Otolaryngology online journal

ISSN 2250-0359

Volume 2 issue 1 2012

Title: LUPUS VULGARIS WITH LARYNGEAL LUPUS – A CASE REPORT

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Abstract :

Objectives : Tuberculosis ,though a common condition in our country is still an enigma because of its varied modes of clinical presentation,characteristics and spread. Our objective is to stress the importance of an complete clinical examination and analysis of the symptomatology in arriving at the diagnosis of laryngeal lupus and provide an update on current knowledge and treatment of lupus vulgaris with laryngeal lupus.

Case Report : A 23 yr old female presented with ulcerative lesion in the upper lip, extending to the nose and also lesions in naso and oropharynx and in the larynx .A provisional diagnosis of lupus vulgaris cuasing laryngeal lupus was made and histopathologically confirmed.Patient dramatically improved with anti tubercular treatment.

Conclusions : A thorough clinical examination, a strong suspicion and judicious use of investigations provided the clinical diagnosis of laryngeal lupus and which showed good improvement with latest regimen of anti tubercular treatment with complete cure in six months with out any significant residual scarring or disfigurement.

Keywords: lupus vulgaris,laryngeal lupus

Introduction

Tuberculosis is an airborne communicable disease that occurs after inhalation of infectious droplets expelled from patients with laryngeal or pulmonary tuberculosis during coughing, sneezing or speaking. The probability that a disease transmission will occur depends on the infectiousness of the patient, the environment in which the exposure takes place and duration of exposure. Because of improved living conditions, BCG vaccination and effective chemotherapy, the incidence and severity shows a downward trend. Yet we are in no position to say that an effective control over the disease has been achieved , there is a resurgence in its occurrence probably due to HIV and multidrug resistant strains.

Lupus Vulgaris is a chronic,progressive and tissue destructive form of cutaneous tuberculosis seen in patients with moderate or high degree of immunity. We are presenting a case of lupus vulgaris of face and nose that has spread back into nasopharynx palate and to the larynx,which is rare.

Case report

A 23 yrs old female presented with an ulcerative skin lesion with crusting over the upper lip and left nasolabial area for 1 yr duration. She had been treated by a general physician initially and was later attended by a general surgeon who has done a biopsy

from the skin lesion which was opined as papilloma by the pathologist. The general surgery people were also considering squamous cell carcinoma and basal cell carcinoma as possibilities and had sought surgical oncology and plastic surgery opinion for complete excision of the lesion and plastic repair. Since she also had a history of change in voice for 4 months the oncologist referred the patient to us for opinion. ENT examination revealed an ulcerative lesion over the upper lip extending into left nostril to involve the anterior end of nasal septum, floor and anterior end of inferior turbinate. Superficial ulcerative lesions were seen on posterior and left lateral walls of the naso pharynx, left side of uvula and soft palate. Videolaryngoscopy revealed irregular granular lesions in epiglottis(which was partially destroyed), ariepiglottic folds, arytenoids and false cords. Left vocal cord could not be visualized. Right vocal cord normal and mobile. Our first impression was tuberculosis of larynx, probably also responsible for other lesions. We wanted to rule out other granulomatous lesions like wegeners,syphilis and complete workout done; renal function tests were normal, VDRL negative, X ray chest showed no evidence of pulmonary tuberculosis, 3 consecutive sputum sample examination for AFB were negative. The dermatologist gave a differential diagnosis of lupus vulgaris, verrucous vulgaris regarding the skin lesion. We proceeded with biopsy from the lesions in the lip,pharynx and larynx. Histopathological examination revealed typical granulomatous tubercle with epitheloid cells, langhans giant cells and a mononuclear infiltrate with minimal caseation.

The lack of evidence for any primary pulmonary tuberculosis, the absence of pain, lesions in the anterior parts of larynx were all contradicting a diagnosis of laryngeal

tuberculosis. Considering the lesions in toto and on going through the literature and the fact that the skin lesions preceded the laryngeal lesion and the chronic indolent course we came to a diagnosis of lupus vulgaris with laryngeal involvement and since histopathology confirmed the diagnosis and ATT was started.

The patient was administered anti tubercular therapy consisting of Rifampicin(450mg),Isoniazid(300mg),Pyrazinamide(1500mg) and Ethambutal(800mg) thrice a week for two months followed by two drugs namely rifampicin and isoniazid for the next four months. This regimen was based on the revised national tuberculosis control pro gramme (RNTCP) guidelines of our country. The patient responded well and the lesions in the lip and larynx regressed showing improvement during the course and on completion of ATT the patient was near normal with no significant scarring or disfigurement.

Discussion

Different forms of cutaneous tuberculosis are lupusvulgaris,scrofuloderma, tuberculosis verrucosa cutis,lichen scrofulosorum, erythema induratum, papulonecrotic tuberculid. Amongst all these commonest is that of lupus vulgaris constituting 59% of total skin tuberculosis [1].This is a chronic, progressive and tissue destructive form of cutaneous tuberculosis seen in patients with moderate or high degree of immunity., occurring mostly in tuberculin sensitive patients.

Lesions appear in normal skin as a result of direct extension of underlying tuberclous foci, of lymphatic or hematogenous spread,or by primary inoculation, BCG vaccination

or in scar of old scrofuloderma[1]. The study done by Singh Gurmohan showed significantly high frequency of this disease in the females [2]. It is the most common form of cutaneous tuberculosis in Europe but is less common in United states. In India it accounts for approximately 59% of cases [3]. Lupus is twice as common in females as in males and is developed most often in early adult life. The mucocutaneous junction of the nasal septum is the most common site of inoculation, as this is frequently exposed to trauma in patients who have the habit of picking the nose [4].The lesion in majority of the cases,involves the skin of the nares and the skin and mucous membrane in front of the nose around upper lip and nasolabial fold [1] . The diagnostic feature is the presence of apple jelly nodules, pin head sized red spots which do not blanch when compressed, for instance with a glass slide [1]. This feature is not made out in many pigmented patients. The course is very slow, the cartilage within the affected area is progressively destroyed. Bone is usually spared .In more advanced cases, there may be more extensive involvement of the floor of nose and turbinates, spreading backwards from the primary site. The surface shows superficial ulcers and crusts. The septum may perforate but only in cartilaginous portion [4]. It may spread back into nasopharynx, the palate and the larynx [1] like what has happened in our case.

Clinical variants are numerous and are seen in the following forms:

Plaque forms: Disease extension occurs with little central atrophy. Scaling can occur,especially on the lower legs where it may resemble psoriasis. Irregular scarring is common and the active edge may be thickened and hyperkeratotic.

Ulcerating form: Scarring and ulceration predominate. Crusts form over areas of

necrosis. Deep tissues and cartilage are invaded by eventual scarring that produces contractures and deformity.

Vegetative form: This form is characterized by necrosis, ulceration and proliferation and papillomatous granulation tissue.

Nodular form: This form is characterized by a relative absence of ulceration and scarring. Large soft tumours occur,especially on ear lobes.

The lesions characteristically progresses by peripheral extension and central healing, atrophy and scarring. Barrie (1975) describes two types of lupus involving the nose(a) a slowly progressive usually non ulcerative tuberculous infection of the skin.,the disease is probably borne by the finger to the nose and the bacillus enters the deep layers of the skin from a finger scratch. The condition is characterized by miliary tubercles forming lupus nodules in the dermis. (b)An ulcerative type of infection of the skin,which may spread rapidly,and which is nearly always secondarily infected by staphylococci .

Lupus of pharynx and larynx occurs in 10-20% of the patients with lupus of the Skin [5]. Lupus of the pharynx is almost invariably secondary to lupus of nose and Face [6]. Laryngeal lupus is the result of spread from the nose. The free part of the epiglottis is the initial site of laryngeal disease but from here it may spread along the ariepiglotic folds as far back as the arytenoids. It has been known to encroach onto the false cords. Ulceration results in more or less destruction, chiefly of the epiglottis. It is painless, indeed often entirely symptomless, but examination of larynx may be prompted by the nasal condition. Lupus is not likely to be confused with laryngeal tuberculosis principally because of its characteristic distribution, the absence of

pulmonary disease and the presence of active or burnt out lupus of the nose [1]. Bhandary and Usha Ranganna have reported a case of Lupus Vulgaris of external nose and have stressed on the importance of taking a deeper biopsy for not missing the characteristic features of lupus vulgaris [8].

Conclusion

Lupus vulgaris is a common morphologic form of cutaneous tuberculosis, but this type of spread to larynx with typical features of laryngeal lupus is rare. A high index of suspicion and a thorough clinical examination of the pharynx and larynx is mandatory for not missing this diagnosis of laryngeal lupus. Diagnosis of lupus from the skin lesion necessitates an adequate and deeper biopsy, as the superficial tissue may show only the non specific inflammatory cell infiltrate missing the characteristic features of lupus vulgaris. The disease was completely cured without any deformity.

References

1. Salmon.L.F.W,chronic laryngitis.In."Scott-Browns Diseases of the Ear,Nose and Throat"The Pharynx and Larynx."Vol. 4Ballantine J,Groves J, editors. 4th Edition,London : Butterworths; 1979,pp 406-409.
2. Singh G(1974) Lupus Vulgaris in India, Indian Journal of Dermatology Venereology and Leprosy 40(6):257-260.
3. Ramesh V,Misra RS,Jain RK (1987) Secondary Tuberculosis of Skin;Clinical features and problems in laboratory Diagnosis, Int Journal of Dermatology 26:578-581
4. Neil Weir , Acute and chronic inflammations of the nasal cavities .In "Scott-Browns Diseases of the Ear,Nose and Throat" Vol. 3 Ballantine J,Groves J,editors. 4th Edition,London Butterworths,1979,pp186-187.

5. Ballenger WL,Ballenger HC In : "Diseases of the Nose,Throat and Ear Medical and Surgical "8th Edition,Philadelphia:Lea and Febiger'1943,P447
6. Mckanzie D.In:"Diseases of Throat,Nose and Ear"1st Edition London:William Heinemann Ltd,1920,pp75-77
7. Bhandary SK,Usha Ranganna B,Lupus Vulgaris of External nose Indian Journal of Otolaryngology,Head and Neck Surgery(oct-dec 2008)Vol 60.No.4 pp373-375

SUMMARY

- Lupus vulgaris ,a cutaneous form of Tuberculosis may sometimes spread to nose,pharynx and larynx.
- A23 yr old female patient with typical features of laryngeal lupus following the skin lesions is presented here.
- A thorough clinical examination of nose,pharynx and larynx., a collective analysis of the symptomatology and clinical findings and a deeper tissue biopsy have helped in diagnosis.

Figures.

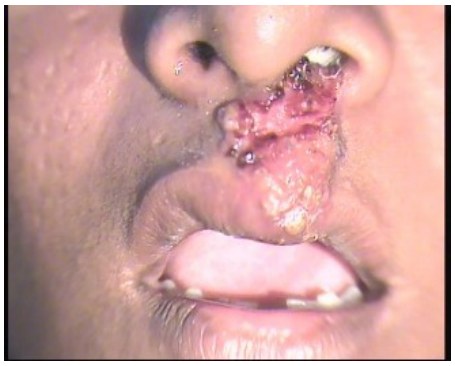

Fig.1

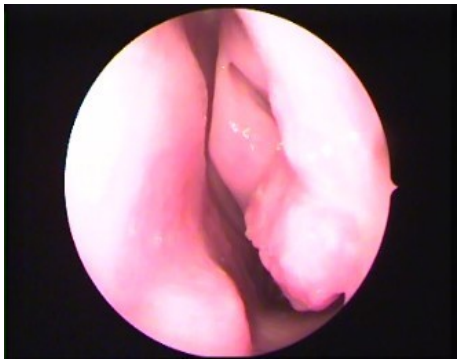

Fig 2.

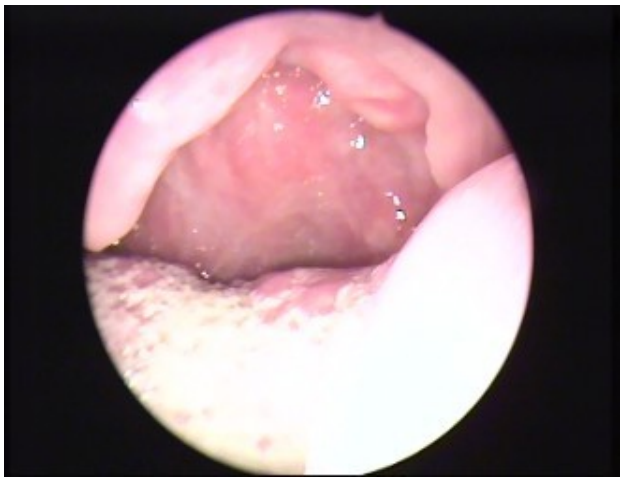

Fig 3.

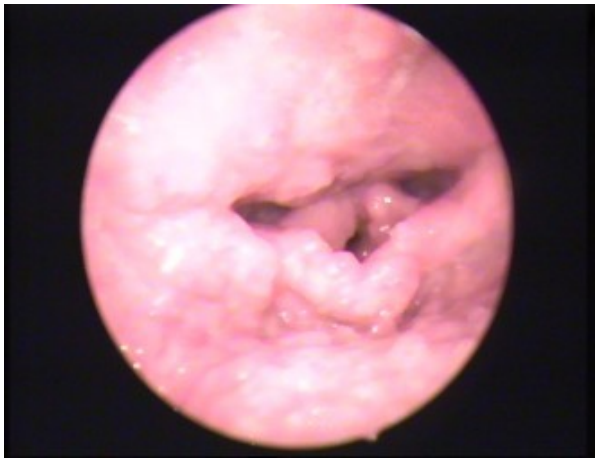

Fig 4.

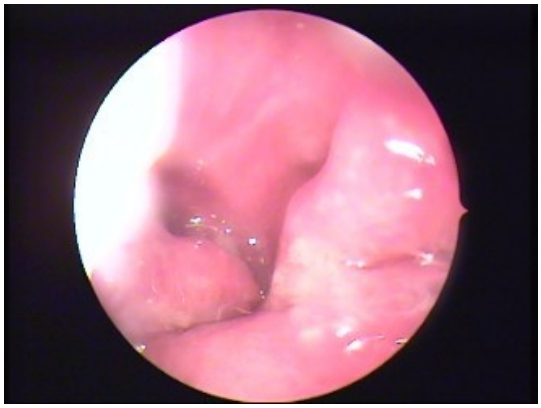

Fig 5.

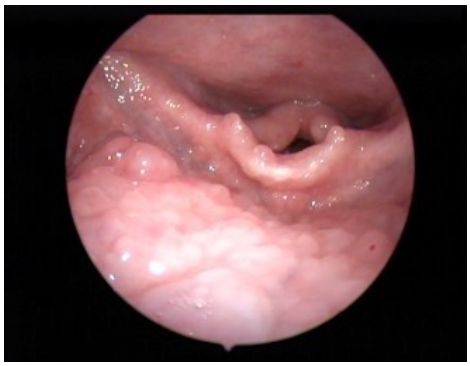

Fig 6.

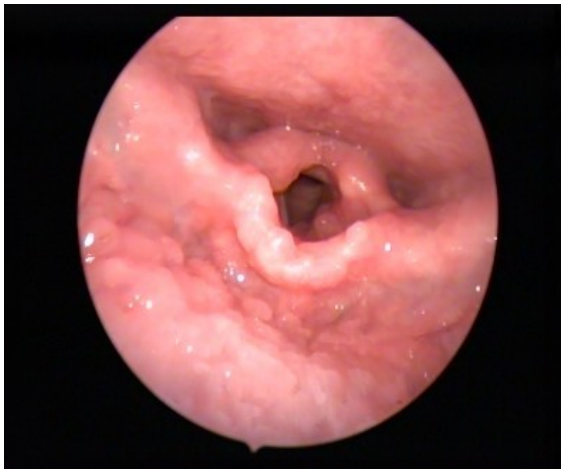

Fig 7

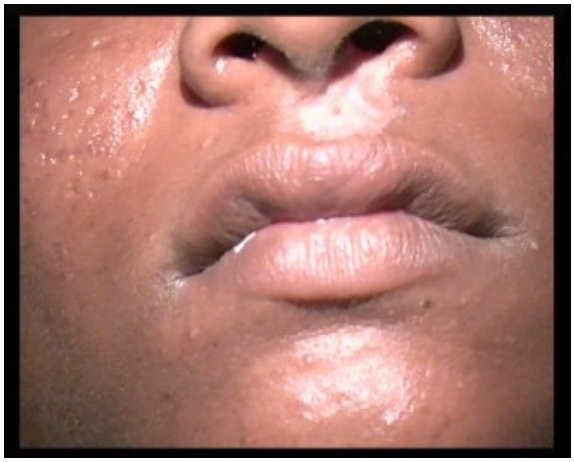

Fig 8.

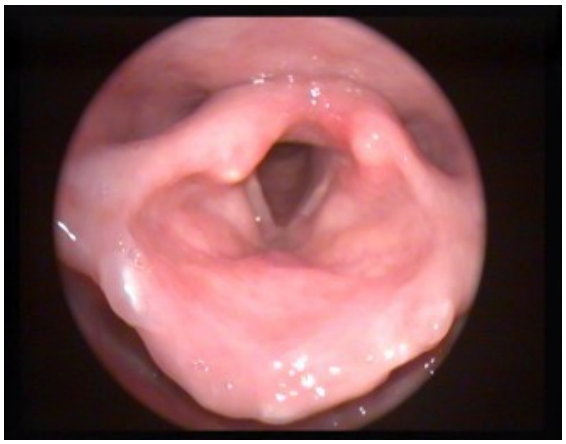

Fig 9.

Figure legend.

Fig.1-Lupus vulgaris in lip extending to nose.

Fig.2 –Lesion in Septum and Inferior turbinate.

Fig.3 –Lesion in Soft palate.

Fig.4 – Lesion in Larynx.

Fig.5 – Lesion in Nasopharynx.

Fig.6 & 7 – During the course of treatment . .

Fig.8 & 9 – After treatment,lesion completely resolved.

Consent For Publication of My case Record

I,Sangeetha do hereby give my express consent to Prof.Sankaranarayanan and his team for publication of my case record with the understanding that this might be useful in diagnosing and treating similar conditions.I also give release to him to publish my photographs as part of the case record.

Sangeetha

(Sangeetha)