European Journal of Molecular & Clinical Medicine
ISSN 2515-8260 Volume 7, Issue 2, 2020

RHINOSCLEROMA – A RARE CASE REPORT

Dr. Anup Kediya[1], Dr. Anil Agrawal[2], Dr. Aanchal Manchanda[3], Dr. Sunita Vagha[4]

[1,2,3,4]*Department of Pathology, Jawaharlal Nehru Medical College, Datta Meghe Institute of Medical Sciences (Deemed to be University), Sawangi Meghe, Wardha*

Dr Anil Agrawal, Email: dranilagrawal.456@gmail.com , 9850309278

Conflict of Interest: None

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.

Keywords: Rhinoscleroma, chronic, granulomatous, Mickulicz cells.

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.[1-2] Species of klebsiella is in its abundance in water, plants and soil but it has been only seem to affect humans.[3] Klebsiella comes under the subgroup of KES of Enterobacteriacae family of which K. rhinoscleromatis can be differentiated from other sub species via various tests like methyl red positivity, urease and citrate reductase negativity.[4] The disease course is insidious in onset which affects most parts of respiratory system beginning from nasal subepithelium, 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, immunocompetant individuals remains unaffected. 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 supported by a bacterial culture of the above histological biopsy specimen.[4-5]

CASE REPORT

A 54 year-old female presented with right sided nasal obstruction for last 3 months with history of similar complaints for past 6 months. He also complained of persistent headache, swelling on cheek, eye and nasal bleeding while blowing. Patient also had a past history of purulent nasal

discharge. On local examination, mild proptosis in the right eye with nasal deviation to right side was noticed. Anterior rhinoscopy showed right nasal cavity mass which was polypoidal and vascular in nature arising from the lateral wall and pushing septum to the left. Hematological and biochemical investigations was in normal limits. On CT scan, right nasal mass compressing the right maxillary sinus was seen. FNAC was done which showed the presence of foamy macrophages in a background of lymphocytes and plasma cells. Stains for acid fast bacilli(AFB), Grocott's silver methenamine were negative. An occasional gram negative bacillus within the macrophage was seen on gram's stain. Histology for the following excised mass was done to rule out rhinoscleroma.

Histopathological examination showed hyperplasia of the epithelium. The zone beneath it was densely infiltrated by chronic inflammatory cells consisting of lymphocytes, plasma cells, and groups of large vacuolated histiocytes with single or multiple cytoplasmic vacuoles and small centrally or peripherally located nuclei(Mikulicz cells) was seen. Nuclear chromatin was fine with indistinct nucleoli. An occasional cell showed intracellular
gram negative bacilli. Russell bodies were also present. An ill-formed noncaseating granuloma comprising of epithelioid cells and lymphocytes were seen. Microbiological culture showed the exact causative microorganism (*Klebsiella rhinoscleromatis*).

Patient was put on the antibiotics for 1 month after which surgical resection of the mass was done followed by course of antibiotics comprising rifampicin and levofloxacin for 3 months.

RESULTS:

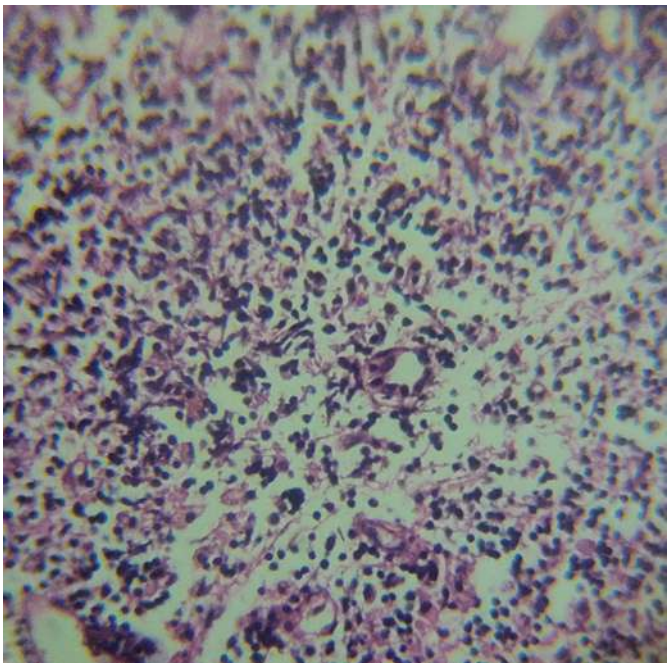

Figure shows a hyperplastic mucosal epithelium with dense, diffuse infiltrate consisting predominantly of plasma cells with many large histiocytes Giant histiocytes with vacuolated cytoplasm (Mikulicz cells) containing gram-negative rods in the cytoplasm.

DISCUSSION

Rhinoscleroma is a rare chronic granulomatous bacterial infection having an insidious onset and an indolent course usually involving the nose and less commonly involve other areas of upper airway. It has a female preponderance affecting in older age groups between 3rd to 6th decade. Initially it presents as a recurrent chronic rhinitis with nasal crustations, which if ill-treated goes under frequent remissions and rarely a slowly growing nasal mass. Predisposing factors can be immunocompromised state of the patient as it it doesn't affect healthy individuals.[1-3]Clinically, patient can present with any of the three stages of disease progression. First stage being the catarrhal stage which is associated with recurrent sinusitis, running nose lasting for weeks or months where there is polymorphonuclear leukocytic infiltration with granulation tissue. Second stage being the granulomatous phase which is diagnostic phase of rhinoscleroma characterized by formation of mass and tissue destruction. Histologically, this stage comprises of dense lymphocytes, plasma cells, Russell bodies and Mikulicz cells which is pathognomic finding with foamy macrophages and cytoplasmic vacuoles containing klebsiella bacilli within it. Last stage being the sclerotic phase where there is extensive sclerosis and fibrosis with less chronic inflammatory cells and few to occasional Mikulicz cells seen. [1,5,6] Although histology holds the gold standard diagnostic modality, stains such as Periodic Acid Schiff (PAS), Geimsa, Gram's stain significantly shows presence of klebsiella bacilli in it. Mikulicz cells are the cells having bacilli within the clear cytoplasmic vacuoles.[4,5] Bhowate et al reported a case of Rhinoscleroma with involvement of the maxillary sinus, orbital floor, and temporomandibular joint[6]. Various studies on related granulomatous lesions of sino-nasal region have been reported by Choudhary et al [7], Kandukuri et al [8,9] and Jain et al[10,11].

Treatment modality consists of intensive antibiotics with different combinations of it mostly effective in catarrhal stage and surgical resection for the later stages where the mass compresses nearby structures leading to complications.

REFERENCES

[1] Umphress B, Raparia K. Rhinoscleroma. Archives of Pathology & Laboratory Medicine. 2018;142(12):1533-1536.
[2] Fawaz S, Tiba M, Salman M, Othman H. Clinical, radiological and pathological study of 88 cases of typical and complicated scleroma. The Clinical Respiratory Journal. 2011;5(2):112-121.
[3] Fevre C, Almeida A, Taront S, Pedron T, Huerre M, Prevost M et al. A novel murine model of rhinoscleroma identifies Mikulicz cells, the disease signature, as IL- 10 dependent derivatives of inflammatory monocytes. EMBO Molecular Medicine. 2013;5(4):516-530.
[4] de Pontual L, Ovetchkine P, Rodriguez D, Grant A, Puel A, Bustamante J et al. Rhinoscleroma: A French National Retrospective Study of Epidemiological and Clinical Features. Clinical Infectious Diseases. 2008;47(11):1396-1402.
[5] Zeinab H El-badawy A. Rhinoscleroma: a detailed histopathological diagnostic insight. Int J Clin Exp Pathol. 2015;8(7):8438-45.
[6] Bhowate, Rahul R., Shirish Degwekar, Shivlal Rawlani, and Surwarna Dangore. "Rhinoscleroma With Involvement of the Maxillary Sinus, Orbital Floor, and

Temporomandibular Joint: A Case Report." *JOURNAL OF ORAL AND MAXILLOFACIAL SURGERY* 70, no. 1 (January 2012): 135–40. https://doi.org/10.1016/j.joms.2011.02.004.

[7] Claude, M. J., L, B., S, N., D, D., S, H., & F, N. (2020). Inhalation of foreign body: "Avoidable lobectomy. A case report''. *Journal of Current Medical Research and Opinion*, *3*(03), 426-429. https://doi.org/10.15520/jcmro.v3i03.263

[8] Choudhary, Sanjiv V., Praveen Khairkar, Adarshlata Singh, and Sumit Gupta. "Dermatitis Artefacta: Keloids and Foreign Body Granuloma Due to Overvalued Ideation of Acupuncture." *INDIAN JOURNAL OF DERMATOLOGY VENEREOLOGY & LEPROLOGY* 75, no. 6 (December 2009): 606–8. https://doi.org/10.4103/0378-6323.57725.

[9] Kandukuri, Rashmi, and Suresh Phatak. "Evaluation of Sinonasal Diseases by Computed Tomography." *JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH* 10, no. 11 (November 2016): TC9–12. https://doi.org/10.7860/JCDR/2016/23197.8826.

[10] Shraddha, Jain, Madhu Priya, and Kumar Sunil. "Mucosal Malignant Melanoma of Sino-Nasal Region with Orbital Involvement: Case Report." *INDIAN JOURNAL OF OTOLARYNGOLOGY AND HEAD & NECK SURGERY* 65, no. 1, SI (July 2013): S178–81. https://doi.org/10.1007/s12070-011-0428-y.

[11] Jain, Shraddha, Pragya Singh, Disha Methwani, and Sanika Kalambe. "Role of Eustachian Dysfunction and Primary Sclerotic Mastoid Pneumatisation Pattern in Aetiology of Squamous Chronic Otitis Media: A Correlative Study." *INDIAN JOURNAL OF OTOLARYNGOLOGY AND HEAD & NECK SURGERY* 71, no. SUPPL 2, 2, SI (November 2019): 1190–96. https://doi.org/10.1007/s12070-018-1259-x.