

RHINOSCLEROMA: A CASE REPORT

KATARZYNA KARPIŃSKA-KACZMARCZYK, MARIA CHOSIA, STANISŁAW WOYKE

Department of Pathology, Pomeranian Medical University, Szczecin

Rhinoscleroma is a chronic inflammatory disease in which granulation tissue with a typical cell content is found. The paper presents the case of a 77-year-old woman with clinically diagnosed nodule in the nasal cavity. The histopathological examination revealed granulation tissue with plasma cells and Mikulicz's cells. The clinical and morphological picture of the case in question is a rare opportunity to bring to mind a disease that used to be common in Poland and which clinically can imitate malignant tumour.

Key words: rhinoscleroma, *Klebsiella rhinoscleromatis*.

Introduction

Although rhinoscleroma is nowadays a very rare disease in Poland, a hundred years ago both Poland and the rest of Eastern Europe had the highest incidence of the illness. There are no epidemiological data on rhinoscleroma incidence in Poland. In France, there were 11 reported cases over a period of 15 years (1990-2005) [1]. Presently, it is endemic in some regions of Asia, Africa and South and Latin America. The endemic regions are characterized by common environmental factors, such as low level of hygiene, malnutrition and overcrowded housing. It most commonly affects young and middle age people, more often women than men [2, 3]. The clinical symptoms of rhinoscleroma of the upper respiratory tract were first presented by Ferdinand von Hebra in 1870. Six years later, Johann von Mikulicz described the microscopic picture of the disease [4]. The first description of ultrastructure of granulation tissue in rhinoscleroma was reported by S. Woyke *et al.* in 1969 [4]. In 1882, Anton von Frisch discovered *Klebsiella rhinoscleromatis* as the possible etiological factor of the disease [4-6]. *Klebsiella rhinoscleromatis* is Gram-negative, nonmotile bacteria, which is the subspecies of *Klebsiella pneumoniae* [1, 7]. The name of the disease can be derived from the Greek term *skleroma* and denotes a hard nodular change.

The inflammatory process usually starts in the nasal cavity and then it spreads on to nostrils, pharynx and larynx [3]. There are also cases of rhinoscleroma of the

lower respiratory tract, maxillary sinuses, orbital cavity, lacrimal ducts and cervical lymph nodes [3, 8-10]. The diagnosis can be established on the basis of histopathological, bacteriological and genetic examinations [7]. The disease can be divided into three morphological stages: catarrhal/atrophic, granulomatous or proliferative and sclerotic. The disease is progressive in character. It starts with acute exudative inflammation of the nasal mucous membrane with serous or mucus secretion. At the second stage characteristic granulation tissue is seen with nodules in the nasal cavity, which clinically can imitate cancerous tumours. Deformities of the tip of the nose are sometimes observed. At the third stage inflammatory infiltrations are replaced by connective tissue with thick cicatricial strips. This leads to deformities of nostrils, which is reflected in the old vernacular names of the disease in Polish: syphilis of the nose, leprosy of the nose and Slavic leprosy [1-3, 5, 6, 9, 11].

The microscopic image is very characteristic of this uncommon inflammatory illness.

Case description

A 77-year-old woman was admitted to the Out-patient Laryngological Clinic with nasal atresia. Under examination, a nodule in the right nasal cavity was found, from which five biopsies (2–5 mm in diameter) were taken for histological examination. The specimens were stained with haematoxylin and eosine and PAS. Immunohistochemistry with CD68,

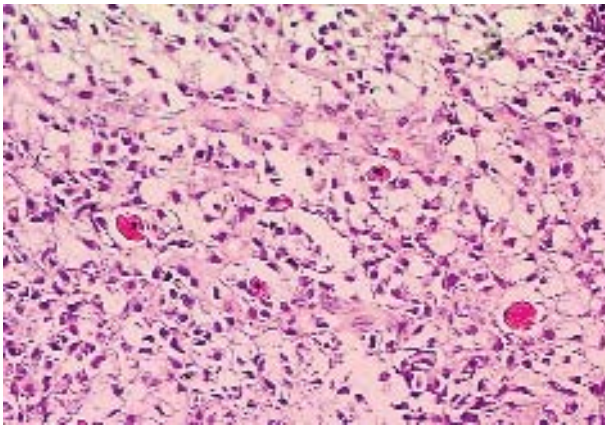


Fig. 1. Plasmacytes with visible traits of hyaline degeneration in the form of Cornil cells and Russell bodies 10×

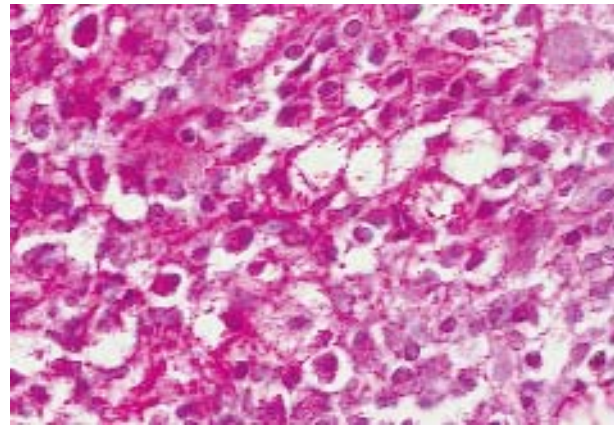


Fig. 2. The cytoplasm of Mikulicz's cells contains PAS-positive inclusion bodies, which correspond to *Klebsiella rhinoscleromatis* 20×

CD138 and plasma-cell antibodies was used. Microscopically granulation tissue composed of large macrophages with light, foamy cytoplasm known as Mikulicz's cells, plasma cells and lymphocytes was found. Plasma cells, more numerous than lymphocytes and Mikulicz's cells, were evenly distributed with a slight tendency to accumulate around small blood vessels. Apart from that, plasmacytes with visible traits of hyaline degeneration in the form of Cornil cells and Russell bodies were seen (Fig. 1). Small neutrophil aggregates were also found mainly in the lumen and around small blood vessels. Glassy, eosinophilic ball-like bodies that can be seen in Cornil cells as well as Russell bodies released from broken cells can be related to containers of rough endoplasmic reticulum, that can be seen in the electron microscope. They are filled with electron thick substance [12]. The cytoplasm of Mikulicz's cells contained PAS positive inclusion bodies, which corresponded to *Klebsiella rhinoscleromatis* (Fig. 2). CD 68 expression was seen in the cytoplasm of Mikulicz's cells. Plasma cells revealed CD138 expression.

Discussion

In the development of rhinoscleroma granulation tissue immunological response plays the major role. Owing to the presence of polysaccharide cell wall, the bacteria effectively inactivate phagocytosis and induce chronic inflammatory process with formation of granulomatous tissue.

Clinical differential diagnosis of the second and third stages of rhinoscleroma involves all destructive lesions associated with granulomas and ulcer formation, including sarcoidosis, Wegener's granulomatosis, leishmaniasis, tuberculosis, syphilis, fungal infections, leprosy, nasopalatine duct cyst, Rosai-Dorfman disease, intranasal narcotic abuse, lymphomas, basal

cell carcinoma, verrucous carcinoma, metastatic renal cell carcinoma [5, 14, 15]. The differential histological diagnosis of the granulomatous stage of rhinoscleroma, which includes the case in question, involves tuberculosis, leprosy, malacoplakia, granular-cell tumour and renal cell carcinoma metastases. The presence of neutrophils and microabscess formation is not characteristic of either rhinoscleroma or leprosy and tuberculosis. They can be present in granulation during the course of rhinoscleroma, but they are not to be found in granulation tissue typical of tuberculosis, in which granulomas are formed by epithelioid cells and multinucleated giant cells with centrally localized caseous necrosis. Apart from this, mycobacteria may be confirmed by Ziehl-Neelsen stain [5]. Mikulicz's cells are characteristic but not pathognomonic for rhinoscleroma, because similar macrophages are also found in leprosy and in non-specific reactions to toxins produced by various pathogens [2, 7-9]. In rhinoscleroma these cells are dispersed and they never form isolated aggregates [9]. They are macrophages with light foamy cytoplasm, sometimes with vacuoles 100µm in diameter, with excentrically located cell nucleus. Using Warthin-Starry, PAS or Giemsa stain as well as in the ultrastructure [13] bacteria can be seen in their cytoplasm. A microscopic examination in leprosy reveals large aggregates of foamy macrophages containing lipids and conglomerates of leprosy mycobacteria (Virchow's cells) [13]. If the cells have eosinophilic cytoplasm, granular-cell tumour should be taken into consideration in the differential diagnosis (tumour cells are S-100 protein positive). In malacoplakia which very rarely affects the nasal cavity, Michaelis-Gutmann bodies can be found. Small aggregations of Mikulicz's cells can mimic clear cells that are seen in renal cell carcinoma, however tumour cells in renal cell carcinoma show keratin and vimentin expression and no CD68 expression.

References

1. de Pontual L, Ovetchkine P, Rodrigueaz D, et al. Rhinoscleroma: A French national retrospective study of epidemiological and clinical features. *Clin Inf Dis* 2008; 47: 1396-1402.
2. Ireland PE: Rhinoscleroma-with the report of a case. *The Canadian Med Assoc J* 1936; 149-152.
3. Lubin JR, Jallow SE, Wilson WR, Grove AS, Albert DM. Rhinoscleroma with exophthalmos: a case report. *Br J Ophthalmol* 1981; 65: 14-17.
4. Simson ME, Granato L, Olivera RCB, Alcantara MPA. Rhinoscleroma: Case report. *Rev Bras Otorrinolaringol* 2006; 72: 568-571.
5. Abalkhail A, Satti M B, Uthman MA, et al. Rhinoscleroma: a clinicopathological study from the Gulf region. *Singapore Med J* 2007; 48: 148-151.
6. DiBartolomeo JR. Scleroma of the nose and pharynx. *West J Med* 1976; 124: 13-17.
7. Botelho-Nevers E, Gouriet F, Lepidi H, et al. Chronic nasal infection caused by *Klebsiella rhinoscleromatis* or *Klebsiella ozaena*: two forgotten infectious diseases. *Intern J Infectious Diseases* 2007; 11: 423-429.
8. Bailhache A, Dehesdin D, Francois A, Marie J P, Choussy O. Rhinoscleroma of the sinuses. *Rhinology* 2008; 46: 338-341.
9. Chan T V, Spiegel J H: *Klebsiella rhinoscleromatis* of the membranous nasal septum. *J Lar and Otol* 2007; 121: 998-1002.
10. Hoffmann EO, Leland DL, Harkin JC. The Mikulicz Cell in Rhinoscleroma. *Am J Pathol* 1973; 73: 1, 47-58.
11. Verma G, Kanawaty D, Hyland R. Rhinoscleroma causing upper airway obstruction. *Can Respir J* 2005; 12: 43-45.
12. Woyke S, Domagała W, Olszewski W. Electron microscopic studies of scleroma granulation tissue. *Acta Med Pol* 1969; 10, 231-242.
13. Stachura J, Domagała W. Patologia znaczy słowo o chorobie 2003; 237-240.
14. Zafar U, Khan N, Afroz N, Hasan SA. Clinicopathological study of non-neoplastic lesions of nasal cavity and paranasal sinuses. *Indian J Pathol Microbiol* 2008; 51: 26-29.
15. Fuchs A, Tanner SB. Granulomatous disorders of the nose and paranasal sinuses. *Curr Opin Otolaryngol Head Neck Surg* 2009; 17: 23-27.

Address for correspondence

Katarzyna Karpińska-Kaczmarczyk MD
 Zakład Patomorfologii,
 Pomorski Uniwersytet Medyczny
 ul. Unii Lubelskiej 1
 71-252 Szczecin
 e-mail: kkapr@pum.edu.pl