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Rhinosporidiosis in Europe

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Abstract
Rhinosporidiosis is an inflammatory disease of the nasal mucosa caused by *Rhinosporidium seeberi*. The disease is endemic in India but very rare in Europe or other continents. The literature concerning aetiology, clinical appearance, morphology and treatment is reviewed. All reported European cases are summarized.

Introduction

Rhinosporidiosis is a chronic granulomatous inflammatory disease characterized by polypoid lesions of the mucous membranes. The principal site of infection is the mucous membrane of the nose and nasopharynx. Other sites described are: conjunctiva, oral cavity, lacrimal sac, urethra, genitalia, tracheobronchial tree, larynx, skin, bone and paranasal cavities (Satyanarayana, 1960; Gupta, 1966; Khan *et al.*, 1969; Kameswaran *et al.*, 1970; Yesudian, 1988). The organism responsible is called *Rhinosporidium seeberi*, long thought to be of fungal origin. Reported cases range from 3 to 73 years with a maximum incidence between the ages of 20 and 30 years (Satyanarayana, 1960; Khan *et al.*, 1969; Lasser and Smith, 1976; von Haacke and Mugliston, 1982). One author reported a maximum incidence between 11–20 years (Moses *et al.*, 1988). Rhinosporidiosis is more common in males than females although the sex distribution varies in different reports (Satyanarayana, 1960; Khan *et al.*, 1969; Kannan-Kutty and Teh, 1975; Moses *et al.*, 1988; Chitravel *et al.*, 1990).

Nasal obstruction and epistaxis are the most common and often the presenting symptoms. Watery discharge, often tinged with blood, is a third symptom and becomes mucopurulent with subsequent infection. Other less common symptoms include sneezing, coryza and a sensation of the presence of a foreign body.

Soft and friable nasal polyps and mucosa can be found upon examination. Grey or yellow spots which represent the bulging sporangia through the attentuated epithelium give a characteristic strawberry appearance (Khan *et al.*, 1969; Vanbreuseghem, 1976).

Associated diseases are incidental but schistomiasis, syphilis, lepra and carcinoma are reported (Satyanarayana, 1960; Vanbreuseghem, 1976; Ahluwalia and Bahadur, 1990).

Rhinosporidiosis is rarely fatal. Haemorrhage and generalized dissemination may however lead to death.

Rhinosporidiosis is endemic in India and Sri Lanka (Satyanarayana, 1960; Karunaratne, 1963; Vanbreuseghem, 1976). Sporadic cases are reported from North, Central and South America, South and East Africa, Japan, Philippine Islands, Malay States, Canada, Uganda and Iran (Engzell and Jones, 1973; Fortin and Meisels, 1974; Lasser and Smith, 1976; Jimenez *et al.*, 1984; Woodard and Hudson, 1984; Goihman-Yahr, 1986; Chetty and Cooper, 1987).

In Europe Rhinosporidiosis is quite rare: only a few cases have been described in the literature (Table I).

Case report

In 1981, at the age of 11, our patient, a Dutch Caucasian male, was referred to our ENT clinic with the complaint of increasing nasal obstruction. There were no other associated symptoms and, especially, no history of epistaxis. Medical history was normal. He was a son of a pig-raiser and had never been abroad. The basic feed products for the pigs were imported from Africa.

On examination, an obstructive polypoid mass was found in the left nasal cavity. It had a reddish aspect. There were no enlarged lymph-nodes present and the remaining ENT examination showed no abnormalities. The mass was removed by endonasal surgery. Bleeding was controlled by a nasal pack. Histological examination revealed a granulomatous infection with the characteristic appearance of *Rhinosporidium seeberi* (Figs. 1 & 2).

The patient has been seen on a regular basis since. Unfortunately the same complaint recurred six years later. A large and, this time, easily bleeding polypoid mass was found in the left nasal cavity upon examination. It had a characteristic strawberry appearance. No other lesions could be found. Blood and urine-chemistry were normal. A CT-scan showed a tumour in the left nasal cavity reaching the epipharynx (Fig. 3).

A left lateral rhinotomy was performed to remove the infected inferior turbinate and lateral nasal wall. Again *Rhinosporidium seeberi* was demonstrated by histological examination. Recurrence of granulomatous mucosa could be seen two years after the lateral rhinotomy. This has been recently removed by local excision.

Discussion

In 1900, Rhinosporidiosis was reported by the Argentinean Seeber as an inflammatory disease of the nasal mucosa caused by a parasite which now bears his name eponymously, as suggested by Ashworth and Logan Turner (1923). They described the responsible microorganism in detail and concluded it was a fungus belonging to the class of phycomycetes. Vanbreuseghem (1973) challenged this fungal hypothesis, mainly because all attempts to culture the organism in vitro had failed.

Studies of the life cycle of *Rhinosporidium seeberi* are still based on the original work of Ashworth and Logan Turner.

The early trophocyte, comparable with a spore, will grow until it reaches about 50 μm in diameter (Ashworth and Logan Turner, 1923; Kannan-Kutty and Teh, 1974). From that moment on the nucleus undergoes a series of mitotic divisions. From about the seventh division, when the developing trophocyte is 100 μm in diameter and contains 128 nuclei, the envelope of the trophocyte is thickened by a deposit of cellulose which increases its total thickness to 8–9 μm over most of its surface and 14–15 μm at the site of the future spore. By the 14th division the sporangium is about 140 μm in diameter and contains 16000

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TABLE I
ALL REPORTED CASES OF RHINOSPORIDIOSIS IN EUROPE

Author and reference number	Age	Sex	Reported in:	Native of
Alonso (1987)	19	M	Spain	Spain
Ashworth (1923)	23	M	England	India
Atav (1955)	12	F	Turkey	Turkey
Fortin (1974)	40	M	Canada/England	India
von Haacke (1982)	38	M	England	India
Karunaratne (1963)	–	M	England	India (4 patients)
				China
				England
				Sth Africa
	–	M	Scotland	India (2 patients)
	–	M	Italy	Italy
	–	F	Italy	Italy
	–	M	Italy	India
	–	M	Switzerland	Japan
	–	M	Rumania	Rumania
Lecluse (1972)	54	M	Netherlands	India
Prins (1983)	34	M	Netherlands	India
Sharp (1944)	31	M	England	India
Vincent (1982)	24	M	Belgium	India
Total		21 M, 2F	23 cases	6 Europeans

young spores about 3 μm in diameter. Maturation of the sporangium typically occurs from the centre outward but about one-third of the spores fail to mature. At the end of this process the sporangium reaches a diameter of about 200–250 μm in diameter and discharges its contents, a phenomenon which can be demonstrated in fixed sections. Enormous numbers of spores escape into the nasal secretions. Electron microscopical studies generally confirm Ashworth and Logan Turner's original view that the spore is uninucleate with multiple inclusions (Kannan-Kutty and Teh, 1974, 1975; Vanbreuseghem, 1976). Ashworth and Logan Turner failed to find any histological evidence for re-infection of the epithelium by spores shed from a neighbouring surface, but demonstrated transformation of the spore to the trophic phase in subepithelial lymph spaces.

The histological appearance of the lesions reveal papilloma-

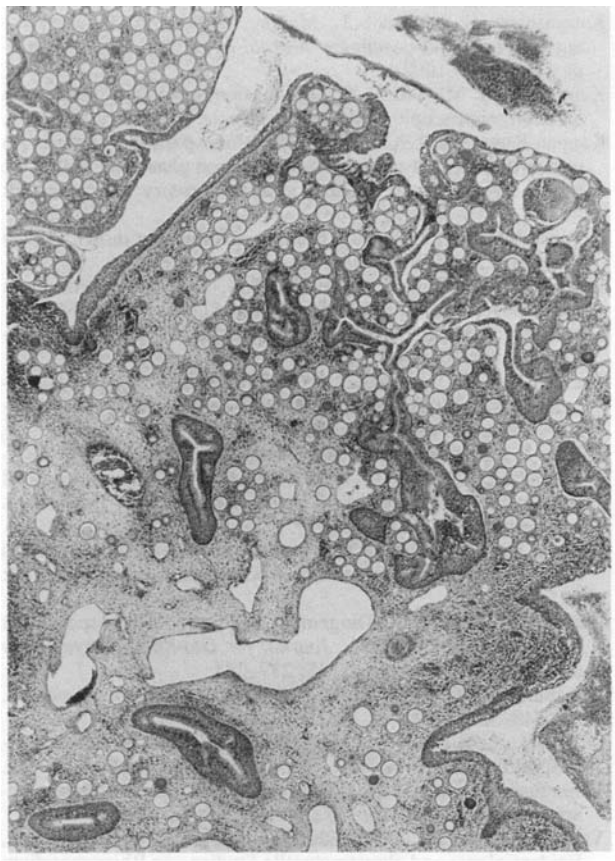

FIG. 1
A low power photomicrograph illustrating features of the polypoid inflammatory mass. There are numerous round Rhinosporidium cysts in the chronically inflamed stroma (Haematoxylin-eosin stain, original magnification ×6.25).

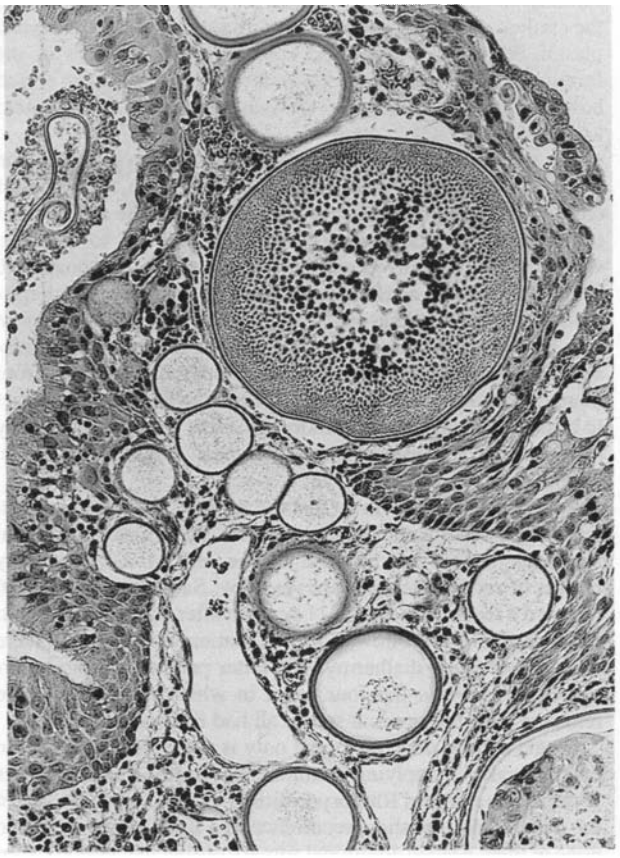

FIG. 2
A high power photomicrograph illustrating an intact thick-walled mature Rhinosporidium sporangium with innumerable endospores (Haematoxylin-eosin stain, original magnification ×50).

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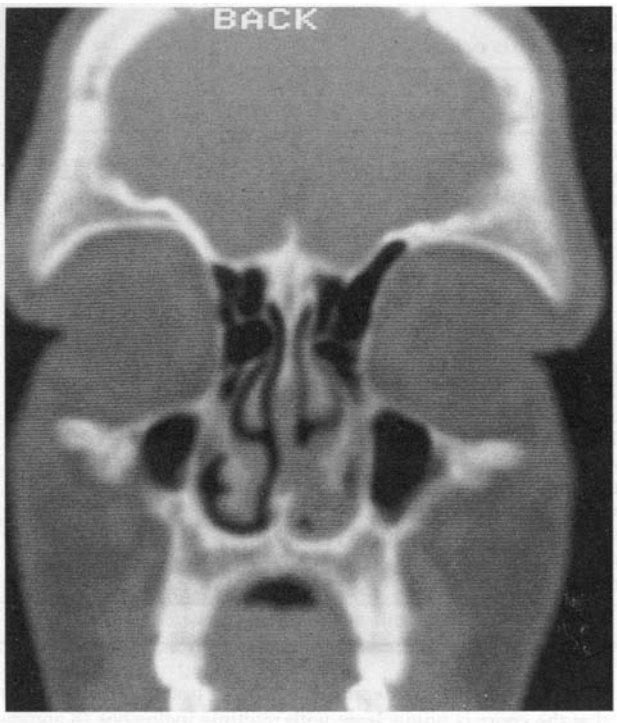

FIG. 3

CT scan of nasal cavity showing an obstructive mass around the left inferior turbinate.

tous hyperplasia of different degrees of the lining epithelium and infiltration with mostly lymphocytes around the spores. The sporangia and spores can be found in subepithelial areas and in the epithelial lining. The stroma is infiltrated with lymphocytes, plasma cells, polymorphs and eosinophils. Giant cells of the foreign-body type appear occasionally around sporangia which have ruptured or degenerated. Neovascularization can be found as well.

No definite evidence has been uncovered concerning the mode of infection and transmission of the disease. A number of cases of Rhinosporidiosis have been found in cattle and pets but transmission of Rhinosporidiosis from animals to humans, which is theoretically possible, has yet to be demonstrated. There seems to be a relationship between the disease and agriculture, suggesting that Rhinosporidiosis most likely lives in soil. Ashworth and Logan Turner (1923) refer to direct transmission of discharged spores as an obvious method of infection from man to man. Although a definite course of transmission has yet to be demonstrated, many authors agree that water is a necessary medium of transmission (Satyanarayana, 1960; Karunaratne, 1963; Chetty and Cooper, 1987).

Several trials have shown that the most successful treatment, with the lowest incidence of recurrence, is surgical extirpation and cauterization of the base of the lesions. Unfortunately reports of recurrence-rates are very few: Satyanarayana (1960) reported a recurrence-rate of 11 per cent after surgical treatment; Khan *et al.* (1969) followed with 22 patients of whom 18 were treated by cutting diathermy. The latter patients had no recurrences, whereas in the four cases in which the lesions were removed with forceps and snares all had recurrent disease.

Single conservative treatment only is not widely used, but in one clinical trial applying diaminophenylsulfone to 32 patients with nasopharyngeal Rhinosporidiosis, 71.4 per cent (*i.e.* 20) of the patients did not show recurrence in a three-year period and none of them needed additional surgery during that period (Nair, 1979).

In conclusion Rhinosporidiosis is very rare in Europe, the mode of transmission remains uncertain and the treatment of choice is surgery.

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