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Unusual case of disseminated rhinosporidiosis with pulmonary involvement

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Abstract : Rhinosporidiosis is a chronic granulomatous disease that commonly affects the nasal and nasopharyngeal mucosa caused by Rhinosporidium seeberi. It rarely presents as disseminateddisease. Pulmonary involvement is even rarer. We describe a patient who had undergone treatment for recurrent nasal rhinosporidiosis and who presented with disseminated disease in the form of multiple subcutaneous swellings and pulmonary lesions.

Keyword :DIsseminated rhinosporidiosis, Pulmonary rhinosporidiosis

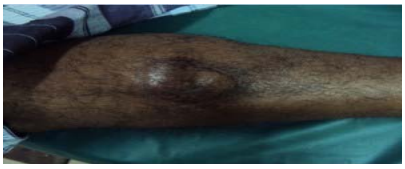
Background

Disseminated rhinosporidiosis and involvement of tissues other than the nasal or nasopharyngeal mucosa are rarely encountered. In 2006, Rajakannu et al1 reported a case of disseminated cutaneous rhinosporidiosis, which presented with pulmonary involvement. Disseminated cutaneous rhinosporidosis without pulmonary involvement have been reported by few authors2,3,4,5,6. Rhinosporidosis involving the lower respiratory tract has also been reported7,8. Disseminated rhinosporidosis involving nail and talus bone has also been reported9,10.Lesions in sites away from the nasal cavity are believed to be brought about by direct infection or by finger-borne autoinoculation11. Rarely, disseminated disease occurs due to hematogenous spread12. The most successful treatment is wide local excision of the lesions and electrodesiccation of the base of the lesions to reduce the chance of recurrence11. Dapsone, a bacteriostatic agent, has been used in the postoperative period to reduce the chance of recurrence13,14.

Case Report

Our case is a 35 year old male who had undergone recurrent surgeries for nasal rhinosporidosis thrice in the last 4 years and a surgery for a cutaneous rhinosporidosis in the gluteal region 6 months back. All the four times the excised tissue was histopathologically proven to be rhinosporidosis. Now the patient presented with a subcutaneous swelling of 8cm * 6 cm size over his right leg (Fig. 1).

Fig 1



Nasal examination revealed a polypoidal mass in his left nasal cavity.Computed tomography scan of paranasal sinuses revealed a growth completely filling his left nasal cavity (Fig 2).

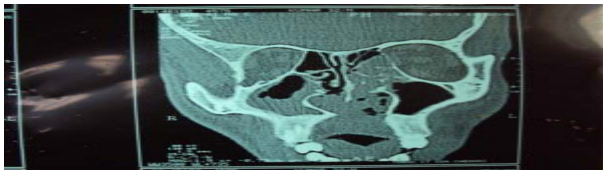
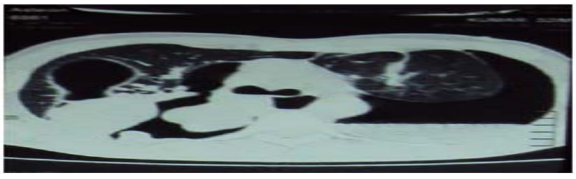


Fig 2

The patient was referred to us in favour of opinion regarding the chest X-ray that showed two homogeneous opacities in his right lung field with areas of localised hyperlucency in both lung fields (Fig 3).

Chest X ray of this patient taken 6 months back did not have any of these findings. Computed tomography scan of the thorax revealed two well-defined mass like lesions in his right lung with bilateral loculated pneumothorax (Fig. 4).

Fig 4



Bronchoscopy was done transorally to avoid contamination by nasal lesion. Nodular lesions were found over the entire trachea bronchial tree. Bronchial wash and endobronchial biopsy were taken from the nodular lesions. Fine needle aspirations were done from the swellings in his leg and nose.All our samples including the bronchial wash and endobronchial biopsy specimen were positive for

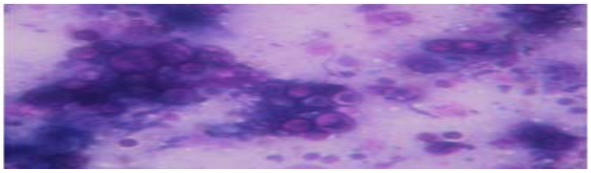


Fig 5

Rhinosporidium spores were not seen in peripheral venous blood or urine. His nasal and leg swellings were surgically excised. The lung lesion was not touched due to high risk for surgical intervention. The patient was started on Tab Dapsone 100mg od and is under our follow up.

Discussion

Introduction

Rhinosporidiosis has been known for over a hundred years since its first description in Argentina. It is a chronic disease, with frequent recurrence after surgery, and occasional dissemination from the initial focus which is most commonly seen in upper respiratory sites. It occurs universally, although it is endemic in south Asia, notably southern India and in Sri Lanka.

Epidemiology

Rhinosporidiosis is an infective disease in the sense that the tissue lesions are always associated with the presence of the pathogen. However there has been no documented evidence of crossinfection between members of the same family or between animals and humans.In addition to cases in humans, Rhinosporidiosis has also been documented in several species of farm, domestic and wild animals like cattle, buffaloes, dogs, cats, goats, horses, mules, several species of ducks, swans, geese and water fowl.The great majority of cases are sporadic. A curious feature in the incidence of the disease is that while several hundreds of persons bathe in the stagnant waters, as in India, only a few develop progressive disease; this might indicate the existence of predisposing factors in the host.

Mode of infection

The presumed mode of infection from the natural aquatic habitat of Rhinosporidium seeberi is through the traumatised epithelium 15 most commonly in nasal sites.

Mode of spread

Auto-inoculation Auto-inoculation was considered by Karunaratne16 in his classical monograph on Rhinosporidiosis, to be the explanation for the occurrence of satellite lesions adjacent to granulomas especially in the upper respiratory sites and for local spread. Spillage of endospores from polyps after trauma or surgery is thought to be followed by auto-inoculation through the adjacent epithelium. Haematogenous spread There is evidence for haematogenous spread of Rhinosporidiosis to anatomically distant sites12. The development of subcutaneous granulomata in the limbs, without breach of the overlying skin, could be attributed to such haematogenous dissemination, from a subclinical, upper respiratory focus of infection. Lymphatic spread Ashworth17 suggested the possibility of lymphatic spread but none of the numerous reports on the histopathology of localised or disseminated Rhinosporidiosis has described the occurrence of lymphadenitis. One reason could be that histological examination of the lymph nodes, especially in disseminated cases, has apparently not been done. The apparent rarity of lymphadenitis in Rhinosporidiosis, contrasting with its frequency in systemic mycotic disease, is remarkable; this rarity might be related to the diverse mechanisms of immune evasion by Rhinosporidium seeberi.

Clinical features

The great majority of cases occur in upper respiratory sites, notably the anterior nares, the nasal cavity - the inferior turbinates, septum and floor. Posteriorly, rhinosporidial polyps occur in the nasopharynx, larynx, and soft palate; the buccal cavity is only rarely affected. Dissemination to the limbs, trunk and viscera has been described in a few cases, with a rare fatality especially when the brain was involved. In rhinosporidial lesions in the limbs, a notable to surgery.feature has been the destruction of underlying bone. Rare cases of spontaneous regression of nasal polyps have been recorded.

Pathology

Characteristically, rhinosporidial lesions in the nasal passages are polypoidal, granular and red in colour due to pronounced vascularity, with the surface containing yellowish pin head sized spots which represent underlying mature sporangia. A covering of mucoid secretions is not uncommon. Naso pharyngeal polyps are often multi-lobed with a variegated appearance, with typical strawberry like regions and other areas which have relatively less vascular lobes with smooth surfaces. Polyps on the face and trunk could simulate verrucous warts, and are either pedunculated, or sessile on broad bases.Rhinosporidial granulomas in disseminated cases occur as subcutaneous lumps with unbroken skin. These sometimes present clinically as ulcerated growths which could mimic malignant lesions such as sarcomas and carcinomas.In most histological sections, the organism is present in all stages of its development. The stroma which is either fibro-myxomatous or fibrous contains chronic inflammatory cells which include macrophages and lymphocytes, while neutrophils are numerous around free endospores. In granulomatous tissue, giant cells occur often within sporangia and in the stroma. Fibrosis is prominent, notably, in non-respiratory sites. A noteworthy feature is the variability of stromal and cellular reactions even within tissues from a single patient.

Diagnosis

Histopathology The definitive diagnosis of rhinosporidiosis is by histopathology on biopsied or resected tissues, with the identification of the pathogen in its diverse stages, rather than the stromal and cellular responses of the host.Cytodiagnosis

Cytodiagnosis on aspirates from rhinosporidial lumps or on smears of secretions from the surfaces of accessible polyps and fine-needle aspirates from lumps provide, with suitable stains, distinctive diagnostic features. Caution was however recommended18 with smears of material from respiratorysites, in the identification of endospores; which could be confused with epithelial cells, especially from the naso-pharynx, in which the residual cytoplasm and large nuclei might simulate the residual mucoid sporangial material around the endospores (referred to as 'comet' forms by Beattie) 20, and the endospores themselves, respectively. The Periodic acid-Schiff stain will discriminate between these, as the endospores stain markedly magenta while the epithelial cells are PAS-negative.

Treatment

Although cases of spontaneous regression have been recorded, they are rare, and the mode of treatment remains surgical. Total excision of the polyp, preferably by electro-cautery, is recommended. Pedunculated polyps permit of radical removal while excision of sessile polyps with broad bases of attachment to the underlying tissues is sometimes followed by recurrence due to spillage of endospores on the adjacent mucosa. Extensive growths, as on the penis, might require amputation of the affected site. While several anti-bacterial and anti-fungal drugs have been tested clinically, but unsuccessfully, the only drug which was found to have some anti-rhinosporidial effect is dapsone13, 20 which appears to arrest the maturation of the sporangia and to promote fibrosis in the stroma, when used as an adjunct

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