

RHINO-ORO-OPHTHALMO-CUTANEOUS LUPUS VULGARIS

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Two unusual modes of presentation of lupus vulgaris are being reported. In one case the lesion started in the nasal mucosa and spread to the nose, upper lip, gums, palate and naso-lacrimal ducts. In the second case the primary nasal lesion led to perforation of the cartilaginous nasal septum.

Key words : Lupus vulgaris, Skin tuberculosis, Nasal mucosa Perforation.

Tuberculosis is still a major health hazard in India, though its incidence has been declining with the advent of potent antitubercular drugs. Tuberculosis of skin though uncommon is not a rarity; its incidence has been reported to be ranging from 0.24%¹ to 0.59%² of all the skin ailments. Primary mucosal involvement however is extremely rare.

Two rare cases of lupus vulgaris (LV) primarily involving the nasal, buccal and lacrimal mucosa are being reported. One of the cases resulted in perforation of the nasal septum, a feature seldom seen these days.³



Fig. 1. Erythematous papulo-nodular lesions involving nose, upper lip, palatal and gingival mucosa (case 1).

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Case Reports

Case 1

Three years ago, a 9-year-old male child developed a small nodule on the inner side of his left nostril. It gradually spread to involve the surrounding skin of the nose and the cheek on the same side with ulcerations accompanied by oozing and crusting at places. In about a year these lesions spread to the upper lip, upper gingival margin and hard palate of the left side (Fig.1). He also had yellowish discharge from the left nostril but without any epistaxis. A diffuse swelling near the medial side of the left eye was also noted, on pressing which a purulent discharge flowed into the eye from the lower lacrimal punctum.

The lesions at present were well-defined erythematous papulo-nodules involving the columella, alae and tip of the nose, left side of upper lip, medial portion of left cheek, gingiva overlying the upper incisor teeth and extending upto the middle of the hard palate on the same side, interspersed with areas of ulceration and crusting. Diascopic examination of the papulo-nodular lesions revealed apple-jelly nodules. Left columella and the alae showed ulceration with oozing and crusting. There was a diffuse swelling near the medial canthus of the left eye with purulent discharge. Regurgitation test was positive. Left cervical lymph nodes were enlarged, soft, non-tender and mobile.

Routine tests on blood were normal, ESR was 22 mm., Mantoux test was highly positive

with blister formation. X-ray chest was normal and VDRL was non-reactive. Dacryocystogram revealed complete blockage at the junction of the lacrimal sac with the nasolacrimal duct. The facial lesion showed an acute flare up on doing the Mantoux test. Biopsies from the upper lip and the hard palate revealed extensive caseating epithelioid cell granulomata with several Langhans giant cells (Fig. 3). Antitubercular treatment with streptomycin and isoniazid led to complete clearing of the lesion within 2 months leaving behind atrophic scarring. At the end of follow-up and treatment for one year, dacryocystitis had completely subsided and there was no septal or palatal perforation.



Fig. 2. LV lesions over the nose and upper lip with a cartilaginous septal perforation (case 2).

Case 2

The second patient was a 14-year-old female with a diffuse swelling and purulent discharge from the medial canthus of the left eye for 8 years and erythematous-nodulo-ulcerative lesions on the margins of nostrils and upper lip with a hole in the anterior part of nasal septum for seven years. She had purulent blood-stained discharge from both the nostrils prior to formation of the septal hole. At the beginning of this episode she had received 15 injections and some tablets from a hospital which considerably

improved her symptoms, but she had discontinued the treatment. She remained asymptomatic for the subsequent six years, but noticed reappearance of the nodulo-ulcerative lesions in the nasal cavity, on the margin of both nostrils and on the skin of the right side of upper lip. Examination revealed a diffuse swelling near the medial part of the left eye, with a positive regurgitation test. There was an erythematous, nodulo-ulcerative, crusted lesion on the skin overlying the upper lip and extending into the margin of the alae nasi. Besides this, there were multiple, discrete, erythematous papules showing apple-jelly nodules on diascopy. Anterior rhinoscopy revealed a perforation approximately 0.75 cm diameter in the cartilaginous part of the nasal septum (Fig. 2). Nasal vestibules revealed thickening with nodules and crusts, more marked on the left side. There was cicatrisation of the lateral nasal wall and the upper margin leading to a tenting appearance of the anterior nares. Posterior rhinoscopy



Fig. 3. Epithelioid cell granulomas and Langhans giant cells in the dermis (case 1 H&E, X250).

revealed only mucopurulent discharge. There were no other abnormal oro-rhino-laryngeal findings. Eye examination was normal except for the presence of an abscess near the medial canthus of the left eye. Tonsillar and submental nodes were enlarged, firm, discrete, mobile and non-tender.

The other systems were normal. Her ESR was 30 mm. Mantoux test was 30 mm, with ulceration. X-ray chest was normal, and VDRL test was non-reactive. Biopsy from the nasal mucosa revealed typical caseating tuberculoid granuloma.

Comments

LV constitutes nearly 74% of all cases of cutaneous tuberculosis.⁴ On the other hand, nasal mucosal involvement in LV is unusual. In a review of the existing American literature on the subject, Havens⁵ found only 15 cases of primary nasal tuberculosis. In another large series, Rohwedder⁶ found no incidence of nasal involvement although the upper respiratory tract was found to be involved in 1.8% of cases.

This infrequent and rare involvement of the nasal mucosa is believed to be due to the bactericidal effect of the nasal secretions and to the mucocilliary barrier of the nasal mucosa⁷. Primary nasal infection in the occasional case is usually acquired either by droplet infection or by local implantations of *M. tuberculosis* by pricking the nose with contaminated fingers⁷.

Involvement of the oro-nasal mucosa due to LV poses a diagnostic problem on account of the wide differential diagnosis, like leprosy, cutaneous leishmaniasis, rhinosclerosis, sarcoidosis, blastomycosis, etc. Further, perforation of the nasal septum as seen in the second case is far rarer with LV than with diseases like leprosy, syphilis, etc.

The absence of bacilli in the tissues in both the cases and the flare up after a Mantoux test in case 1 indicate a high immunological state in both patients. This is at variance with the observations of earlier workers⁸ who found the immune status in oral mucosal tuberculosis to be depressed. Further, no roentgenographic evidence of pulmonary involvement was seen in either of the above cases although Laskaris and Nicilis⁸ found evidence of pulmonary tuberculosis in all the 4 of his cases of mucosal LV.

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