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Case Report

A rare case of tertiary nasal syphilis: an unusual ear, nose, and throat manifestation of a rare disease

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ABSTRACT

Tertiary syphilis is a rare entity in present antibiotic era. Syphilis of the nose is considered to be very rare by all authorities. It is mainly a systemic disease caused by the spirochete Treponema pallidum. The infection acquired may be congenital or acquired. Tertiary syphilis shows most marked manifestations in nose, causing superficial and deep ulcerations, and gumma. Nasal septum, especially the junction of the cartilaginous with the bony septum resulting in perforation is a usual manifestation. In cases where the bony septum is involved, existence of syphilis is unquestionable. The nasal deformity results from destruction of bony framework of nose and shrinking of fibroid tissue, thus, produces the typical saddle nose which is characteristic of syphilis. The treatment line for syphilis has remained relatively unchanged in recent years and continues to vary with stage of infection. However, local treatment consists of clearance of crusts and regular cleansing of the nasal passages by copious alkaline douches. Yellow mercury oxide ointment may be applied locally. The purpose of local treatment is to remove the discharge and crusts, kill spirochetes, promote wound healing and epithelial growth, prevent secondary infection.

Keywords: Nasal syphilis, Granuloma, Saddle nose deformity

INTRODUCTION

Syphilis is one of the oldest systemic disease having an inexpensive treatment and is completely curable. It is caused by spirochaete treponema pallidum (TP). Sexual transmission is the main mode of transmission. The infection does not lead to immunity against reinfection and repeated episodes of syphilis are reported to occur, predominantly in men who have sex with men (MSM) with a high rate of partner change.[1] The exact prevalence of syphilis in India is unknown due to of several reasons-the stigma related to the sexually transmitted diseases (STDs), poor reporting at STD clinics, lack of common registry for reporting of STDs, symptomatic management at small centres and missing many asymptomatic cases/ partners that do not report for appropriate treatment.[2] As per global statistics, in 2017, the annual rate of primary and secondary syphilis in the United States was 11% higher than in 2016: more than 60% of infected individuals were MSM. The rate continued to rise consecutively through 2020. Despite the antibiotic era, resurgence in cases of syphilis has revived interest both in developing and developed countries. This is because syphilis still presents several diagnostic challenges, identifying reinfections, monitoring appropriate therapy with serological testing and formulating a correct approach to asymptomatic neurosyphilis.[1]

Also, most of the studies from India involve only one group of patients i.e., those reporting at health care centres for treatment and hence, may not be representative of the true situation in the community.[1]

Tertiary syphilis is a rare entity in present antibiotic era. It may manifest in nose as superficial or deep ulcerations and granuloma. Granuloma may involve any part of the nose. Saddle nose deformity that is characteristic of syphilis results from the destruction of the bony framework of the nose and the shrinking of fibroid tissue. The diagnosis of syphilis usually reached after carefully ruling out all other clinical prospects and confirming treponemal infection by laboratory analysis. Syphilis of the nose is regarded by all authorities to be very rare.[3] One such case is presented here for its rarity.

CASE REPORT

A 76 years old presented with complaints of recurrent epistaxis since 01 month. Patient had a history of previous nasal surgery for bilateral nasal obstruction 02 months back. The patient, however, denied any complaint of trauma to nose, cough, chest pain, dyspnoea, hematuria, sexual promiscuity, anaesthetic skin patches, raised lesions on skin, joint pains, redness or swelling of pinna, mouth or genital ulcers, redness of eyes, diabetes, hypertension or weight loss.

His vital signs were found to be normal on nasal endoscopy excessive crusting in both nasal cavity and septal perforation involving both cartilaginous and bony septum was evident (Figure 1). Throat examination revealed a mass covered with slough extending from nasopharynx into oropharynx, involving left lateral pharyngeal wall and soft palate, palatal movements were normal (Figure 2). Orbital ridges, eyeballs, and rest of the face were normal.

Blood counts, liver functional tests, renal function test, pANCA, cANCA, HIV, HBsAg, Anti HCV, routine and microscopic examination of urine, Mantoux test, chest radiograph were normal. CSF examination revealed negative VDRL reaction and normal biochemical parameters. Echocardiography showed normal study. C-reactive protein turned out to be positive, suggesting an inflammatory pathology. CECT PNS was suggestive of post operative status with pansinusitis. Nasal biopsy suggestive of acute inflammation were inconclusive.

Diagnosis was clenched when VDRL came as reactive and subsequently serum TPHA and serum FTA-ABS came positive. The patient was treated in lines of tertiary syphilis with 2.4 million units of intramuscular injection of benzathine. Penicillin every week for 3 weeks and simultaneous surgical debridement. Histopathological report of debrided tissue confirmed the lesion suggestive of non caseating granulomatous lesion (Figure 3). Presently patient is under regular follow up and shown signs of improvement (Figure 4). He is presently using local nasal douching with normal saline and baking soda with good benefit. His symptoms of nasal discharge and crusting in nasal cavity are well under control.

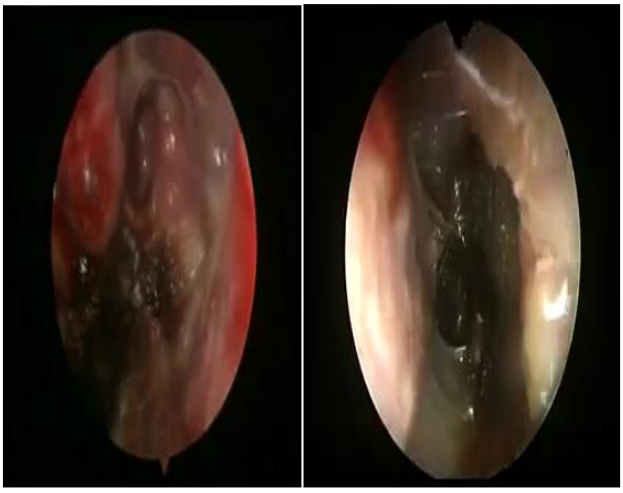

Figure 1: Extensive crusting involving bilateral nasal cavities.

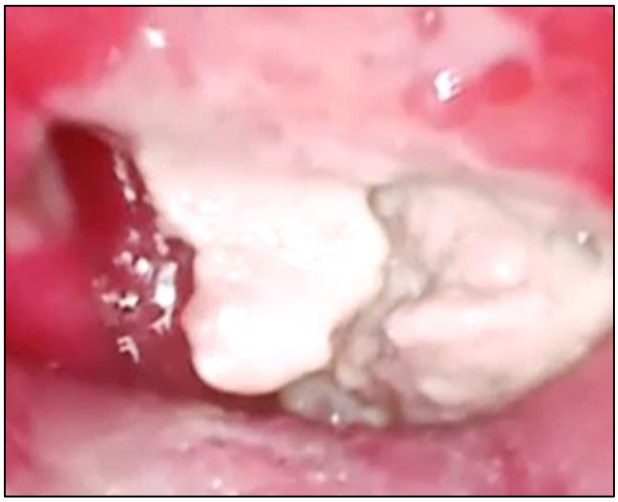

Figure 2: Mass involving left lateral pharngeal wall and soft palate.

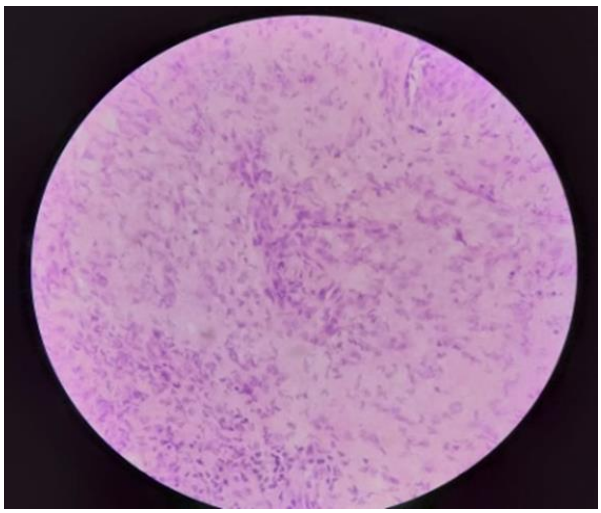

Figure 3: HPR suggestive of chronic granulomatous disease.

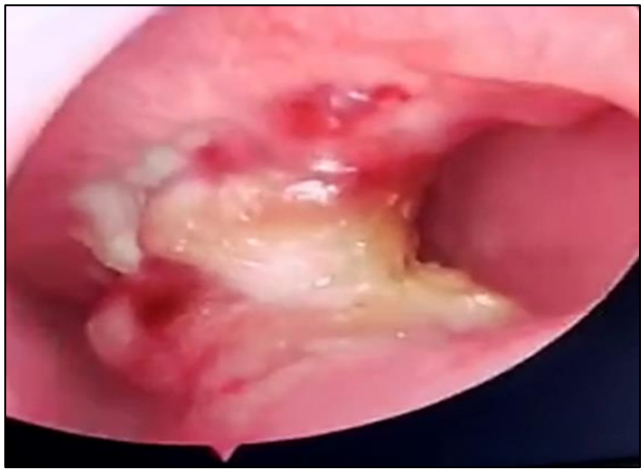

Figure 4: Post op crusting reduced, septal perforation involving cartilagenous and bony septum.

DISCUSSION

Syphilis is a systemic disease caused by the spirochete TP.[4] It occurs exclusively in humans; there is no animal reservoir.[5] Approximately 90% of all syphilis is sexually transmitted during oral, anal or vaginal intercourse. Transmission occurs through direct contact with infectious exudates from moist skin lesions or mucus membranes of infected persons during sexual contact.[6]

The disease is classified as congenital or acquired. Congenital syphilis is divided into early (first 2 years) and late, including stigmata of congenital syphilis. Acquired syphilis is divided into early and late. The European centre for disease prevention and control (ECDC) defines early syphilis as syphilis acquired <1 year previously and world health organization (WHO) as syphilis acquired <2 years previously. Early syphilis includes primary, secondary and early latent syphilis. Late syphilis includes late latent and tertiary syphilis (gummatous, cardiovascular and neurosyphilis). The ECDC defines late syphilis as syphilis acquired >1 year previously and the WHO defines it as syphilis acquired >2 years previously.[7,8] All stages of syphilis may manifest with head and neck findings.[9]

Tertiary syphilis shows most marked manifestations in nose, causing superficial and deep ulcerations, and gumma. Gummatous deposit may occur in any portion of the nose. The most frequent site is the septum and floor of the cavity. It commences most frequently in the submucous tissues, extending both to the surface and the deeper tissues with subsequent degeneration, resulting in superficial or deep ulcerations. The periosteum or perichondrium becomes involved, and later there is necrosis of the bony structures. The septum is a frequent site of pathology, especially the junction of the cartilaginous with the bony septum, resulting in perforation. Where the bony septum is involved, existence of syphilis is unquestionable.

The deformity resulting from destruction of bony framework of nose and shrinking of fibroid tissue produces the typical saddle nose which is characteristic of syphilis.[10] Certain manifestations like interstitial keratitis and Clutton's joints are due to hypersensitivity and not because of the direct effects of the organism. In our patient the septal perforation was due to chronic necrosis by the localized gumma.[11]

Co-infection of syphilis and HIV is common as both are sexually transmitted infections. Syphilis can enhance the acquisition of HIV. Syphilis in the HIV-infected individual can be highly aggressive. Patients can progress from primary to tertiary syphilis over several years, as opposed to several decades in individuals not infected with HIV. They are at increased risk to manifest a more protracted and malignant course which includes more constitutional symptoms, greater organ involvement, atypical and florid skin rashes, multiple genital ulcers, concomitant chancre during the second stage, and a significant predisposition to develop symptomatic neurosyphilis, especially uveitis.[10]

Patients suspected of having syphilis are usually screened with nontreponemal tests, including the venereal disease research laboratory (VDRL) test.[12] Although the chancre may develop within one week of exposure, IgM antibodies take 2 to 3 weeks to be detectable during which time patients may have negative nontreponemal tests.[13] During this gap, dark-field microscopy is an invaluable tool for directly visualizing pathogens from chancre fluid; however, this method requires special equipment and experienced technicians. Patients with a positive VDRL test should undergo specific treponemal testing, such as the fluorescent treponemal antibody absorption (FTA-ABS) assay or the T. pallidum particle agglutination (TPHA) test to confirm infection with T. pallidum. Persons with confirmed syphilis should always be tested for HIV.

The characteristic lesion of tertiary syphilis on histopathological examination is 'gumma', which is characterized by nodules of plasma cells, lymphocytes, epithelioid cells and fibroblasts. Perivascular cuffing by these cells and endarteritis will cause a reduction in the lumen of blood vessels causing necrosis and ulceration.

The treatment plan for syphilis remains relatively unchanged in recent years and continues to vary with stage of infection. Primary, secondary, and early latent syphilis can be treated with a single intramuscular dose of 2.4 million units of benzathine penicillin. A longer treatment course of 2.4 million units of intramuscular Benzathine penicillin every week for three weeks is recommended for late latent syphilis, for tertiary syphilis, or if infection duration is unknown. Neurosyphilis requires 3 to 4 million units of intravenous aqueous crystalline penicillin G every four hours for 10 to 14 days.[14] Local treatment consists of clearance of crusts and regular cleansing of the nasal passages by copious

alkaline douches one to three times a day. Yellow mercury oxide ointment may be applied locally. The purpose of local treatment is to remove the discharge and crusts, kill spirochetes, promote wound healing and epithelial growth, prevent secondary infection. Gumma responds rapidly to general antisyphilitic treatment, but atrophic rhinitis and deformity may persist after the disease is cured and this may need further reconstructive surgery.[15]

CONCLUSION

Tertiary syphilis is rarely seen these days; but collapsed nasal bridge with destruction of nasal septum and turbinates even without clear history of genital sore or lesions of secondary syphilis in the past should evoke high index of suspicion. Diagnosis can be made by characteristic organ involvement, histopathological picture, TPHA test being positive in spite of VDRL being negative and by response to adequate antibiotic therapy. It is very important to rule out other possible disease pathologies such as tuberculosis, lupus vulgaris, sarcoidosis, yaws, atrophic rhinitis, leprosy, scleroma, chronic glanders, leishmaniasis and benign or malignant neoplasm.

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