Association of rhinoscleroma with rhinosporidiosis*

Awad M. Al-Serhani[1], Ali S. Al Qahtani[1], Maha Arafa[2]

[1] Department of ENT/Head & Neck Surgery, King Abdul Aziz University Hospital, Riyadh, Kingdom of Saudi Arabia
[2] Department of Pathology, King Khalid University Hospital, Riyadh, Kingdom of Saudi Arabia

SUMMARY

Rhinoscleroma caused by the bacillus Klebsiella rhinoscleromatis and rhinosporidiosis caused by the fungus Rhinosporidium seebri are rare, specific nasal infections, both of which have a certain geographical distribution. To the best of our knowledge no association between them has been reported in the international literature. We have documented such an association in two male Indian patients aged – 32 and 27 years, respectively – both presenting with unilateral blood-stained discharge and nasal blockage. They showed strawberry-like polypoidal masses, and histological examination confirmed the diagnosis. Klebsiella rhinoscleromatis was cultured twice in the first case. The patients were treated with complete excision and a long course of septrin, for which Klebsiella rhinoscleromatis is sensitive. The purpose of this paper is to report the first association of these two granulomatous infections, to show the impact of immigration on the differential diagnosis, and to review the relevant literature.

Keywords: rhinoscleroma, rhinosporidiosis, granulomatous disease.

INTRODUCTION

Rhinoscleroma and rhinosporidiosis are granulomatous infections affecting the nose and sinuses as the commonest site. Both have been reported in India where our patients came from. Although an immunological defect (impairment of T-lymphocytic function with normal humoral immune system) has been indicated in patients with rhinoscleroma (Dogheim et al., 1986), little is known about the immunology of rhinosporidiosis (Kornita et al., 1990). Tracing the international literature failed to reveal any association between them. Our patients presented with clinical lesions highly suspicious of rhinosporidiosis, which was documented histologically. The incidental histological and microbiological findings of rhinoscleroma makes this a unique association, worth reporting and carefully looking for in the future.

PATIENTS

Case 1

A 32-year-old Indian male who arrived in Saudi Arabia one year prior to his admission presented with a 4-month history of left-sided persistent nasal obstruction, associated with blood-stained watery discharge. He had previous "nasal polypectomy" three times between 1990 and 1994 in India. No histological information was available. There was no history of severe epistaxis.

When examined, the left nostril was completely obstructed by a reddish polypoidal friable lesion, with a strawberry-like appearance and bleeding on manipulation. The exact origin of the mass was unclear. There was slight swelling of the medial canthus, but vision was not affected and eye movement was normal. Axial and coronal CT-scans revealed opacification of the whole left paranasal sinuses and nasal cavity. The lesion was enhanced and showed destructive features. A biopsy of the lesion was taken, and this was suggestive for an association of rhinoscleroma and rhinosporidiosis (see Results). *Klebsiella rhinoscleromatis* was cultured twice, once from a nasal swab and crust, and another from fluid aspirated from a swelling that developed on admission beneath the left lower eyelid – where it was associated with *Haemophilus influenzae* – and responded to cefuroxime.

Through a left lateral rhinotomy, the mass (which was found to be extending to the lacrimal sac) was excised. The patient received a course of septrin, for which *Klebsiella rhinocleromatis* is sensitive, for 2 months and did well post-operatively. One-year follow-up showed no recurrence.

Case 2

A 27-year-old Indian male presented with left-sided nasal obstruction associated with recurrent epistaxis for one year. He

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had a history of recurrent nasal polyps which had been removed previously, 5 times during the past years in India. No histopathological information was available. Nasal examination showed at the left side a mass which was strawberry-like in appearance and originated from the middle meatus. A CT scan of the paranasal sinuses showed that the disease was limited to the left ethmoid sinus. A biopsy of the lesion revealed a diagnosis similar to case No. 1 (see Results). The mass was excised intranasally with uneventful recovery. The patient developed a recurrence 2 months later and a revision was performed through an external approach, and this confirmed the association of rhinoscleroma and rhinosporidiosis.

RESULTS

Routine histological sections from the resected nasal lesion in both cases revealed polypoid mucosa lined by pseudostratified columnar ciliated epithelium, exhibiting focal areas of squamous metaplastic changes. There was a marked lymphoplasmocytic infiltration with formation of prominent lymphoid follicles. Stromal fibrosis was more prominent in case No. 2.

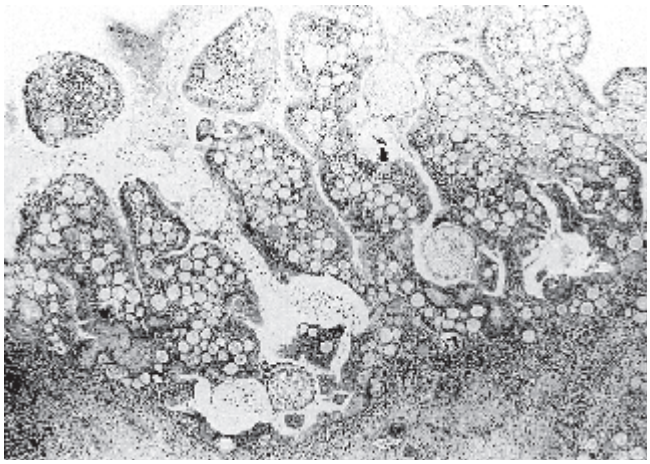

Figure 1. Low-magnification micrograph showing polypoid projection of nasal mucosa containing trophocytes and sporangia. Note released spores in the crypt (Haematoxylin and eosin staining; ×40)

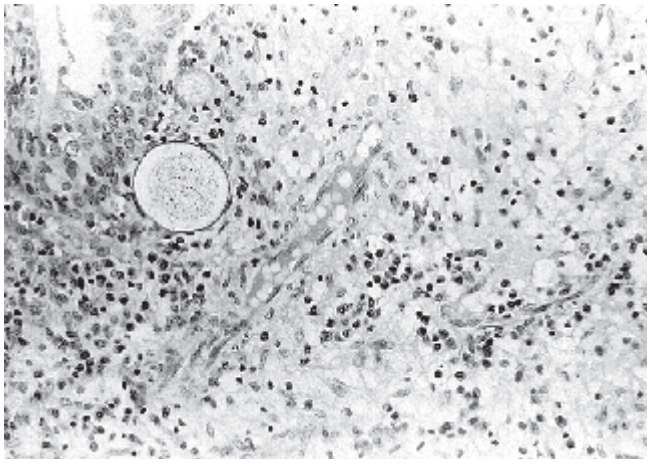

Figure 2. Light micrograph showing sporangia near the lining epithelium (left) and a few clusters of Mikulicz's cells (right) (Haematoxylin and eosin staining; ×250).

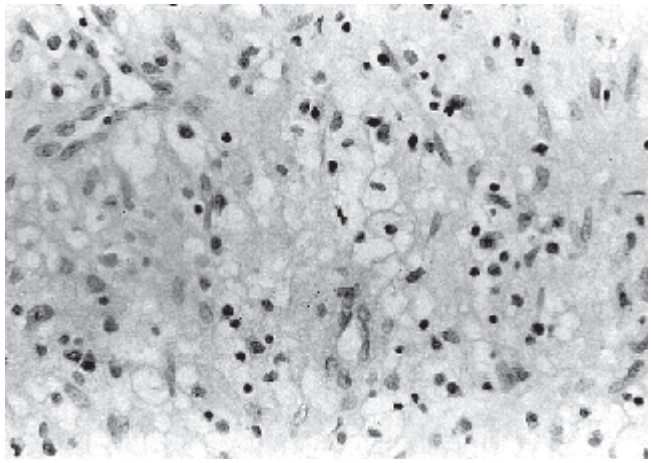

Figure 3. High-magnification micrograph showing clusters of macrophages having a thick cytoplasmic membrane and vacuolated cytoplasm (Haematoxylin and eosin staining; ×400).

The lamina propria contained numerous globular cysts with a sharply-defined eosinophilic wall, some of which contained round spores. Various developmental stages of the infective organisms were found within the mucosa. Some of the crypts contained released spores. Some sporangia were surrounded by foreign-body-type giant cells. In addition, there were groups and clusters of large macrophages with a foamy-looking cytoplasm, containing loosely clustered bacilli (Miculicz's cells). The surrounding tissue showed an infiltration by lymphocytes and plasma cells containing Russell's bodies (Figures 1–3).

DISCUSSION

Rhinosporidiosis is a fungal infection that is thought to be contracted from infected soil and contaminated water, although Koch's postulates for transmission have not been fulfilled (Batsakis and El Naggar, 1992). The disease is endemic to South India and Sri Lanka where 88% of the cases have been reported (Blitzer and Lawson, 1993). It is also seen in East Africa, Italy, Brazil, and Malaya (Kornita et al., 1990; Van Der Goer et al., 1992; Batsakis and El-Naggar, 1992).

The commonest site is the nasal cavity, where the septum, turbinates and floor of the nose are affected. The polypoidal mucosa with a strawberry-like appearance leads to nasal obstruction, epistaxis, and blood-stained discharge. An associated ocular involvement (case No. 1) is often seen. Other sites include the urethra, vagina, larynx, and skin (Van Der Goer et al., 1992; Mears and Amerasinghe, 1992). There is a male preponderance of 4:1, which is probably secondary to occupational and traditional exposure (Satyanarayan, 1960). The diagnosis is confirmed by detecting the infective organism during routine light microscopy. The characteristic trophocytes and sporangia containing spores, are present in substantial numbers in various developmental stages within the polypoidal mucosa. However, attempts to culture it have been unsuccessful (Blitzer and Lawson, 1993). Association with schistosomiasis, syphilis, leprosy and carcinoma have been reported (Satyanarayan, 1960; Van Der Goer et al., 1992), but not with rhinoscleroma.

Although some forms of medical treatment have been advocated (Van Der Goer et al., 1992; Blitzer and Lawson, 1993; Job et al., 1993), surgical excision is still the standard treatment, preferably with cauterization of the margins (Van Der Goer et al., 1992; Batsakis and El-Naggar, 1992; Blitzer and Lawson, 1993). The recurrence rate is approximately 11% (Van Der Goer et al., 1992; Mears and Amerasinghe, 1992). Our cases had presumably recurrences (3 times in case No. 1, and 5 times in case No. 2), probably due to incomplete removal.

Rhinoscleroma is a granulomatous infection caused by *Klebsiella rhinoscleromitis*. It has an affinity to the upper respiratory mucosa and can be cultured from infected tissue in 98% of the cases (McDonald, 1993). The disease is endemic in 25 countries (Eastern Europe, North and Central Africa, and South and Central America), and environmental factors are conducive. It is commonly reported in Poland, Egypt and Indonesia, and is being recognized in increasing numbers in non-endemic areas with an influx of immigrants from the endemic zones (Batsakis and El-Naggar, 1992). Poor hygiene, crowded living and malnutrition are predisposing factors for rhinoscleroma. However, it is not highly contagious or hereditary (Batsakis and El-Naggar, 1992).

The disease manifests itself in four (overlapping) stages, and appearing as a slowly progressive midfacial necrotizing lesion presenting initially with nasal obstruction and crusting. The destruction of bone and cartilage produces deformities of the nasal pyramid.

The *catarrhal (exudative) stage* is an acute or active chronic inflammation with oedema, congestion, and suppurative necrosis characterized by foul-smelling and purulent rhinorrhoea for a few weeks. The infective organism may be obscured by the inflammation, so that histopathological diagnosis is not possible (Abou-Seif et al., 1991). The *atrophic stage* simulates atrophic rhinitis with nasal plaque and crust formation. In the *granulomatous (proliferative) stage* the diagnostic features are most commonly found. There are clusters or sheets of large vacuolated histiocytes (Mikulicz's cells) and plasma cells containing Russell's bodies. The organisms are found within the cytoplasm of these cells or between the cells after release. If numerous, they can be seen after haematoxylin and eosin staining, but sometimes Giemsa staining, silver impregnation or immunocytochemical staining are required (Batsakis and El-Naggar, 1992). The *fibrosis stage* often causes stenosis requiring surgical excision (McDonald, 1993). Histological findings are usually nonspecific and may be devoid of organism.

Although a definitive cure for rhinoscleroma is difficult, various forms of medical therapy – including systemic and local antibiotics and local acriflavin – have been tried with different degrees of success (Shaer et al., 1981; Hashash et al., 1983; Gamea and El-Tatawi, 1990; Job et al., 1993). Surgical excision, preferably with laser, is the standard treatment for the fibrotic stage (Maher et al., 1990; Batsakis and El-Naggar, 1992), especially since steroids and radiation are not effective (McDonald, 1993).

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Dr. Awad M. Al-Serhani
Department of ENT/Head & Neck Surgery
King Abdul Aziz University Hospital
P.O. Box 245
Riyadh 11411
Kingdom of Saudi Arabia