

CASE REPORT

An erosive mass involving the nasal cavity and maxillary sinus: rhinosporidiosis

Nazal kavite ve maksiller sinüsü tutan eroziv kitle: Rhinosporidiosis

Gürkan KESKİN, M.D.,¹ Emre ÜSTÜNDAĞ, M.D.,¹ Sevgiye KAÇARÖZKARA, M.D.,² Sinem KESKİN, M.D.¹

Rhinosporidiosis is a chronic granulomatous inflammatory disease of the mucous membranes caused by *Rhinosporidium seberi*. The disease is endemic in India but is very rare in other countries. A review of the literature shows that our patient is the second documented case from Turkey. The involvement and erosion of the anterior wall of maxillary sinus as in this case is very rare. In this article we describe the clinico-pathological features of a case presenting in 47-year-old-male and discuss transmission of the disease.

Key Words: Rhinosporidiosis/pathology/immunology.

Rhinosporidiosis, *rhinosporidium seberi*'nin neden olduğu mukus membranların kronik granülomatöz inflamatuvar bir hastalığıdır. Hastalık Hindistan'da özgündür fakat diğer ülkelerde oldukça nadirdir. Literatür araştırıldığında hastamız Türkiye'de kayıtlara geçen ikinci olgudur. Bu olguda olduğu gibi maksiller sinüsün tutulumu ve ön duvarının erozyonu oldukça nadirdir. Bu makalede 47 yaşında erkek hastanın klinikopatolojik özellikleri sunuldu ve hastalığın yayılımı tartışıldı.

Anahtar Sözcükler: Rhinosporidiosis/patoloji/immünoloji.

Rhinosporidiosis is a chronic granulomatous inflammatory disease of the nasal mucous membranes caused by *Rhinosporidium seberi*.^[1] The disease is characterized by formation of polyps on the mucous membranes of the infected host. Rhinosporidiosis is not a life-threatening disease unless it affects tracheo-bronchial tree. Its treatment is basically limited to surgical removal of polyps. It is most prevalent in Asia, but the disease has been reported sporadically in other continents.^[2]

Transmission of the disease is not clear but the organism is thought to be contracted from infected

soil and water, probably as a result of contaminated drinking water.^[3]

We report a case of aggressive rhinosporidiosis that eroded the right maxillary sinus. We also describe the clinico-pathological features of rhinosporidiosis and discuss transmission of the disease.

CASE REPORT

A 47-year-old-male was referred to our ENT clinic with complaints of increasing nasal obstruction and bloodstained watery discharge from the right nostril. He had also a three month history of

◆ Departments of ¹Otolaryngology and ²Pathology, Medicine Faculty of Kocaeli University (Kocaeli Üniversitesi Tıp Fakültesi 'Kulak Burun Boğaz Hastalıkları Anabilim Dalı, ²Patoloji Anabilim Dalı), Kocaeli, Turkey.

◆ Received - December 2, 2003 (Dergiye geliş tarihi - 2 Aralık 2003). Request for revision - June 1, 2004 (Düzeltilme isteği - 1 Haziran 2004). Accepted for publication - June 18, 2004 (Yayın için kabul tarihi - 18 Haziran 2004).

◆ Correspondence (İletişim adresi): Dr. Gürkan Keskin. Kocaeli Üniversitesi Tıp Fakültesi KBB Hastalıkları Anabilim Dalı, 41900 Kocaeli, Turkey. Tel: +90 262 - 233 59 82 / 144 Fax (Faks): +90 262 - 233 54 88 e-mail (e-posta): gurkankeskin@yahoo.com

loosing the right molar teeth. He had never been abroad.

On examination; an obstructive, fleshy, haemorrhagic polypoid mass was found in the right nasal cavity. The mass was fluctuating and filling the right fossa canina. Palpation of the fossa canina revealed softening of the underlying bone, indicating erosion of the anterior wall of the right maxillary antrum.

A fine needle aspiration biopsy from fossa canina was performed with the tentative diagnosis of malignancy and interpreted as to be suggestive of rhinosporidial infection. The sporangias were observed within the metaplastic epithelial cells as well as distributed freely on the smears (Fig. 1). As it is extremely rare in our country, an insicisional biopsy was taken and histopathological examination confirmed the cytologic diagnosis. PAS (periodic-acid-Schiff) stain showed positivity for the capsule of the

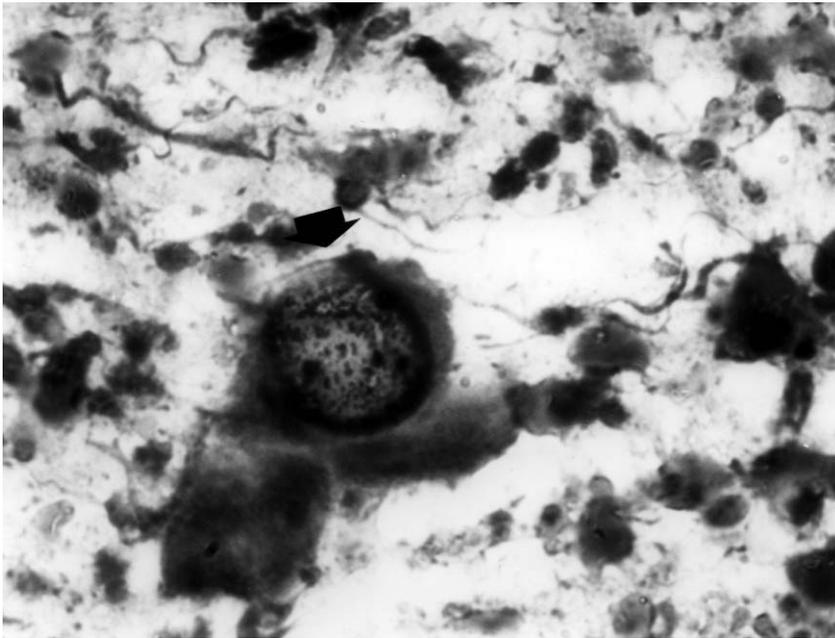


Fig. 1 - The typical spherule (arrow) of Rhinosporidium seeberi on fine needle aspiration cytology; well-circumscribed, globular structure with several endospores within (May-Grünwaldt-Giemsa, x 1250, oil immersion).

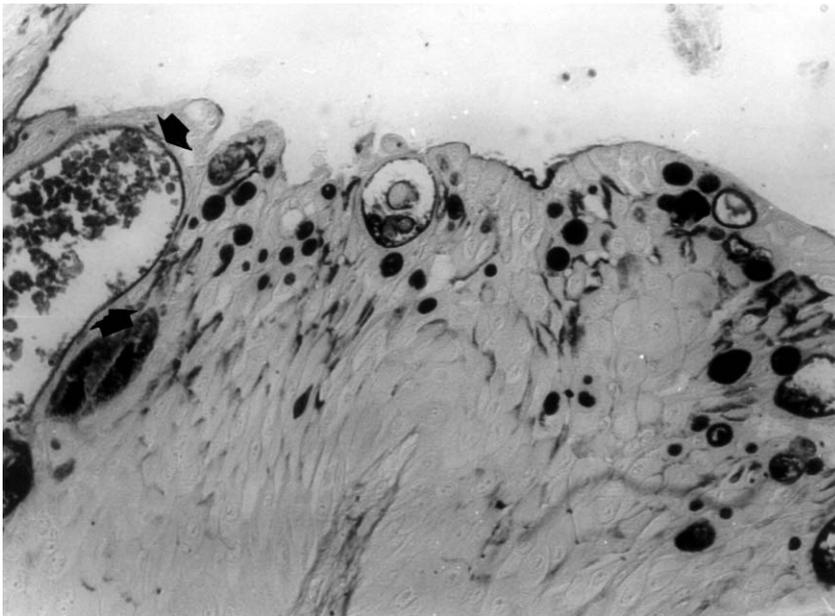


Fig. 2 - The spherules showing great variability in size, up to ten fold. The mature sporangium (arrow) is together with immature trophic forms along the lining epithelium (Grocott-Gomori's methenamine silver stain, x 310).

microorganism. Grocotte-Gomori's methenamine silver impregnation technique also stained both the trophic forms and sporangia (Fig. 2).

A coronal CT scan showed a large mass involving the right nasal cavity and maxillary antrum. The mass was extended anteriorly, and eroded anterior wall of the right maxillary sinus (Fig. 3).

It was removed by transantral-transnasal approach (Fig. 4). Excessive bleeding was controlled by anterior and posterior nasal packs. The excised mass was a multiple polypoidal lesion measured 11x8x3 cm (Fig. 5). On histopathological examination, the immature sporangia of *Rhinosporidium seeberi* as well as the mature ones were seen within the metaplastic squamous epithelial cells together with intraepithelial lymphocytes and polymorphs. The stroma was infiltrated with lymphocytes, plasma cells, polymorphs and eosinophils. No granulomatous inflammatory reaction was observed (Fig. 6). The same histochemical stains were repeated with the identical results.

The patient was followed up for two years after the surgery and no recurrence occurred.

DISCUSSION

Guillermo Seeber from Argentina reported an inflammatory disease of the nasal mucosa caused by a parasite, which now bears his name eponymously. The disease is reported most commonly from India

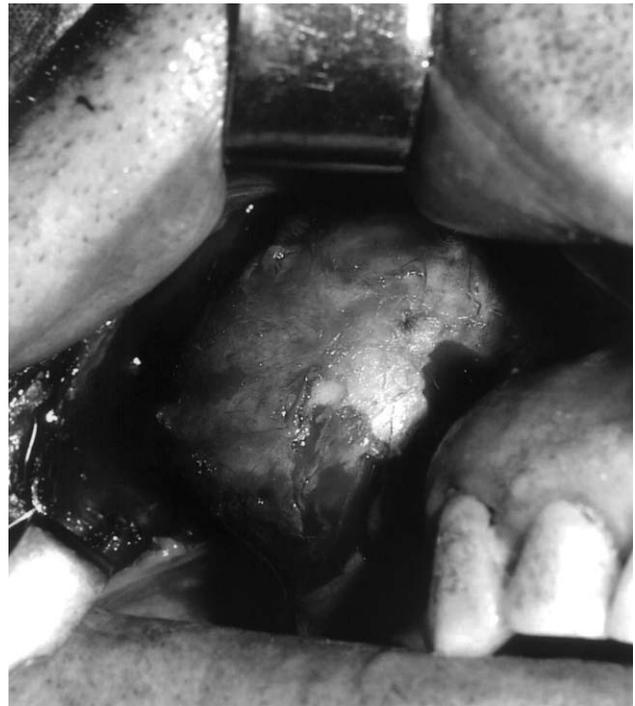


Fig. 4 - The lesion was removed by transantral-transnasal approach.

and Sri Lanka, but very rare in Europe and other continents. According to van der Coer et al.,^[4] there are 23 reported cases in Europe only six of them are Europeans. Radovanovic et al.^[5] published an outbreak that began in 1995 affected 21 individuals; five of them had nasal involvement. The first case in



Fig. 3 - The coronal CT scan showed a large mass involving the right nasal cavity and maxillary antrum. The mass eroded anterior wall of the right maxillary sinus (arrow-head).

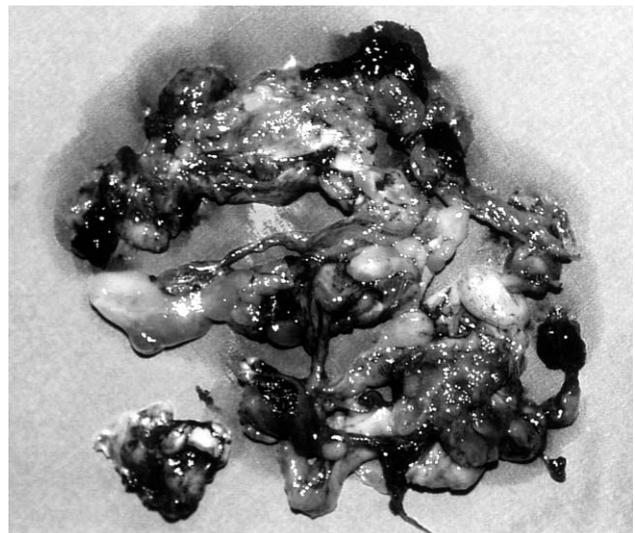


Fig. 5 - Macroscopical appearance of the excised polypoid mass lesion.

Turkey was encountered in 1955 and published in the literature.^[4] The presented case is the second documented one from Turkey.

Rhinosporidium seeberi is the causative organism of rhinosporidiosis. It is a chronic granulomatous inflammatory disease characterized by polypoid lesions of the mucous membranes. The most common effected site of infection is the mucous membranes of the nasal cavity and the nasopharynx. Other sites of involvement include the conjunctiva, lacrimal sac, oral cavity, larynx, vagina, rectum, urethra, bone and brain.^[1,6]

Nasal obstruction and epistaxis are the most common and often the presenting symptoms. Watery discharge, often tinged with blood is another symptom and become mucopurulent with subsequent infection. Nasal polyps and hypertrophic mucosa can be seen upon examination. Grey or yellow spots, which represent the bulging sporangia through the attenuated epithelium, give a characteristic appearance.^[7] The involvement and erosion of the maxillary sinus as in the presented case is very rare. Misdiagnosis of this condition by the physicians who are not familiar with this disease is possible. An erosive form of rhinosporidiosis as our case should be differentiated from malignancy. Fine needle aspiration biopsy features of rhinosporidiosis are typical. The spherules are well-circumscribed,

globular structured with several endospores within. Preoperative diagnosis of rhinosporidiosis is possible even in cases with unusual clinical presentation and from the countries where the condition is not endemic.

The mode of transmission, pathogenesis and taxonomic position of the pathogen are still not clear. The mode of transmission is supposed to be via the contaminated dust or water. There seems to be a relationship between disease and agriculture, suggesting that rhinosporidiosis most likely lives in soil.^[3] According to Radovanovic et al.,^[5] 21 patients had been exposed to the same source of stagnant water. The custom of mechanically cleansing of the nose may create trauma that might predispose to infection. Our patient had a contact (three days long) with soil after an earthquake in our city. This might play a role in transmission for this patient.

Rhinosporidium seeberi has been classified as a fungus on the basis of morphologic and histochemical characteristics. In the recent studies, using polymerase chain reaction, phylogenetic relationships have shown that *Rhinosporidium seeberi* is a protist from a novel clade of parasites that infect fish and amphibians.^[8,9] On the other hand, Ahluwalia^[10] suppose that a prokaryotic cyanobacterium is the causative agent of the disease and Rhinosporidiosis is the first human disease to be caused by a cyanobacterium.

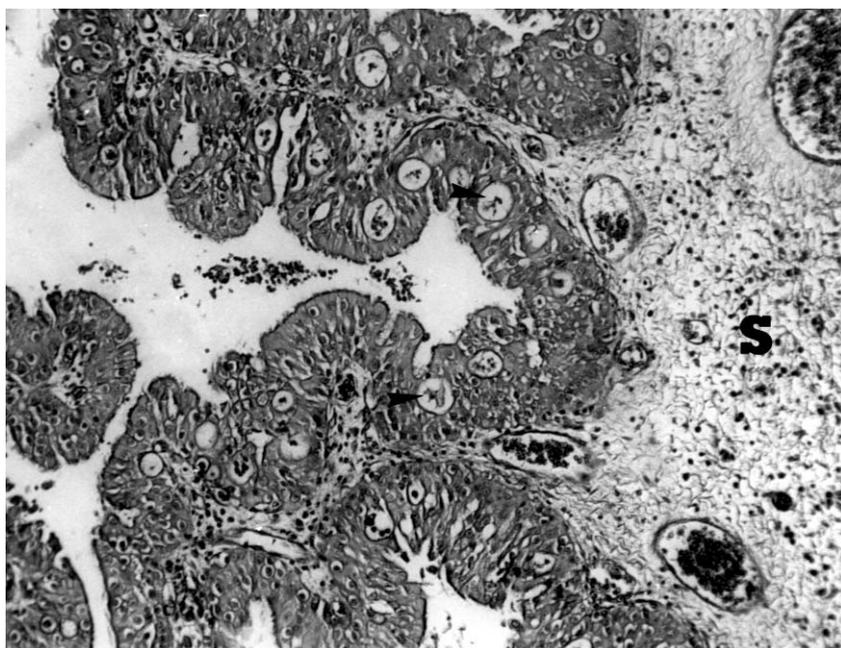


Fig. 6 - Papillomatous hyperplasia of the lining stratified squamous epithelium containing *Rhinosporidium seeberi* (arrowhead). Underlining stroma (s) is edematous, vascular and infiltrated with inflammatory cells (Hematoxylin & Eosine, x125).

The treatment, with the lowest incidence of recurrence, is surgical extirpation and cauterization of the base of the lesions, no medical treatment without surgery has been found to be effective.^[4] Amphotericin B has been used locally after surgery to prevent re-infection and spread but the drug has no effect on reoccurrence.^[3] In Nair's^[11] clinical trial with diaminodiphenylsulfone (dapson), on 32 patients with nasal and nasopharyngeal rhinosporidiosis, 71.4% (20 patients) did not show recurrence in a three year period, and none of them needed additional surgery in that period.

In conclusion the erosive form of rhinosporidiosis is very rare especially in non-endemic areas. The mode of transmission of the disease remains uncertain and the treatment of choice is surgery.

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