

Rhinoscleroma with Malignancy- A Case Report and Review of Literature

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Abstract

Rhinoscleroma (scrofulous lupus, scleroma) is a rare, chronic, specific granulomatous disease of the nose and upper respiratory tract. It occurs most frequently in young female adults between the ages of 10-35 years in low socioeconomic class in developing countries. The course is usually chronic, relapse can occur and it is non-neoplastic. Inflammatory compressive destruction of bone and soft tissue may occur and thus could lead the clinician and radiologist to suspect malignancy. We report a 25-year-old woman who developed undifferentiated tumor three months after initial diagnosis of RS. She was the first case of RS with malignancy in our institution.

Keywords: Rhinoscleroma; Malignancy

Introduction

Rhinoscleroma (RS) is an uncommon chronic, specific granulomatous infectious disease caused by *klebsiella rhinoscleromatis* (*Kr*) a gram-negative diplobacillus [1-3]. The disease is endemic in Tropical African, South East Asia, Mexico, Egypt Central and South America Eastern and central Europe [4-7]. Its importance as a public health problem in developing countries is underestimated due to paucity of epidemiological data and literature on this disease [8]. Sporadic cases of RS have been reported in U.S and Saudi Arabia because of migration from endemic areas [9-11]. Therefore, RS is an important differential diagnosis in immigrants from the endemic regions suffering on chronic obstructive, pseudoneoplastic inflammations in the upper airways [8]. It is most frequently recognized in adolescents and young adults female and less than 10% of children were reported in the literature [2,12,13]. The case presented was a young woman whose nasal swab and biopsy confirmed RS. Three months after initial diagnosis she relapsed, a repeat tissue biopsy; histopathology confirmed malignancy. We want to raise awareness that inflammatory compressive destruction of bone and soft tissue in RS could suggest to the clinician and radiologist to suspect malignancy.

Case Report

A 25- year old female, a groundnut farmer referred from a peripheral hospital to our Clinic with a three and a half months history of gradual onset but progressively worsening bilateral nasal obstruction and yellowish offensive nasal discharge, anosmia, nasal tone of voice, mouth breathing, snoring and weight loss. There was no epistaxis or symptoms of nasal allergy. However, she had sores on her palate with odynophagia. She neither smoked cigarette nor snuffed tobacco.



Figure 1: Patient at Initial Presentation (Front View)

General physical examination revealed chronically ill looking patient, mild palour with *Tenea coporis* affecting the face, chest and upper limbs. No cervical lymphadenopathy. The external nose was asymmetrically (Figures 1 and 2).



Figure 2: Patient at Initial Presentation (Side View)

Rhinoscopy showed mucopurulent rhinorrhoea, with yellowish crusts and contact bleeding on the walls of both nasal cavities. There

was an ulcer on the left side of the posterior ½ of the hard palate (Figures 3 and 4).

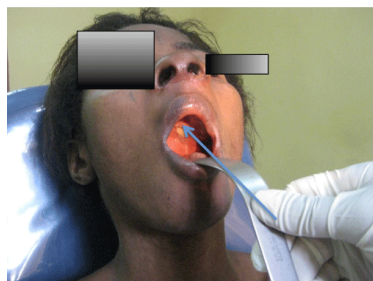


Figure 3: Shows Oro-nasal Fistula in the Patient

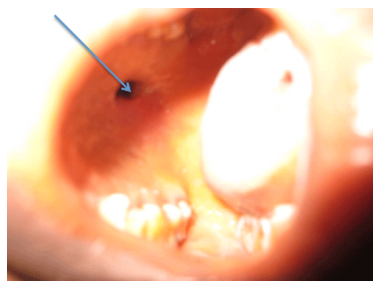


Figure 4: The Same Patient with Oro -Nasal Fistula

Plain radiographies of paranasal sinuses revealed only features of chronic rhinitis. CT scan and MRI Paranasal sinuses were considered but she could not afford the cost. She was HIV- negative and VDRL non-reactive. Nasal swabs microscopy culture and sensitivity [M/C/S] yielded growth of *klebsiella species* sensitive to Roicephin, and Zinnat and Carbencillin. Clinical diagnosis of RS was made. She was treated according to sensitivity pattern. Tissue biopsy was taken under general anaesthesia and histopathology result confirmed RS (Figures 5 and 6). Patient however defaulted from treatment because of financial constraints and resorted to self-medication.

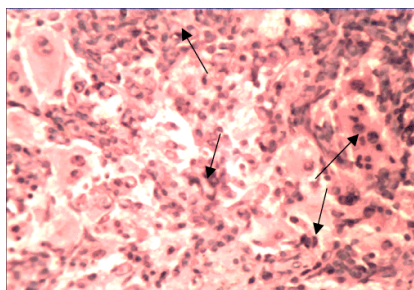


Figure 5: Histological section of the tumour showing diffuse infiltration by predominant chronic inflammatory cells mainly lymphocytes (arrow) and plasma cells

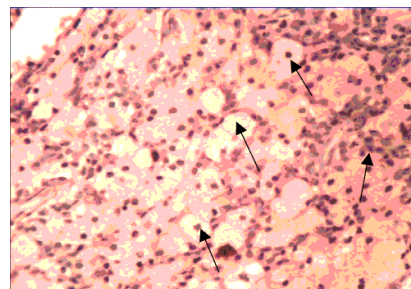


Figure 6: Histological section of the tumour showing reactive histiocytic cells with abundant eosinophilic cytoplasm (Milculitz like cells) (arrow) amidst mature lymphocytes

Three months later, the symptoms relapsed with extensive destruction of soft tissue, septal cartilage and bone of the face and oronasal fistula on the left side of the hard palate (Figure 7). Clinical diagnosis of RS to rule out malignancy was made.



Figure 7: Re-presentation of the patient with ulcer extending to the left upper lip three months after initial diagnosis and treatment

Repeated tissue biopsy confirmed malignancy (Figures 8 and 9).

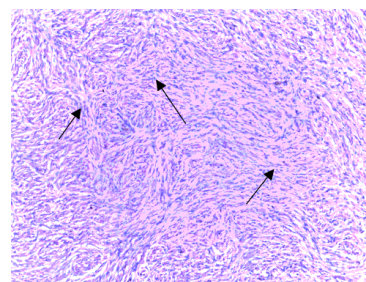


Figure 8: Photomicrograph of the anaplastic lesion from the lesion having predominantly spindle shaped cells (arrow)

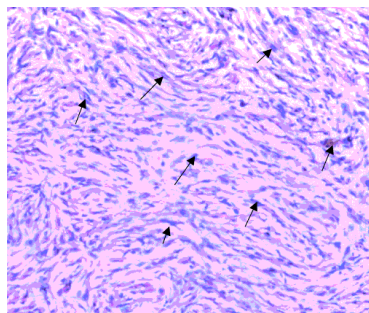


Figure 9: Histology section showing spindle shaped cells with hyperchromatic nuclei and irregular nuclear outline and coarse chromatin pattern (arrow)

Immunohistochemical stain was requested for as diagnostics assistance but could not be done because patient could not pay for it. Radiotherapy was recommended and patient was referred to appropriate Centre. However, the patient did not go for radiotherapy but discharge against medical advice and thereafter she was lost to follow up.

Discussion

RS, a granulomatous disease, was first described in Austria and Germany [8]. von Hebra [6], Viennese dermatologist, originally described the disease and gives its name in 1870. In 1877, von Mikulicz [5,14] described the histological features of this disease and established its non-Neoplastic, inflammatory nature. In 1882 Von Frisch [15] identified a Gram- Negative encapsulated bacillus, which could be isolated from the nasal lesions of patients with scleroma [3]. The standards of therapy of RS at first time were discussed on the international congress of Otorhinolaryngology in 1932 in Madrid [8].

RS is transmitted by means of direct inhalation of droplets or contaminated material and humans are the only identified host. It has affinity for mucosa of nose; usually starts in the subepithelium of vestibules spreading to other area such as nasopharynx, oral cavity, paranasal sinuses; maxillary sinus being the most commonly affected, larynx and trachea and bronchi [2,12,16-19] in a progressive and often-destructive fashion [1,6] rarely orbit and cervical nodes. Almost all patient of RS have nasal involvement with nasal obstruction [94%] nasal deformity [32%] and epistaxis [11%] being presenting feature. The disease involves oral cavity in 18%, Pharynx in 18-43%, Eustachian tube 27%, Larynx 26% trachea and bronchi in 10% cases. The nose involved in almost all cases; 95-100%, yet the reported rate of sinus involvement is low with maxillary sinus being involved in about 22% cases [20]. Sinus involvement may also be due to secretion retention secondary to ostium blockage in the nasal cavity [1,21,22]. Our patient's disease started from the nose then spread to right maxillary sinus, upper lip, oral cavity and the pharynx.

The peak age of presentation of RS is 2nd and 3rd decade [12]. The female sex predilection has been reported in some literature; ratios of 13:1 [12,23,24] while other report no sex preponderance [2,8]. The case reported was a 25 -year old female.

Clinical presentation of RS is often non-specific resembling that of chronic rhinitis; hence symptoms may be on for over 10 years prior to diagnosis [25]. Symptoms depend on the anatomical site affected,

ranging from nasal obstruction, dysphagia to severe respiratory distress or asphyxia [8]. The rate of occurrence [infection and transmission] is probably associated with poor hygiene, crowded living environment and malnutrition [1,6,12,19,23,26] and host susceptibility even in immune-competence subjects [6,23]. Thus, the disease condition is also known as the disease of Great Unwashed [27]. The index patient was from low socioeconomic group. She was a groundnut farmer married to a palm wine tapper. She lived in a room apartment with her husband and five children.

RS manifests in three progressive but ill-defined stages: exudative, proliferative and cicatricial [1,2,17]. The exudative stage, the first stage is characterized by abundance of polymer-mono-nuclear leukocytes and cellular debris [27,28] the symptoms resemble that of infective rhinitis. Often present with purulent rhinorrhoe that is foul smelling [10]. Diagnosis often missed at this phase but high index of suspicion could lead to early diagnosis.

Proliferative stage [hypertrophic, granulomatous], the second stage characterized by appearance of granulomatous nodules, deformity and functional difficulty. This stage takes months to years.

The third stage is cicatricial [fibrotic, sclerotic, scarring] stage. This phase is marked with extensive scar formation [17,27] with abundance of connective tissue and paucity of Mikulicz cells and plasma cells [27,28]. Our patient was seen in the later exudative stage and progressed to the proliferative stage. Diagnosis of RS is made by isolation of Kr from nasal swab culture in MacConkey agar and through histology evidenced by the presence of Mikulicz cells [20,29]. Demonstration of Kr in nasal culture is diagnostic since this not among the normal nasal flora. The immunoperoxidase technique is highly sensitive and specific in identifying Kr organism [20]. Polymerase chain reaction has been described as useful in confirming the diagnosis of RS [30]. The index case had swab taken from her nose and m/c/s yielded Kr on which clinical diagnosis of RS was made.

It is known that granuloma spreads mainly in the soft tissues and in the connective tissue spaces and that it does not invade bony structures. Sometimes inflammatory compressive destruction of bone and soft tissue may occur and thus should lead clinician and radiologist to suspect malignancy [31]. Bonacina et al. in their case report of RS in immigrant from Egypt had extension of disease into the ethmoidal sinuses. They used MRI to monitor the progress of treatment of their patient. There was no histopathology report of malignancy in their case. Also Razeq et al. in their article on MR Appearance of RS reported of some bone resumption but no associated malignancy was reported. Our case reported developed extensive destruction of soft tissue of left side of her face with exposure of bony structures of the left nasal cavity, maxillary antrum and palate. At this point we considered a repeat tissue biopsy necessary. The second histology report show malignant proliferation of spindle to oval shaped cells in a dense and compact stroma. The cells are with irregular nuclear edges and coarse chromatin pattern. There is mitotic activity of 4-5 per high power field. There are also foci of chronic inflammatory cells infiltrate mainly mature lymphocytes. A diagnosis of undifferentiated tumour was made (Figures 8 and 9).

It is not recorded that RS assumes malignant characters or predisposes to malignant change [32]. Atta reported two cases of RS with malignancy that clinically malignancy was not suspected. Is malignancy and RS coincidence finding? Or the malignancy occurs as a result of chronic irritation and sepsis produced by RS and which, as we know, lead to squamous metaplasia of the covering nasal mucosa,

in the long run to malignant change in this epithelium. Does RS have malignancy transformation tendency [32]?

However, RS can mimic fungal infections, other nasal granuloma such as tuberculosis, leprosy, vasculitis, Wegner's disease, saiciodosis, and verrucous carcinoma [10,12,17,30,33]. The following are other differentials, basal cell carcinoma, mucocutaneous leishmaniasis, rhinosporidosis, sporotrichosis, blastomycosis and paracoccidioidomycosis [30].

Clinical remission and relapse is a common finding in RS [30]. Treatment option depends on the stage at presentation. RS recurrence rate is high therefore a long duration of antibiotics treatment is advisable [23]. The following recommended antibiotics have been reported to give a good outcome; streptomycin, doxycycline/tetracycline, second and third generation of cephalosporines, ofloxacin, rifampicin and sulfonamides [19,23,34]. Ciprofloxacin also reported to offer complete resolution of RS [2,23,33,34,35]. Recommended antibiotics treatment duration varying from six weeks to six months [2,23]. Topical antibiotics like acriflavin and 3% rifampicin ointment has been reported with resolution of symptoms [23,36]. The case presented had initial treatment with Zinnat for 2 weeks and change to streptomycin because of poor finance. She defaulted, only to reappear 3 months later with recurrence. RS is usually not fatal when diagnosed early and appropriate antimicrobial treatment instituted promptly. However patient may die from respiratory obstruction or intracranial extension [10].

Conclusion

Although RS often mimics malignancy it often respond to treatment if diagnosis is made early and treated. There is need for high index of suspicion in our environment when ruling out malignancy.

Consent

Informed consent was obtained from the patient relative for the case report to be published.

Conflict of Interests

The authors state that they have no conflict of interests to declare regarding the publication of this paper.

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