Case study

An anesthesiologist's perspective of disseminated cutaneous rhinosporidiosis; 'Block resection' of the lesions!

ABSTRACT :

Rhinosporidiosis is a chronic granulomatous disease caused by a fungus-like organism, Rhinosporidium seeberi, which is endemic in India and Sri Lanka. Polypoidal nasal mass is the most common clinical manifestation, with scattered case reports of lesions in the nasopharynx, eye (nasolacrimal duct) and lower aerodigestive tract. Isolated disseminated cutaneous lesions, though reported, are very rare.

Chronic recurrent disseminated cutaneous rhinosporidiosis poses novel challenges for the anaesthesiologist in the perioperative period, right from simple issues such as intravenous access to the more pressing concern of choosing an anaesthetic technique tailored to the wide variety of clinical presentation and hence planned surgical intervention. This case report aims to throw light on the path less travelled - management of such a rare and challenging patient through the eyes of an anesthesiologist.

Keywords : Rhinosporidiosis , disseminated , anesthetic management.

INTRODUCTION :

Rhinosporidiosis is a chronic granulomatous disease caused by a fungus-like organism, Rhinosporidium seeberi, which affects primarily the mucous membranes of the nose and nasopharynx. The causative agent is taxonomically placed in "DRIP" clade (Dermocystidium, Rosette agent, Ichthyophonus, and Psorospermium) which has been renamed as Mesomycetozoea.[1]

The biological agent has a mature stage that consists of large, thick-walled spherical structures called 'sporangia' which contain smaller 'daughter-cells' called 'sporangiospores' and it can be visualized with fungal stains such as Gomori Methanamine Stain, Periodic Acid Schiff and even standard Hematoxillin & Eosin staining.

Rhinosporidiosis is endemic in India and Sri Lanka and more commonly seen in males [2]. The transmission occurs through dust, infected clothing and fingers and through swimming in infested

stagnant water, when spores come in direct contact with nasal mucosa. Polypoidal nasal mass is the most common clinical manifestation, with scattered case reports of lesions in the nasopharynx and lower aerodigestive tract [3]. Surgery is the mainstay of treatment but excision poses a challenging problem of recurrence. Isolated disseminated cutaneous lesions, though reported, are very rare [4-9], more so, in immunocompetent individuals, and causing negligible morbidity. Disseminated disease has been primarily attributed to hematogenous spread. Additionally, the delayed presentation at extra nasal sites may pose a diagnostic dilemma. Our case report contains an amalgamation of all the abovesaid sporadic entities which we believe would make for a compelling scientific read and valuable addition to existing literature.

CASE REPORT:

A 36-year-old male ASA 1 patient, presented to the outpatient department with complaints of bleeding associated with purulent discharge from a mass in the left forearm since two days. A detailed history revealed that the mass had been present since 1 year which gradually increased in size and started bleeding 2 days back. The patient mentioned that he was diagnosed with nasal rhinosporidiosis 25 years ago and was operated for the same 5 times (last 10 years back) for excision of the nasopharyngeal mass lesions and was also tracheostomized 6 years ago in view of respiratory obstruction. He recovered well post-surgery and currently had no signs of respiratory distress. The patient presently only had complaints of multiple nodular swellings all over the body. The nodules, which appeared 4 years ago and have been gradually increasing in size, were present over his face, both arms and forearms, peri umbilical region, back, and bilateral lower limbs. Wide local excision for the cutaneous lesion over the right arm was done one year ago when the nodule became a bleeding ulcero - proliferative growth and cutaneous rhinosporidiosis was diagnosed post excision biopsy. The patient had infection localized in the nasal mucosa for greater than a decade and despite multiple surgeries, he showed progression, recurrence and dissemination of the disease in the form of widespread cutaneous lesions all over the body.

On thorough examination, there were large necrosed friable swellings on the right arm (10x8 cm) (Figure 1) and left forearm(5x7cm) associated with bleeding. Apart from these there were lumpy, verrucous necrosed nodules on the back, umbilical area, and in the lower limbs. All these swellings had developed over the last few years (Figures 2& 3). On inspection, the lesions were lying singly as well as in clusters with moist and shiny surfaces and a few of them showed hemorrhagic crusts. On palpation, these lesions were granular, polypoid fleshy masses, which were firm, smooth, non-tender and mobile with well-defined margins.

Figure 1- Necrosed lesion on Right arm

Figure 2- verrucous fleshy nodules in para umbilical areas

Figure 3- crusted lesions with nodules in right lower limb

There was no lymphadenopathy. X-rays of shoulder and chest were normal as was ultrasonography of abdomen. Screening for human immunodeficiency virus was negative. Rest all routine blood investigations were normal.

Wide local excision was planned for all cutaneous lesions involving arms and legs.

The patient has a history of multiple surgeries and disease recurrence over a span of 25 years. The major goal in present scenario was to excise as many lesions as possible. The first challenge posed was intravenous cannulation as the patient had cutaneous nodules in bilateral upper limbs and both lower limbs were swollen. Hence vascular access was secured by inserting a central line in the right internal jugular vein prior to the surgical intervention. The second major challenge was plan of anaesthesia in view of history of tracheostomy and multiple nasopharyngeal surgeries. There was a possibility of difficult intubation along with increased infection risk through iatrogenic inoculation as he had disseminated cutaneous lesions all over the body.

In view of the above concerns, regional anaesthesia was concluded to be the safest choice for this patient. In order to offer anesthetic coverage to both forearm and shoulder region, combined interscalene and supraclavicular brachial plexus block was given. Subarachnoid block was administered for excision of lower limb lesions.

DISCUSSION:

The cutaneous form of Rhinosporidiosis was initially described by Forsyth. Bathing or working in stagnant contaminated water harboring the infectious agent are the biggest risk factors for contracting the disease.[8] Rhinosporidiosis is more prevalent in tropical climates, such as those of South India and Sri Lanka. [9] According to Sudarshan et al., it is also endemic in Central India in Chhattisgarh. [10] The pathogen enters the body via traumatized epithelium (i.e., transepithelial infection), most commonly the nasal mucosa. Endospore penetrates the nasal cavity mucosa and matures into sporangium in the submucosa.[9] No person-to-person transmission is noted. The disease can manifest itself in four forms – (1) Nasal, (2) Ocular, (3) Cutaneous and (4) Disseminated. Rhinosporidiosis has a male preponderance (M: F = 4:1), with most patients presenting between the second to fourth decade of life [3].

The most common sites of infection are nasal cavity and nasopharynx, followed by the eye. Occurrence is rare in the larynx, trachea, lungs, urethra, genitalia, lacrimal sac, and bones.[10] Clinical presentation of Rhinosporidiosis in an isolated widespread disseminated cutaneous distribution throughout the body, with no concurrent nasal or laryngotracheal symptoms, is even rarer. [4-9] Disseminated rhinosporidiosis is usually seen in immunocompromised patients and can have high mortality.[11]

The cutaneous lesions in Rhinosporidiosis usually start as friable papillomata that later become pedunculated. It may also present as warty papules and nodules with whitish spots, crusting and bleeding on the surface. The term 'Dermosporidiosis' has been recently suggested to denote the predominant skin lesions of the disease [12]. Presentation of rhinosporidiosis as a tumour, ulceroproliferative growth or arthropathy may cause diagnostic dilemma. The cutaneous lesions occur due to auto-inoculation or due to hematogenous spread.

Implantation of R. seeberi spores into a traumatized nasal mucosa is very likely in a habitual pond bather, coupled with intermittent nose picking. In our patient, the nasal disease recurred several times after surgical removal, which may represent regrowth of residual Rhinosporidiosis. It is quite surprising that despite a persistent presence of behavioral risk factors in this patient, the nasal disease remained localized for more than a decade before dissemination. The dissemination was most probably due to hematogenous spread, as depicted by tumoral lesions of the shoulders, arm, thigh, leg and wrist, affecting the soft tissues without initial involvement of the overlying skin.

The diagnosis of rhinosporidiosis can be made by FNAC but this may also be inconclusive.[13] The 'gold standard' for rhinosporidiosis diagnosis is histopathological identification of the pathogen, showing different stages of maturation, in biopsied or resected tissue.

The next challenge encountered is the preoperative anesthetic evaluation prior to surgery. These patients in case of extensive involvement may present with airway obstruction that can either be symptomatic or asymptomatic. In case of asymptomatic patients having suspected laryngeal involvement, we must anticipate difficult intubation whereas in case of patients in distress, emergency tracheostomy is done. In the present scenario, the patient gave history of multiple nasal surgeries in a decade long span along with the history of being tracheostomized once. Since this patient had a history of vigorous airway handling in the past and was currently having disseminated cutaneous lesions, general anesthesia was avoided and the best modality of anesthesia was considered to be regional anesthesia.

CONCLUSION:

Disseminated rhinosporidiosis is extremely rare. Although autoinoculation of R. seeberi spores by infected fingernails may explain the occurrence of rhinosporidiosis in various anatomically unrelated parts of the body, the involvement of the soft tissue mass with intact overlying skin suggests the possibility of additional spread of rhinosporidiosis via a hematogenous route. Rhinosporidiosis presents a therapeutic challenge amenable primarily to surgical excision with the necessity of prolonged adjuvant drug therapy.

The chronicity of the disease coupled with its propensity for recurrence means the anaesthesiologist may encounter patients with history of multiple surgeries and therefore previous interventions to the

airway, adding to the preexisting challenging anaesthetic concerns (eg: difficulty in securing an intravenous line, a depressed patient on medications in whom renal function and hemogram need to be taken into account). A wide variety of presentation means the anaesthetic technique needs to be individualized to a particular patient and often improvised, as in our case. Taking all factors into consideration, this patient was operated under regional anesthesia, under brachial block for the arm forearm lesions and subarachnoid block for lower limbs.

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