CASE REPORT

Rhinoscleroma Revisited

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ABSTRACT

Rhinoscleroma (more appropriately 'scleroma') is a chronic, disfiguring inflammatory lesion that is rarely encountered in the present-day otolaryngology practice. The diagnosis often becomes difficult especially when it recurs and presents late with complications. This report illustrates the clinical and histologic features of rhinoscleroma in a defaulter patient who represents advanced-stage disease with orbital and intracranial extensions. Special emphasis has been provided on the computed tomography impressions such that the evolution of scleroma with time can be studied.

Keywords: Intracranial extension, Mikulicz cell, orbit, rhinoscleroma, scleroma.

INTRODUCTION

Rhinoscleroma is a chronic, recalcitrant disfiguring lesion caused by *Klebsiella pneumoniae* subsp. *rhinoscleromatis* that is seldom encountered in the present-day clinical practice. The disease is endemic to the arid tropical regions of South America, Eastern Europe, the Middle-East, and some parts of Asia and Africa (1, 2). In spite of a surge in immigration, the restricted geographic limit, and, more importantly, susceptibility to the fluoroquinolones (3) have rendered the disease as one of the rare pathologies today (1). This report illustrates a classic case of scleroma that initiated in the nasal cavity and had subsequently spread to the orbits with intracranial extension.

CASE REPORT

A 48-year-old farmer presented with blindness, right-sided proptosis, headache, nasal obstruction and facial deformity. He had history of surgery three times in our hospital; a biopsy from a nasal mass 8 years previously suggested rhinoscleroma. He was prescribed oral ciprofloxacin; however, he was a defaulter, and was lost to follow-up several times. As the disease progressed, he underwent excision of the nasal mass from the left and right sides 8 and 6 years back, respectively, followed by enucleation of his left eye 4 years back. Histopathology in all three instances suggested rhinoscleroma.

He had intermittent headache, nasal obstruction and progressive protrusion of the right eye since 1 year. His vision also deteriorated since 2 months. He suffered from occasional running nose and stuffiness for 4 years prior to his first surgery. His right eyeball was fixed and hard, with scleral oedema and exposure keratitis (Fig. 1), and had no perception of light. The perinasal facial skin appeared glistening and taut due to fibrosis.

Computed tomography (CT) scan (Fig. 2) revealed an enhancing mass occupying both sides of the nasal cavity, with involvement of the anterior cranial fossa. The right eyeball was pushed anteroinferiorly due to the mass effect. Bone windows showed diffuse moth-eaten appearance of the entire facial skeleton, especially those forming the sinus walls, with prominent sclerotic areas. While the soft-tissue characteristics remained unchanged, sclerosis and remodelling of the facial skeleton were more prominent when compared with the previous scans (Fig. 2).

Diagnostic naso-endoscopy revealed smooth-walled rubbery lesions bulging in the lateral nasal walls (Fig. 3).

Biopsy from the right lateral nasal wall showed subepithelial inflammatory infiltrates, chiefly plasma cells and monocytes, with occasional Mikulicz cells and Russell bodies, in a grossly fibrocollagenous stroma (Fig. 4).

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Considering the orbital and intracranial extensions, we consulted the ophthalmologists and neurosurgeons for appropriate surgical interventions. However, the patient refused to give consent for further surgery, and

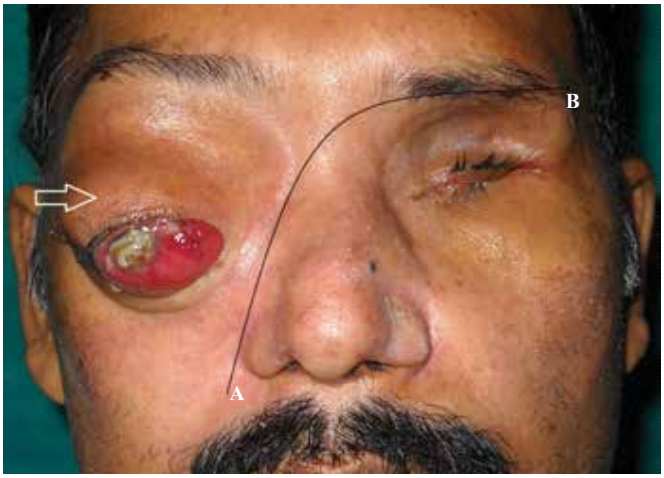

Fig. 1: The clinical appearance of the patient: He presented with right-sided proptosis with fixed, hard globe, which was pushed anteroinferiorly by the mass occupying the orbit (arrow). There was scleral chemosis and exposure keratitis. Note the right-sided malar fullness and raised eyebrow suggesting a sino-orbital lesion. The perinasal region had a glazed appearance due to fibrosis. The alae and alar grooves were collapsed with stenosed, triangular alar rim; the dorsum of the nose was depressed. The imaginary line (AB) divides the disease activity into two zones: the area below AB denotes predominance of cicatrization, while the area above it represents the proliferative stage.

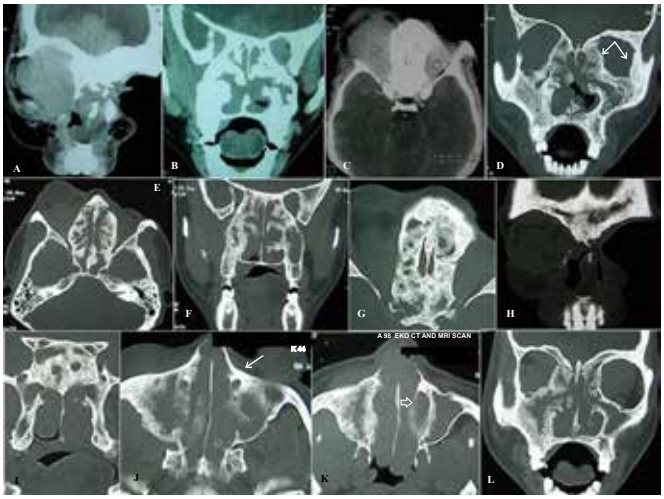

Fig. 2: Computed tomography (CT) features of the facial skeleton (A–I: present scans; J–L: earlier scans): CT showed enhanced mass lesion in the nasal cavities, orbits (A–D) and intracranial compartment (A, B). The facial skeleton had a peculiar moth-eaten appearance with areas of sclerosis and resorption, suggesting prolonged inflammatory process. This was more prominent in the perisinus bony framework, involving maxilla (D), ethmoids (E–G), frontal (H) and sphenoids (I). Note the porous, sieve-like appearance of the cribriform plate (G)—the potential route for intracranial spread in this patient. Also note the areas of sclerosis around the left orbit (D) and nasolacrimal duct (J) (arrows). There was characteristic involvement of the left middle turbinate (K) (hollow arrow) with destruction of a large part of the septum (D). A comparison between the present and previous scans (D and L, respectively) showed a gradual decrease in the sinus spaces due to progressive bone remodelling and mass effect.

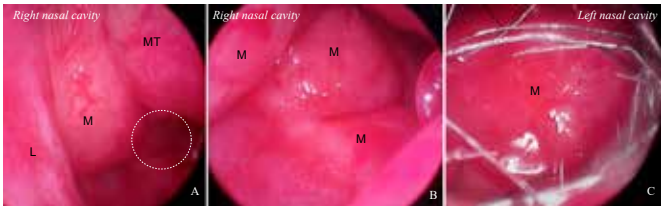

Fig. 3: Diagnostic naso-endoscopy before taking the biopsy: Naso-endoscopy showed thick rubbery submucosal lesions mostly in the lateral nasal wall (A). The scope could not be progressed beyond a point (area within the dotted circle in A, viewed closer in B) where the mass almost occluded the nasal lumen. It filled up the nasal cavity just beyond the vestibule on the left side (C). M = mass; MT = middle turbinate; L = lateral nasal wall.

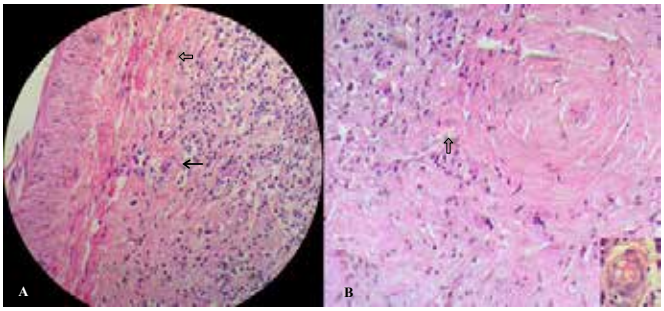

Fig. 4: Histopathologic features of rhinoscleroma: There was dense aggregate of subepithelial inflammatory cells (A) but with chiefly fibrocollagenous stroma signifying a cicatricial stage (B), with relatively scanty Mikulicz cells (solid arrow; inset) and Russell bodies (hollow arrows). Haematoxylin-eosin; ×400 (inset: ×1000).

was quite depressed on his present status of health. He is presently under a counselling program in our psychiatry department, and has been put on long-term oral ciprofloxacin.

DISCUSSION

A classic illustration of rhinoscleroma in advanced stage is presented here. The term ‘scleroma’ seems more appropriate as the disease, almost always originating in the nasal cavities, often progresses beyond. Mostly the spread is contiguous in caudad direction, involving pharynx, the laryngotracheal complex and lungs (4). However, cephalad extension into the orbits and intracranial compartment is rare (2).

A unique characteristic noted here is the dynamicity of the disease, and that the existence of the three stages (catarrhal/atrophic, proliferative/granulomatous, cicatricial) is not mutually exclusive. While the nasal component here was chiefly fibrotic, the disease was in its proliferative stage in the right orbit, and perhaps in the anterior cranial fossa (Fig. 1). The facial skeleton showing extensive remodelling was intermediate between the two, with areas of sclerosis coexisting with the osteogenesis–osteolysis couplet. This explains its peculiar fluffy, irregular, heterogeneous appearance with

a sort of malleability such that the contour of the maxillary and ethmoid sinuses was compromised due to the lesion's mass effect. Progressive osteolysis and sclerosis had also made the facial bones, especially the perisinus and periorbital framework, more pervious (Fig. 2). We assume this might explain the intracranial spread through the porous cribriform plate, and to the orbits through the lamina papyracea, apart from the natural conduits of superior orbital fissures and nasolacrimal ducts.

Histopathology depicted predominantly cicatricial stage with fibrocollagenous stroma, but also had appreciable inflammatory cells signifying active disease process. This was evident clinically as residual fibrotic lesion in the nose and left orbit, with the disease in the proliferative stage in the right eye. We considered the left naso-orbital lesion as residual, because the patient was a defaulter and surgical excision might not be exhaustive. Also, Mikulicz cells and Russell bodies were relatively less, and granulomas were scarce. This could be due to the predominant scarring of the endonasal lesion. Alternatively, deficiency of interleukin-10, the key molecule that recruits and matures the histiocytes (future Mikulicz cells with phagocytic property), may result in ineffective containment of *K. rhinoscleromatis*, exposing host tissue to disease invasion (1). This could also explain the unusual aggressive nature of the disease in our patient.

Evidently, scleroma is difficult to eradicate, and although sensitive to quinolones, a prolonged drug therapy is needed (4). Thus, compliance to medication and proper long-term follow-up are required to check disease progression. Scleroma and its surgical treatment can be mutilating, and recurrence is common. Consequently, it has a significant impact on the patients' quality of life. As illustrated here, the disease is polymorphic and fleeting in character with contiguous spread, associated with characteristic alterations in imaging and histology. The diagnosis is challenging given its rarity, insidious onset and misleading presentation. Management needs to be individualized—but above all, the patience needed to deal with this slow-progressing disease is of utmost importance.

This article briefly recapitulates the cardinal features of rhinoscleroma from its clinical, imaging, pathologic (histologic and molecular aspects) and management perspectives. Seldom encountered in the present-day clinical practice, this chronic, recalcitrant disease entity has a high chance of being misdiagnosed, and as evident from this case description, still holds importance in the endemic regions. The present case of scleroma in its advanced stage with contiguous spread into the intracranial compartment and orbits, along with a prolonged past history in a defaulter patient, represents a completed spectrum of disease manifestation. This has made it worth presenting, both for its archival values and as a stand-alone reference illustration.

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AUTHORS' NOTE

Written informed consent has been obtained from the patient for publication of the clinical photographs. The necessary forms have been uploaded during submission of the manuscript and are available at the Editorial Office.

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