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RESEARCH ARTICLE

RHINOSCLEROMA - ARARE CASE REPORT WITH SINUS INVOLVEMENT

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Abstract

Rhinoscleroma is a rare chronic infection with granuloma formation which is precipitated due to recurrent chronic sinusitis. Clinically patient presents with mass or swelling in upper airways most commonly in nose with obstructive features. Though there are many tests for its diagnostic evaluation, histology still holds the mainstream modality for its confirmatory diagnosis.

Introduction:-

Rhinoscleroma is one of the gradually developing, rare scleromascausing chronic granulomatous infection found to be endemic in areas like India, Middle East, Africa, Indonesia. The causative pathogen *Klebsiella rhinoscleromatis* (subspecies of *k.pneumonae*), is a gram negative bacilli usually affecting the upper airways along with nasal tract.[14-15] Species of *klebsiella* is in its abundance in water, plants and soil but it has been only seem to affect humans.[16] *Klebsiella* comes under the subgroup of KES of *Enterobacteriaceae*family of which *K. rhinoscleromatis* can be differentiated from other sub species via various tests like methyl red positivity, urease and citrate reductase negativity.[17]The disease course is insidious in onset which affects most parts of respiratory system beginning from nasal sub epithelium, with slowly spreading down to pharyngeal subepithelial region and further to Eustachian tube, antrum of the maxilla, oral cavity, larynx, trachea, bronchi. It transmits via airborne route with its secretions; however, immunocompetent individuals remainunaffected. This disease progression can be stated into three stages as, stage 1 or catarrhal stage or stage of atrophy, stage 2 or proliferative stage or stage of granuloma formation and stage 3 or fibrotic or the stage of sclerosis. The histological diagnosis remains the mainstay of diagnosis. Also, it can support by a bacterial culture of the above histological biopsy specimen. [17-18]

Case Report:

A 25- year old male patient reported to the Department of Oral and Maxillofacial Surgery, Dental College, RIMS with the chief complaint of swelling and pus discharge from buccal vestibule in the upper left back teeth region associated with painin the past 2 weeks from the date of reporting. Solitary diffuse swelling obliterating buccal vestibule from the left 1st premolar region to 2nd molar region. The swelling was gradual in onset, moderate and has progressed to the present state. The pain was sudden in onset, moderate in intensity, throbbing type, radiating to ears, aggravated on eating and relieved by medication. No relevant medical history. He gave history of root canal treatment irt (24,25,26) 2 years back and undergone extraction irt (28) 1 year back with no associated complications. He also gave history of smoking tobacco (2-3 sticks/day) for 3 years.

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General examination did not reveal any deviation from the routine findings. Extraoral swelling on the left side of face, 2-3 cm from the lateral side of nose.

Intraorally, the lesion presented as a diffuseswelling involving the left side extendingfrom the buccal vestibule corresponding toleft 1st premolar up to 1st molar. The swellingwas grossly 25×43×46 mm in size withregular, diffuse margin, andthe overlying surface was smooth, pus like discharge andof the same color as the adjacent palatalmucosa. On palpation, the swellingwas nontender and soft to firm in consistency.

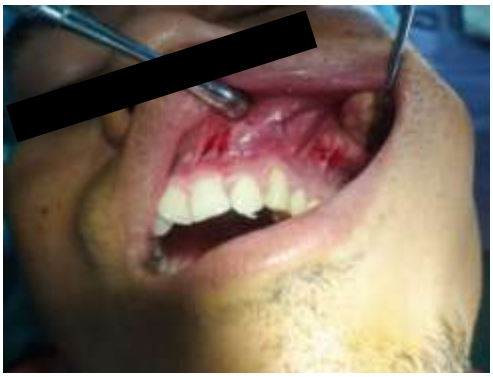


Figure 1:- Diffuse swelling obliterating buccalvestibule

Based on the history and clinical findings, a provisional diagnosis of diffuse swelling obliterating buccal vestibule of Rhinoscleroma was made.

Minor salivary gland tumor, radicular cyst, and periapical abscess were considered in differential diagnosis. Subsequently, radicular cyst and periapical abscess were ruled out the due absence of any dental involvement.

Investigations

Intraoral periapical radiograph reveals diffuse radiolucency in the periapical region of 24,25,26.

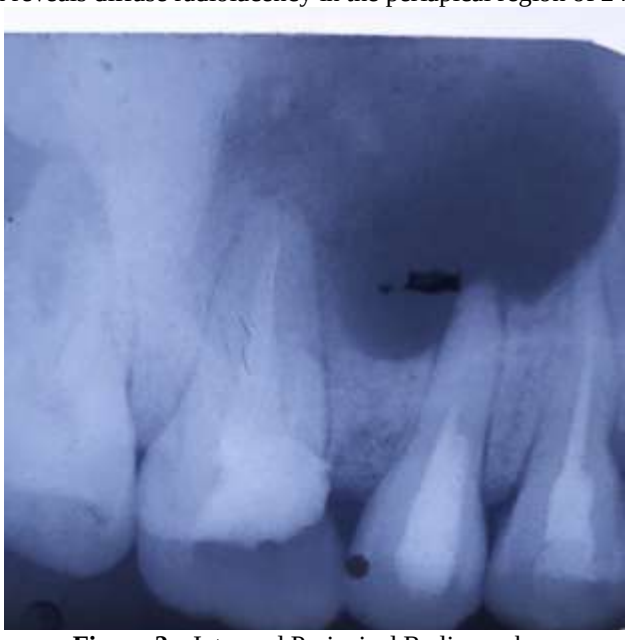
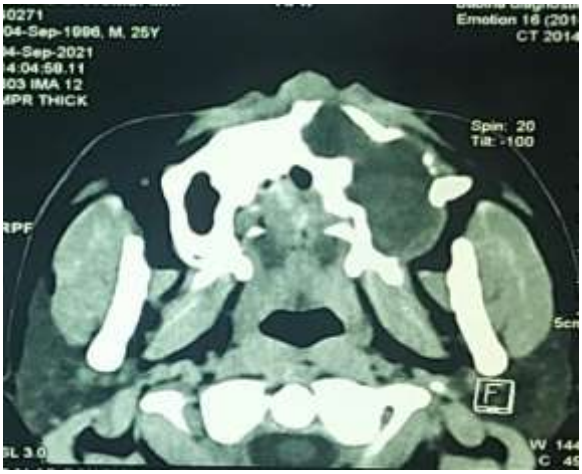
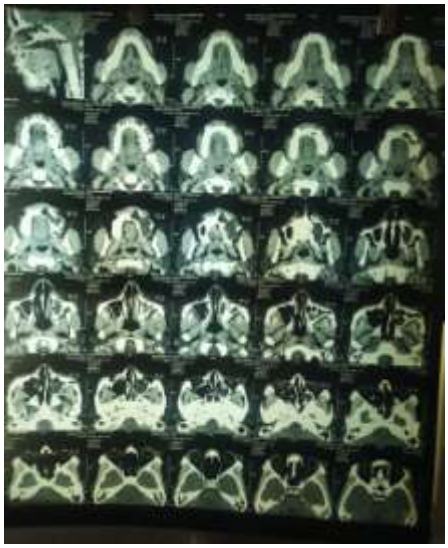


Figure 2:- Intraoral Periapical Radiograph.

CT face reveals expansile lytic (cystic) lesion in left maxilla. The roots of left incisors are affected. Antero superiorly and laterally it is causing bony erosion in the anterior wall of maxillary sinus. Complete opacification of left maxillary sinusis noted.





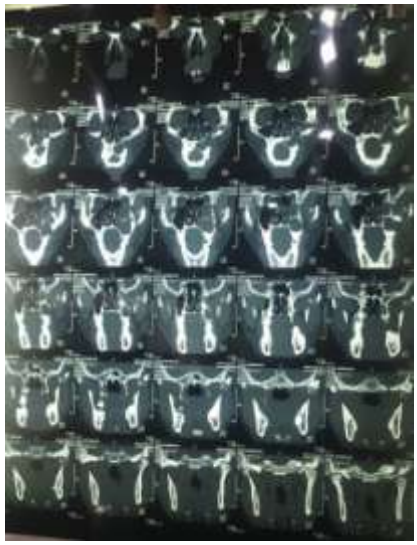


Figure 3, 4 & 5:- CT Scan of face.

Figure 6:- Specimen.

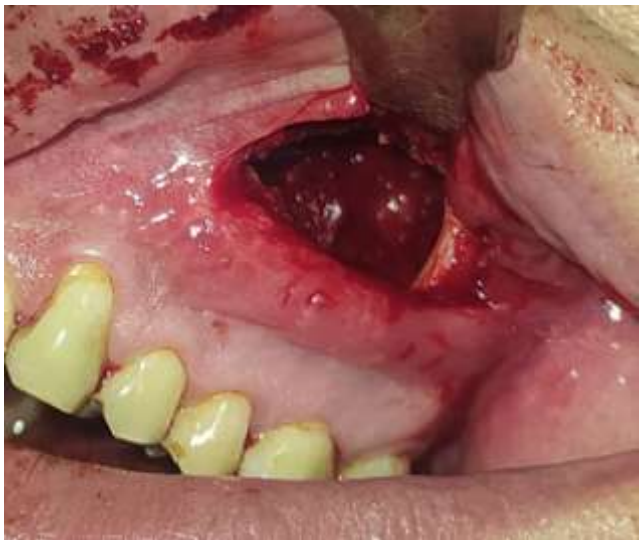


Figure 2:- Caldwell- Luc procedure.

Hematological investigation suggests mild eosinophilia.
Biochemical investigation reveals total protein: 8.5 gms% by Biuret method, globulin: 4.1 gms%

Cytological evaluation of the aspirate revealed clumps of red blood cells and few inflammatory cells.

Histopathological examination reveals multiple sections studied from the tissue pieces from left maxillary sinus show mostly ulcerated epithelial lining with a few foci lined by metaplastic stratified squamous epithelium. In subepithelial area, there is marked infiltration by lymphoplasmacytic cells with plasma cells with plasma cells arranged in sheet, foamy histocytes (Mikulicz cells), focal areas of hemorrhagic necrosis and sclerosis. Periodic acid- Schiff (PAS) stain for bacilli in the foamy histocytes is negative.

Based on the histopathological findings, the diagnosis was given in favor of Rhinoscleroma.
The lesion was surgically curettage from the left maxillary sinus under local anesthesia(Caldwell- Luc procedure) followed by antibiotics therapy including cefoperazone and sulbactam (1.5 gm) twice daily for 5 days, metronidazole (400mg) thrice daily for 5 days. For steam inhalation, decongestant and anti- inflammatory drugs was given.The patient is kept under regular observation to monitor any incidence of recurrence.

Discussion:-

RS or scleroma is a rare, chronic, indolent, granulomatous, and debilitating bacterial disease primarily involving the nose and is designated as respiratory scleroma when it involves sub glottis, trachea, and bronchi.[4] Its notoriety in involving other neighboring sites such as nasopharynx, eustachian tube, paranasal sinuses, soft tissues of lips and nose, and rarely orbit is well established. The current case is one such example of extranasal RS. Anton Von Frisch in the year 1882 is credited with the identification of the causative organism *K.rhinoscleromatis*, a Gram- negative aerobic bacteria belonging to the *Klebsiella pneumoniae* family. [5,6] RS has female predilection (13:1) and usually occurs in the middle- aged population. Low socioeconomic status, malnutrition, and iron deficiency anemia are considered as the predisposing factors that lower cellular immunity while preserving humoral immunity.[2- 4] The presence of HLA- DQA103011- DQB0301 haplotype has been found to be a strong risk factor.[7] Clinically and pathologically, RS is characterized by three overlapping phases in terms of progression: rhinitic, florid, and fibrotic. The rhinitic/catarrhal stage causes earliest symptoms of nonspecific rhinitis which are known to last for weeks or months and often evolves into purulent and fetid rhinorrhea with crusting. Histologically, squamous metaplasia along with subepithelial infiltrate of polymorphonuclear cells is usually observed. Bacteria are incompletely digested in the subepithelial layer and further released into tissues. The second florid/granulomatousstage is characterized by the development of bluish- rednasal mucosa and intranasal rubbery nodules or polyps andmanifests with epistaxis and nasal deformity, destruction ofthe nasal cartilage, and bone destruction. In advanced cases,the destruction of the nasal cartilage with the formation ofnodules causes a severe deformity referred to as Hebranose. Histology shows the appearance of Mikulicz cellsthat are pathognomic of RS. [8,9]Interleukin- 10, an

anti- inflammatory cytokine, has beendemonstrated as being crucial for the establishmentof a proper environment leading to the phenotypicmaturation of Mikulicz cells. It controls the metabolicreprogramming of macrophages through inhibition ofmechanistic target of rapamycin (mTOR) signaling pathway.Deregulation of mTOR signaling may lead to metabolicchanges such as hyperproliferation of macrophages andgranuloma formation which in turn will contribute to diseaseprogression in human granulomatous inflammations.[9,10]The last phase is the sclerotic/fibrotic stage that ischaracterized by formation of adhesions, extensive fibrosisleading to scarring, and possible nasal stenosis which willultimately result in distortion of anatomy.[8,9]In our case, the patient reported with a palatal swelling andinvolvement of the posterior part of the oral cavity whichdiffered strikingly from its usual manifestation like nodularenlargement observed in the lower nasal and upper labialregion.

In 1969, Mikulicz and Woyke et al. described themicroscopic picture of the disease along with the firstdescription of the ultrastructure of granulation tissue inRS.[3] Microscopic deduction shows an initial infiltrationof neutrophils, plasma cells, and macrophages which isfollowed by granulomatous inflammation, characterized bynumerous plasma cells along with Russell bodies and MCs.Russell bodies, which represent excess immunoglobulinsproduced by the plasma cells, are round to ovoid,homogeneous, eosinophilic, and up to 40 um in diameter.MCs are large phagocytes with a single shrunken nucleus,displaced to the cell periphery because of the presenceof multibacillary cytoplasmic vacuoles, 10–100 um indiameter.[7,11,12] In adjunction to the routine histopathology,various special stains have also been employed in detectionof the *K. rhinoscleromatis*, these include PAS stain thatstains the bacilli pink, Giemsa stain which gives paleblue color to the organism and Warthin–Starry silverstain which imparts a distinct black color to the bacillimaking it more prominent as compared to the previouslydiscussed stains. Recent advances in the diagnosis, inconjunction with the existing methods, include serotypingand immunohistochemistry. De Pontual et al. in theirstudy noticed seropositivity in culture for O2:K3 type of*Klebsiella*.[6]

Among the wide spectrum of nasal and palatal lesions, thedifferential diagnosis of RS may include:
1. Fungal infections such as histoplasmosis,blastomycosis,and rhinosporidiosis
2. Bacterial infections – tuberculosis, leprosy, and syphilis
3. Noninfectious conditions – Wegener's granulomatosis,natural killer cell lymphoma, lethal midline granulomaetc. [13]

The fungal etiology may be ruled out by the negativeculture of the causative organisms as well as by applyingthe special stains such as PAS, Gomori Silver Methenamineto demonstrate the fungal hyphae. Presence of acid–fastbacilli is the diagnostic feature of other granulomatousinfectious diseases such as Tuberculosis and leprosy. Efaredet al. in 2017 applied immunochemical histiocyte stainingfor CD68 to rule out the diagnosis of certain lymphomas ormalignancies which may mimic the morphology of RS.[5]Routinely followed treatment for management of RSincludes surgical correction and antimicrobial treatmentwith ciprofloxacin, streptomycin, rifampicin, sulfonamides,clofazimine, or fluoroquinolones.[7] Treatment must becontinued for months, which frequently leads to poorpatient compliance. Recurrence is quite common in casesof RS and its chronicity also plays a pivotal role inunsatisfactory prognosis of the disease.

Conclusion:-

RS usually presents with very deceptive clinical featureswhich often may lead to its misdiagnosis. This reportpresents one such incidence of this disease with widespreadinvolvement including oral cavity. In this scenario,histopathology has proven to be an excellent aid in clearingthe dilemma surrounding the diagnosis of the condition.

Currently, newer advances in the diagnosis by serotypingand immunohistochemistry are being explored which willlead to more prompt diagnosis thus resulting in better prognosis.

Declaration of patient consent:

The authors certify that they have obtained all appropriatepatient consent forms. In the form, the patient has given hisconsent for his images and other clinical information to bereported in the journal. The patient understands that name andinitials will not be published and due efforts will be made toconceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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