as the standard therapy¹. For penicillin-allergic patients, doxycycline, minocycline, tetracycline, clindamycin, erythromycin and cephalosporins have been found to be effective¹. In the present case, coming from a far off village and being the only breadwinner in the family, patient was not willing for a long term hospital stay. So we treated him with high dose injectable penicillin (4 lakh IU) every 3 weekly along with oral penicillin (800mg per day) and the result of our therapy was equivocal to the of the standard therapy described in literature.

Surgical treatment may include incision and drainage of the abscesses, resection of necrotic tissue, and curettage of bone¹. An early diagnosis can often avoid the need for surgery and prevent unnecessary morbidity and mortality. Hyperbaric oxygen therapy can also be used to treat Actinomycosis by increasing oxygen tension which often proves lethal to this anaerobic organism.

Regular and meticulous follow up every 2 to 3 months should be ensured to assess the response to the therapy. Monitoring the impact of therapy with CT and MRI is advisable when appropriate¹.

The preventive measure include maintenance of good oral hygiene and adequate regular dental care, in particular removal of dental plaque, may reduce the density if not the incidence of colonisation and low-grade periodontal infection with *Actinomyces* species.

The prognosis for the treated Actinomycosis infection is excellent if recognised early. The cure rates are as high as 90 % and neither deformity nor death is common. The overall mortality from Actinomycosis ranges from 0-28% depending mainly on the site of infection and the time of diagnosis; CNS involvement being associated with greatest mortality².

CONCLUSION

Based on our case report, it could be concluded that Actinomycosis poses a great diagnostic challenge as it can mimic many other disease because of its insidious course and non specific symptoms. Since medical therapy alone can frequently cure the disease, challenge lies in considering the possibility of Actinomycosis, diagnosing it in the least invasive way and avoid unnecessary surgery. A greater index of clinical suspicion, early diagnosis and appropriate treatment can ensure high cure rates and reduced morbidity and mortality.



Figure 1 : Neck swelling was seen with multiple non healing ulcerations. Whitish discharge was noted.

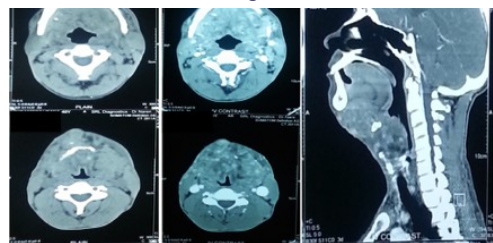


Figure 2 : CECT neck showing the extent of the disease and narrowing of supraglottic airway

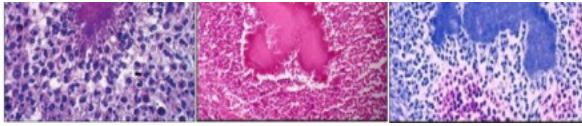


Figure 3 : Histopathological examination showing Actinomycosis israelii and its confirmation on special stains (PAS and ZN stain)

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