

Destructive Potential of Spirochete: Perforation of Hard Palate: A Case Report

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Abstract: Syphilis is a chronic disease with a waxing and waning course, the manifestations of which have been described for centuries. Syphilis has re-emerged as an important cause of morbidity, mortality and a possible transmission factor in the spread of HIV infection. It occurs worldwide, and the incidence varies significantly with geographic location. In the pre-antibiotic era, syphilis was often called "the great imitator" because its signs and symptoms are similar to those of various other diseases. Syphilis manifests in four stages primary, secondary, latent and tertiary. Primary stage manifests as painless ulcer called as chancre at the site of inoculation. Nonpruritic, papular rash, condylomata lata and mucous patches are seen in secondary stage and tertiary syphilis manifests as gumma and neurosyphilis. Gumma is commonly seen on the palate or tongue and rarely with palatal perforation. In this article, we discuss a rare case of palatal perforation caused by tertiary syphilis along in a 55 years old male patient along with the review of literature.

Keywords: Syphilis, Sexually transmitted disease, Gumma, Palatal perforation.

I. Introduction

Syphilis is a sexually transmitted disease (STD) caused by the *Treponema pallidum* spirochete, named because of its resemblance to a twisted thread (*Treponema*) and the pale color (*pallidum*). The term syphilis originates from the poem "Syphilis, Sive Morbus Gallicus" by Girolamo Fracastoro, an Italian physician and a poet (1478-1553), in which the shepherd in poem, Syphilus, is a victim of the disease. *Treponema pallidum* is transmitted by direct sexual contact through vaginal, oral, anal, or vertically across the placenta from mother to fetus. The incidence of syphilis has a myriad of presentations and can mimic many other infections and immune-mediated diseases². Syphilis manifests in four stages: primary, secondary, latent and tertiary stages. Tertiary syphilis may manifest as a gumma in the palate or tongue and also involving the heart, neurosyphilis and perforation of hard palate may also be seen rarely. As syphilis is known to facilitate HIV transmission, and therefore all patients with syphilis should be tested for HIV coinfection, routine screening for syphilis in HIV-infected patients is recommended. Thus, the control of syphilis requires early identification and treatment of such cases. We report a rare case of palatal perforation due to gumma caused by tertiary syphilis as a clinical presentation in a 55 years old male patient.

II. Case Report

A 55 years old male patient reported to Department Of Oral Medicine And Radiology, with chief complaint of inability to eat food since 2 months. Patient had noticed 3 years back a small vesicular eruption on the hard palate which ruptured after 2 months leaving a pinhole opening which had gradually enlarged to attain the present size. Presently patient has a complaint of escaping food through the nose via the palate and change in his speech (nasal tone) since 2 months. Medical history and family history were noncontributory and there was no history of trauma or injury to the palate. Patient gave the history of extramarital affairs since 10 years and been smoking bidis and cigarettes, approximately 2 packets/ day since 40 years. On general physical examination all the vital signs were within normal range. Examination of the lesion revealed, an oval shaped palatal perforation in the middle of the palate extending anteroposteriorly from 24 to 28 region and mediolaterally, 1cm crossing the midline to 1.5cm from gingiva, measuring approximately 2x3 cm and nasal septum was seen through the palatal perforation. It was nontender and soft in consistency. Patient was edentulous except for upper right 1st molar which was carious and root stump was present in canine. (Fig. 1).



Fig 1: Picture of lesion

Based on the history and clinical examination palatal perforation due to tertiary syphilis was given as provisional diagnosis with a differential diagnosis of malignancy of the palate, mucormycosis, and midline lethal granuloma. Patient was advised with various investigations, and on haematological investigations all the values were within normal limits except for the elevated ESR. On serological investigations, VDRL (Venereal disease research laboratory) showed positive reaction and Treponema Haemagglutination Test (TPHA) was reactive and the titer of TPHA was raised to 1:1280. ELISA (Enzyme Linked Immunosorbent Assay) for HIV were non-reactive and Montoux test was negative. Maxillary occlusal cross sectional projection showed bony destruction in the middle of the palate, more on the left side (Fig. 2).

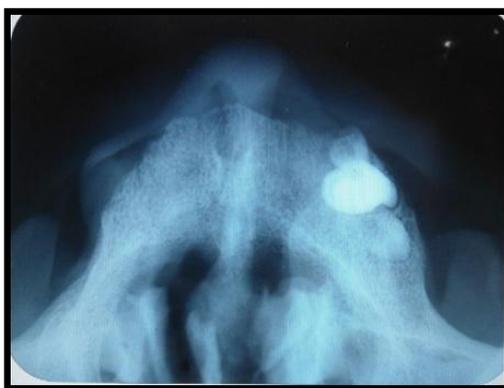


Fig 2: Maxillary Occlusal radiograph

To rule out systemic involvement ultrasound of abdomen, chest- x ray and ECG were performed and no systemic findings were found. Based on the history, clinical findings and investigations a final diagnosis of tertiary syphilis with palatal perforation was given. Patient was advised for obturator and referred to Medicine department for further management and patient's wife was also asked to undergo examination but she did not turn back. Unfortunately patient did not turn out for the follow up and eventually met fatal.

III. Discussion

The origins of syphilis have been discussed for many centuries. Regardless of the origins, however, it remains clear that by 1495 a widespread syphilis epidemic had spread throughout Europe. From there the disease spread to India in 1498 and China in 1505. Early names for syphilis included the Great Pox, Lues venereum (venereal disease), Morbus gallicus (French disease), and the Italian, Spanish, German, or Polish disease, but the name that was to become part of the everyday language was syphilis. In 1905, the association of *Treponema pallidum* with syphilis was described by Schaudinn and Hoffman, who demonstrated spirochetes in Giemsa-stained smears of fluid from secondary syphilitic lesions. August von Wassermann devised a serum reaction test for syphilis in 1906, and thus serologic tests for syphilis were born¹.

Etiology:

The main causative organism of syphilis is a spirochete *Treponema pallidum* (*T. pallidum*) which is a member of the order Spirochaetales, family Spirochaetaceae, and genus *Treponema*, that includes four human pathogens and at least six human nonpathogens. The pathogenic species are *T. pallidum* which causes venereal syphilis, *T. pallidum* endemicum, which causes endemic syphilis (bejel), *T. pallidum* Pertenuae, which causes

yaws, and *T. carateum*, which is the etiologic agent of pinta². *T. pallidum* is a spirochete varying from 0.10 to 0.18 mm in diameter and from 6 to 20 mm in length, making it invisible by light microscopy. Dark-field microscopy is generally used in clinical practice for visualization. The average number of windings is 6 to 14, and the organism has pointed ends and lacks the hook shape seen in some commensal human spirochetes. The bacterium exhibits characteristic corkscrew motility due to endoflagella, with rapid rotation about the longitudinal axis and flexing, bending, and snapping about the full length³.

Epidemiology & Pathogenesis:

The primary mode of transmission is by sexual contact, and the next most common is transfer across the placenta. The majority of infants with congenital syphilis are infected in utero, but the newborn can also be infected by contact with an active genital lesion at the time of delivery⁴. *T. pallidum* is presumed to penetrate through small breaks in the skin. Magnuson et al. were able to demonstrate that two organisms inoculated intracutaneously in rabbits produced a dark-field positive lesion in 47% of cases. This increased to 71 and 100% when 20 and 200,000 organisms were inoculated, respectively. On intracutaneous inoculation, the incubation period varied with the size of the inoculum, so that with a large inoculum, e.g., 10⁷ organisms, a chancre appeared in 5 to 7 days⁵. Animal studies have shown that the organisms appear within minutes in lymph nodes and disseminate widely within hours⁶. Although the exact mechanism by which *T. pallidum* enters cells is not known, it has been shown to attach to mammalian cells in vitro. Attachment may occur by specific attachment ligands. Invasion appears to be a critical virulence factor for *T. pallidum*, as demonstrated by its ability to penetrate endothelial cell monolayers and intact membranes⁷. The pathologic findings of all stages of syphilis are characterized by vascular involvement with endarteritis and periarteritis and in the gummatous stage by granulomatous inflammation. In most primary lesions, the epidermis demonstrates hyperplasia with widening and elongation of the rete ridges⁸. The ulcer surface is covered with an exudate consisting of fibrin, necrotic tissue fragments, and polymorphonuclear leukocytes. A dense inflammatory infiltrate is seen in the adjacent dermis, with predominantly lymphocytes and plasma cells, but histiocytes and polymorphonuclear cells are also seen. The perivascular area is particularly involved and associated with endothelial-cell swelling. Silver staining invariably demonstrates the presence of spirochetes, mainly in the dermal-epidermal junction in the perivascular area. Histopathologic examination of syphilitic aneurysms demonstrates invasion of the aorta by spirochetes. An inflammatory exudate of lymphocytes and plasma cells forms about the vasa vasorum of the adventitia and is followed by obliterative endarteritis of nutrient vessels⁹.

Clinical manifestations:

The acquired syphilis may be classified accordingly to the type of lesion and temporal course of the disease in to primary syphilis, secondary syphilis, latent syphilis and tertiary syphilis. **a) Primary syphilis:** The classic lesion of primary syphilis is the chancre, which is a single, painless, indurated ulcer with a clean base¹⁰. Intraorally chancre may be seen on the lower lip or tongue. The average time from exposure to the appearance of chancre is 3 weeks. The chancre may persist for 4 to 6 weeks and usually heals spontaneously even without treatment¹¹. **b) Secondary Syphilis:** Secondary syphilis is the result of hematogenous dissemination of *T. Pallidum*, which subsequently causes systemic symptoms. This stage typically occurs 6 to 8 weeks after the primary infection¹¹. Secondary syphilis is often a subtle disease; the skin lesions may be easily overlooked and may mimic other dermatologic diseases. The appearance of the skin rash ranges from macular to papular, follicular, and occasionally pustular. Classically, the lesions are described as “raw ham” or copper colored. They tend to be universally distributed on the palms and soles. Skin lesions may be associated with diffuse inflammatory involvement of the pharynx and tonsils, which may cause a symptomatic sore throat¹². The typical lesion of the mucous membranes is the mucus patch, occurring in 5 to 22% of patients. Mucous patches are small, smooth, erythematous areas or superficial grayish white erosions found on the mucous membrane of the vulvar, penis or oral cavity, involving the tongue, buccal mucosa, and lips. Condylomata lata describes grayish moist flat topped extra large papules, sometimes coalescing into plaques, which are commonly found on moist mucocutaneous surface¹³. **c) Latent syphilis:** the disease lies dormant, without any clinical signs except for positive serology and later may manifest as cardiovascular or neurosyphilis²³. **d) Tertiary syphilis:** The characteristic lesion is the gumma, a chronic destructive granulomatous process, which can occur anywhere in the body including the oral cavity. Gummatous destruction of the palatal bone is known to cause perforation of the palate¹³. However, very few cases had been reported so far with the palatal perforation. In the present case palatal perforation was seen due to gumma exposing the nasal conchae. Similar cases have been reported by M.K. Bains and M. Hosseini- Ardehali with palatal perforations caused by tertiary syphilis with associated cocaine habit²¹.

Currently, gummas are rare, reflecting effective treatment with antibiotics. However due to obliterating small vessel arteritis, involving the aorta and the large vessels as well as the CNS, which may or may not require persistence of the organism, continues as the major manifestations of tertiary syphilis. Its involvement usually

occurs between 10 and 30 years after the initial infection. Syphilitic aortitis is the most common manifestation and typically involves the ascending aorta. The most common complication of untreated syphilitic aortitis is aortic regurgitation. Coronary ostial stenosis occurs in 20% of patients with syphilitic aortic insufficiency. Angina is the most frequent symptom but is rarely associated with myocardial infarction. Neurosyphilis produces a wide variety of symptoms on the location and the extent of the lesion. It is manifested as tabes dorsalis and general paresis. General paresis is syphilitic involvement of the cerebral tissue and tabes dorsalis is syphilitic involvement of the dorsal column of the spinal cord and the dorsal root ganglion. Tabes dorsalis affects males more frequently and short, shooting, knife like pain may be experienced in the abdominal region known as tabetic crisis. Patients lose the positional sense in his lower extremities and walks with a characteristic slapping step associated with burning or pricking sensation of the extremities along with paresthesia or anesthesia. With disease progression sensory ataxia affecting mainly the lower extremities becomes a prominent feature. Such individuals are unable to stand erect unaided with his eyes closed, the positive Romberg's sign. Involvement of cranial nerves results in pupils that react to accommodation but not to light suggestive of positive Argyll Robertson pupil which is reported in only 48 to 64% of the patients^{16,17}. Dorsal root ganglion involvement leads to destruction of large joints suggestive of positive Charcot's joints and deep perforating ulcers. Personality changes are often the first manifestation of paresis with irritability, fatigue, mental sluggishness and carelessness in personal habits the patients may have unrealistic ideas of grandeur, wealth and ability. Cerebral gumma may produce symptoms suggesting a brain tumor.

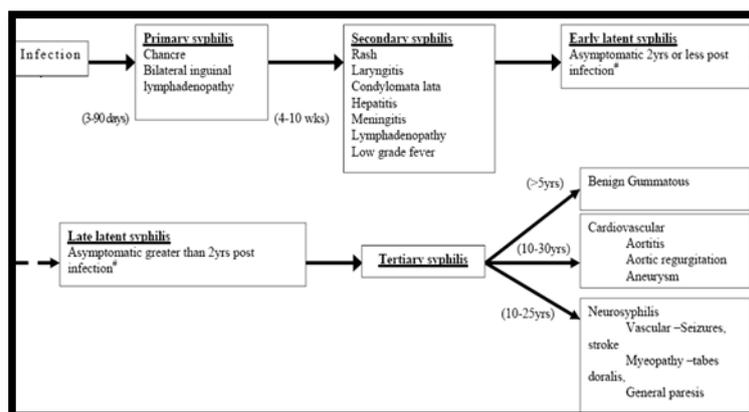


Fig 1: Various Stages of Syphilis

IV. Diagnosis

Syphilis can be diagnosed with dark field microscopy, treponemal and non-treponemal tests. In primary syphilis it is necessary to take exudates from the lesion to demonstrate the presence of living treponemes under dark field microscopy. This examination is useful in genital lesions but not in oral lesions due to the presence of morphologically similar treponemas in the oral cavity. More commonly syphilis is diagnosed using a combination of treponemal and non-treponemal serological tests¹⁸. Non treponemal testing includes the Venereal Disease Research Laboratory (VDRL) and Rapid Plasma Reagin (RPR) tests. These are sensitive tests that are easily analyzed, inexpensive and reliable. Specificity is variously reported as 93-98% with sensitivity varying with stage of disease. Treponemal tests are Treponemal enzyme immunoassay (EIA), T. pallidum haemagglutination assay (TPHA), T. pallidum particle agglutination (TPPA), fluorescent Treponemal antibody absorption test (FTA-abs), and T. pallidum recombinant antigen line immunoassay¹⁹. Enzyme immunoassays with highly purified Treponemal pallidum antigens are more commonly used for screening for syphilis. Diagnosis of neurosyphilis is centered on clinical findings and cerebrospinal fluid (CSF) analysis. Since Treponemes will invade the CNS in 25-60% of cases, Lumbar puncture (LP) for the examination of CSF is recommended²⁰.

V. Treatment

Syphilis has been treated with various remedies over the ages. Previous therapy, before the antibiotics era, with mercury, gave rise to the saying "a night in the arms of Venus leads to a lifetime on Mercury". Arsenic was the preferred remedy but since the advent of penicillin in the 1940's this has been the mainstay of syphilis treatment and indeed the first with the ability to cure. Penicillin therapy is generally given intravenously in the form of aqueous crystalline penicillin G or intramuscular as aqueous Procaine or Benzathine penicillin. The adverse effect of penicillin is anaphylactic reaction. In case of syphilis the Jarisch-Herxheimer reaction occurs, comprising acute febrile reaction with chills, headache and myalgia commencing within the first 24hrs. This

systemic reaction is believed to be due to significant release of cytokines when large numbers of *T. pallidum* are killed by antibiotics. Specific management of the palatal defect may be to seal the defect or to repair the defect. Obturators are a successful method of managing the speech and masticatory problems. Surgery is another option but extensive scarring in syphilitic lesions makes any attempt at palatal repair hazardous. The success was attributed to the excellent vascular supply and the proximity of the donor and recipient sites. Speech therapy can be advised as part of the rehabilitation programmes²¹.

However, palatal perforations are produced by many conditions. The unusual causes are exemplified by the gumma of tertiary syphilis. Hopefully such cases will remain rare. Though medications are available in the present century for treatment of syphilis, rehabilitation by using obturators is simple and cost effective which remains the mainstay of treatment in case of palatal perforations along with the medications. Hence dentists should be aware of such cases and manage appropriately.

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