Rhinoscleroma

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Introduction

Rhinoscleroma is a progressive, granulomatous infectious disease, usually limited to the upper respiratory tract and produced by Klebsiella rhinoscleromatis[1,2]. Sporadic cases and endemic foci of the disease have been found in 68 countries[3,4].
Clinically, the disease has been described in progressive stages; however overlapping of the stages occur frequently in the individual patient[3,5-7]. Other Investigators[8,9] have considered the Mikulicz cell as a macrophage. The foamy mononuclear cell in rhinoscieroma has all the basic morphologic structures characteristic of m acrophages[10].
The typical histopathology consists of plasma cells, lymphocytes, Russell bodies, Mikulicz cells and gram negative bacilli[11]. The Mikulicz cells are foamy vacuolated histiocytes under going hydropic degeneration in different stages[12]. These cells are well stained with hematoxylin eosin. Klebsiella rhinoscieromatis are found inside the Mikulicz cells and inter cellular spaces as gram negative diplobacilli.
Microscopically the tumor shows polypoidal fragments. The subepithelial tissue reveals aggregates of foamy macrophages admixed with plasma cell. Many plasma cells contain fine cytoplasmic vacuoles, and the cytoplasm of others appear more eosinophilic than the amphophilia more commonly observed.

Case Report

We report a case of Rhinoscieroma arising in the nasal cavity of a 60 years old male, presenting at the department of ENT, Jinnah Postgraduate Medical Center.

Clinical Features

The patient presented with history of nasal obstruction since 2 years. The physical and CT scan examination suggested a growth involving the nasopharynx hanging in to the oropharynx and pushing the left maxillary wall.

Pathological Findings

Gross Examination

On gross examination multiple, polypoidal, soft to firm, tan to brown tissue pieces measuring 20m1 in volume, with a single piece of bone was received. The largest piece measuring about 4.5 x 3.2x 2.5 cms.

Figure 1 Gross appearance of the tumour, soft, polypoidal masses measuring 20 ml in volume.

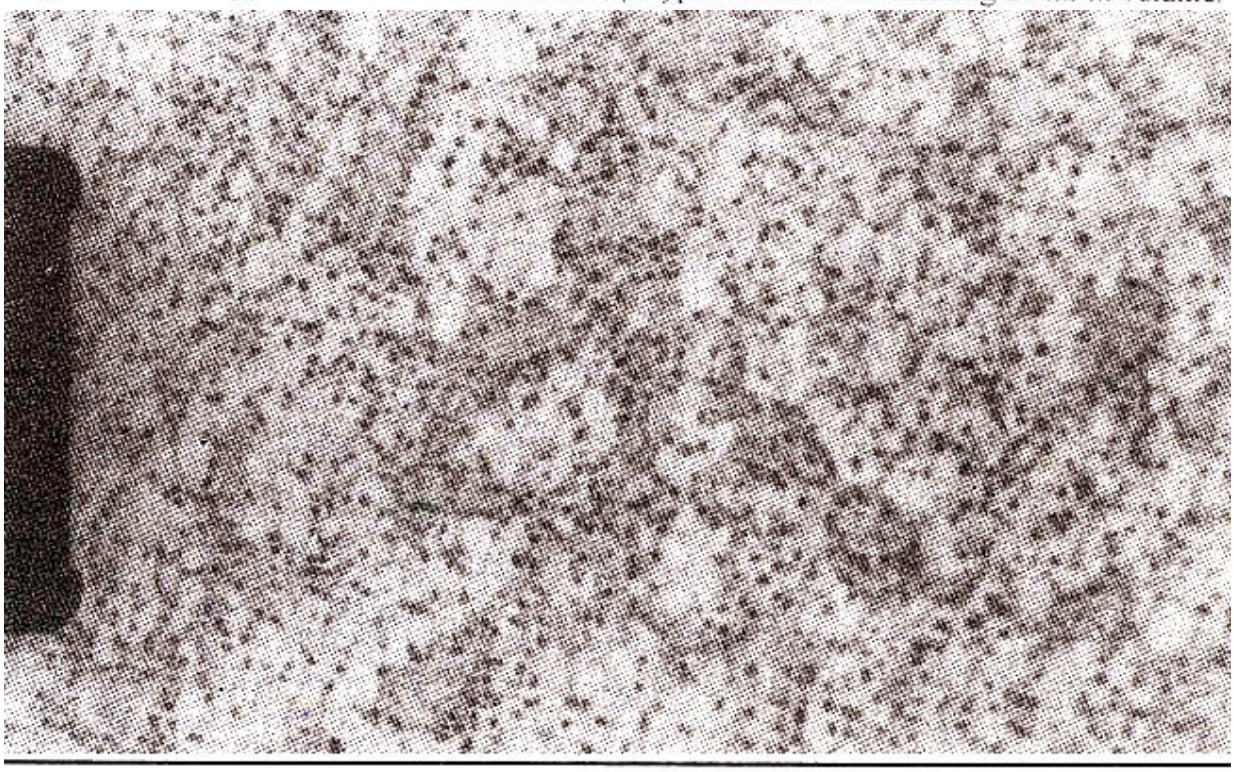

Figure 2 Microphotograph of the tumour showing aggregates of foamy macrophages admixed with plasma cells. (H & E x 200).

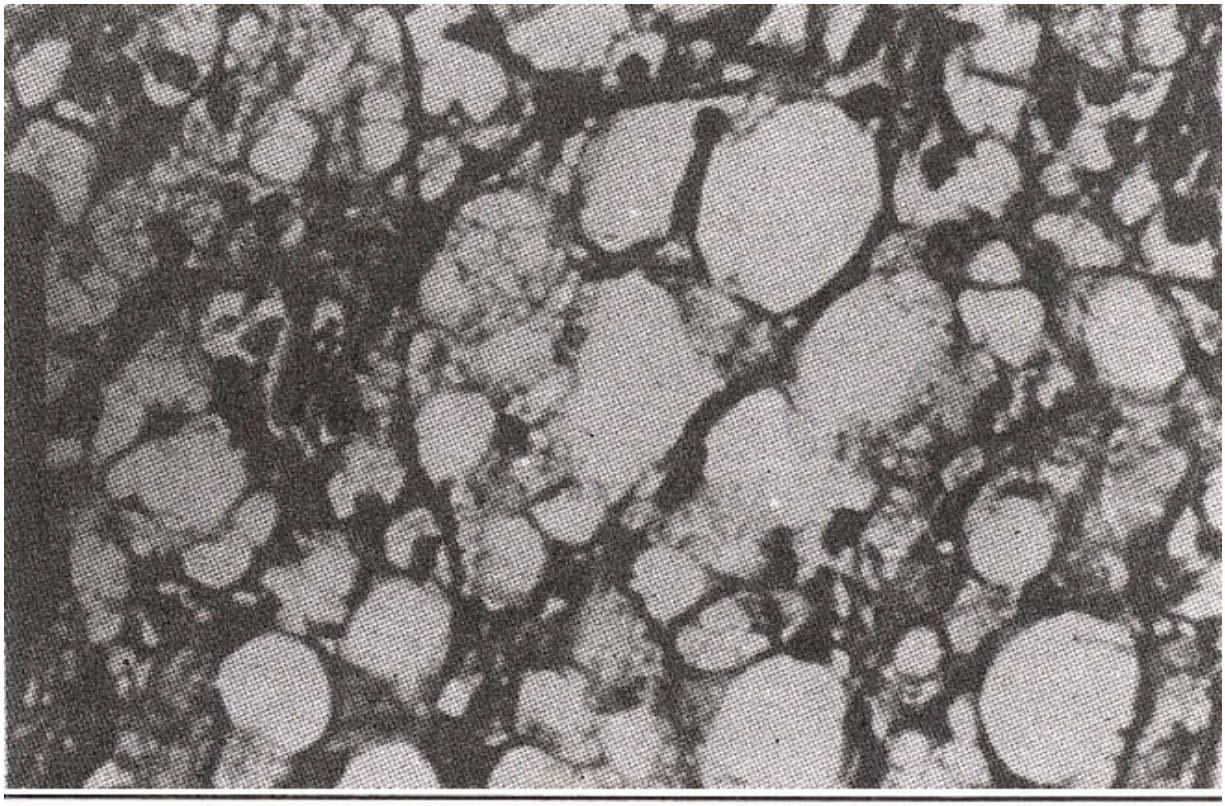

Figure 3. Microphotograph of the tumour revealing foamy macrophages with Gram negative bacilli (Gram's stain x1000).

Microscopic Examination

Microscopically, the tumour showed polypoidal fragments covered by the nasal mucosa. The subepithelial tissue had aggregates of foamy macrophages admixed with plasma cells and neutrophils. Gram's stain revealed gram negative diplobacilli within the macrophages giving a diagnosis of Rhinoscleroma.

Discussion

Rhinoscieroma is a chronic, possibly specific granuloma of benign symptoms at the beginning and characterized by a slow progress. In 1876 Mikulicz[13] published his studies and described the "Swollen cells" for the first time which bear his name.

Scleroma is characterized by granulornatous infiltration of the soft tissues and by the formation of nodules or tubercles, which are present during the late stages of the disease. Cartilage and bone are always spared; originating in the nose, it can involve any part of the respiratory tract[14]. The incidence of nasal extension to the oral cavity and pharynx varies from 18 to 43% in different series.

Clinically, rhicoscieroma begins with rhinorrhea and nasal obstruction. It differs from atrophic rhinitis by presence of granulomatous nodules on the nasal septum, inferior turbinate and floor of the nasal vestibule. These nodules coalesce and fill the nasal cavity. In our case, the patient presented with nasal obstruction since 2 years. The lesion was polypoidal, soft, brown measuring 20ml in volume.

The microscopic findings show almost three diagnostic elements. The Mikulicz cells, the Russell

bodies and bacillus Klebsiella rhinoscleromatis. The light microscopic feature in our case included aggregates of foamy macrophages admixed with plasma cells and neutrophils.
While the patient is immunologically competent, the pathogenesis may lie in the interaction between the Mikulicz cells and the intracellular Kiebsiella rhinoscleromatis. The cells are histiocytes in different stages of phagocytosis. In acute inflammation, numerous bacilli each with a slime coating were found in phagosomes of the Mikulicz cells. This non-digestible mucopolysaccharide material is important in the pathogenesis of the disease[12]. It increases the osmotic pressure and forms multiple hydropic vacuoles that rupture both the phagosomes and the Mikulicz cell. Further, the premature rupture of the Mikulicz cell allows liberation of some morphologically viable bacilli[15]. This ineffective destruction allows the infectious process to continue.
Rhinoscieroma can be mistaken for tuberculosis, leprosy rhinosporidiosis, inflammatory pseudotumor. In leprosy mycobacterium leprae proliferate in the upper respiratory passages from the nares to the larynx, with extensive infiltrations of M. Ieprae-Iaden histiocytes in the submucosa.
Rhinosporidiosis is a chronic inflammatory disease produced by Rhinosporidium seeberi, a spore producing fungus that has never been cultured[16], characterized by hyperplastic polypoidal lesion of the nasal cavity. Histological examination reveals thick walled sacs or sporangia containing numerous spores.
Inflammatory pseudotumors are tumor like lesions, morphologically heterogenous consisting of edematous or hyalinized fibrous tissue infiltrated with varying numbers of plasma cells, Russell bodies and lymphocytes.
Tuberculosis can involve the nasal cavity, nasopharynx or paranasal sinuses. Many of the cases are associated with cervical lymphadenopathy[17].
To our knowledge no case of rhinoscleroma has been reported. This is the first case reported from our center in the last ten years.

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