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CASUISTIC PAPER

Favre-Racouchot syndrome and chronic obstructive pulmonary disease – a common link

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ABSTRACT

Introduction and aim. Favre-Racouchot syndrome though mostly reported in Caucasian men (with an estimated prevalence of 6% in adults older than 50 years), cases have been reported in dark-skinned population including Indians, albeit rarely. It is characterized by large open and closed comedones along with epidermal cysts over the nose, cheeks, temples, forehead and periorbital areas. The association of this condition with chronic heavy smoking is what it makes compelling.

Description of the case report. We report a case of elderly male, chronic heavy smoker who was diagnosed as a case of chronic obstructive pulmonary disease (COPD) as per standard guidelines. He presented with multiple nodulo-cystic lesions and had undergone a skin biopsy. Histomorphology features were consistent with Favre-Racouchot syndrome.

Conclusion. Early identification of this skin condition in mildly symptomatic and asymptomatic smokers may help clinicians to forewarn the patients regarding development of chronic obstructive pulmonary disease (COPD).

Keywords. comedones, COPD, Favre-Racouchot syndrome

Introduction

Favre-Racouchot syndrome though mostly reported in Caucasian men (with an estimated prevalence of 6% in adults older than 50 years), cases have been reported in dark-skinned population including Indians, albeit rarely. It is characterized by large open and closed comedones along with epidermal cysts over the nose, cheeks, temples, forehead and periorbital areas.[1-3]

Aim

To describe the link between Favre-Racouchot syndrome and chronic obstructive pulmonary disease (COPD) in an Indian patient.

Description of the case report

We report a case of 58 years old elderly male, resident of Bhomkhera village in Rajasthan, tailor by occupation with minimal field work and sun exposure, who was admitted to our hospital with complaints of gradually progressive breathlessness correlating to modified Medical Research Council (mMRC) grade 1 to grade 3 dyspnoea along with cough and minimal expectoration since past six months. He has been a chronic heavy smoker smoking more than 20 cigarettes a day for past three decades. The patient denied any history of headaches, dizziness, syncope, paroxysmal nocturnal dyspnoea or pedal oedema. His personal and family history was unremarkable. There was no history of pulmonary or extrapulmonary tuberculosis in the past.

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On initial evaluation, patient had pulse oximetry saturation of 90% at room air with heart rate of 110/minute and blood pressure of 110/70 mmHg. His general physical examination was remarkably normal except for some hyper-pigmented multiple nodular cystic lesions over dorsum of his nose, nasolabial folds and forehead. Respiratory system examination revealed decreased breath sounds over both lung bases. Cardiovascular system examination was normal.

His laboratory parameters revealed haemoglobin of 14.5gm/dl, total leukocyte count (TLC) of 10,800/mm^3, with normal platelet counts. Renal and liver function tests were within normal limit. Sputum culture was sterile and sputum for Ziehl-Neelsen stain was negative. Electrocardiogram (ECG) showed normal sinus rhythm with tachycardia. Arterial blood gases revealed mild hypoxemia. Chest X Ray (PA view) (Figure 1) showed bilateral hyperinflated lung fields with prominent broncho-vascular markings suggestive of chronic bronchitis. Pulmonary function testing revealed a moderate obstructive ventilatory defect with forced expiratory volume in one second (FEV_1) of 0.94 l (63% of predicted). Thus, based on relevant clinical history, examination and investigations including spirometry, this patient was diagnosed as a case of chronic obstructive pulmonary disease (COPD) as per standard guidelines.

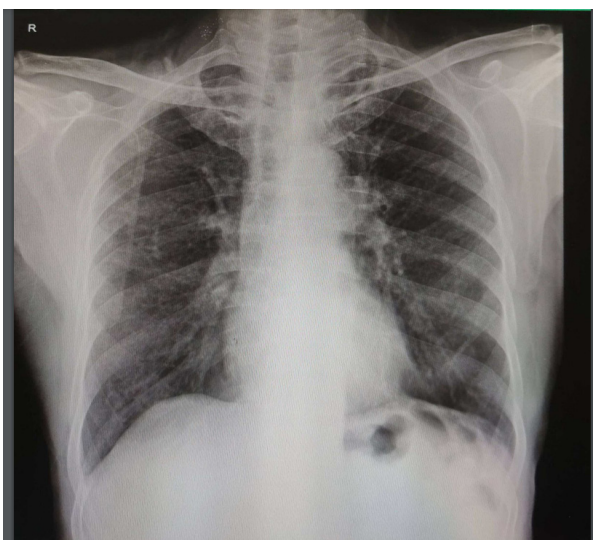

Fig. 1. Chest X-Ray Postero-Anterior view shows bilateral hyperinflated lung fields with prominent broncho-vascular markings

The patient was emotionally disturbed with the unappealing appearance of his face and gave a history of slowly expanding dark coloured lesions on his nose. Dermatologist's opinion was sought and on examination, multiple nodule cystic lesions (largest measuring 1x2 cm) were described over dorsum of his nose and forehead (Figure 2). There was no history of any drug intake, exposure to chemicals or toxins, application of

Fig. 2. Multiple nodulo-cystic lesions bilaterally over dorsum of nose, nasolabial folds and forehead

cosmetic creams to face or any other associated systemic illness.

He was advised to undergo skin biopsy. Incisional skin biopsy was done from forehead lesion under local anaesthesia. Histopathology revealed keratinized stratified squamous epithelium having dilated pilosebaceous openings and cyst like spaces lined by flattened epithelium, filled with lamellated keratinous material. Histomorphology features were consistent with Favre-Racouchot syndrome (Figure 3).

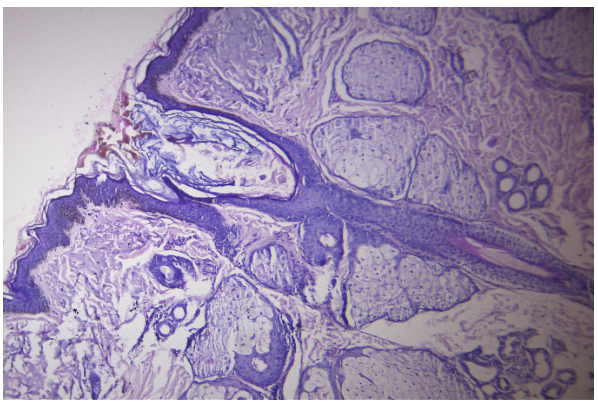

Fig. 3. Histopathology reveals keratinized stratified squamous epithelium having dilated pilosebaceous openings and cyst like spaces lined by flattened epithelium, filled with lamellated keratinous material (H+E stain; 40 X)

Patient was managed with antibiotics along with inhaled corticosteroids and bronchodilators and was symptomatically better. Patient was simultaneously counselled regarding smoking cessation and sun protection for prevention of recurrence of skin lesions.

Discussion

The ancient "Zang-Fu" theory proposed that skin is functionally linked to lung. Vierkotter et al (2010) showed that cigarette smoking and ambient soot levels were important factors in causing pigment spots and wrinkles.[1] Cutaneous manifestations are frequently associated with pulmonary diseases.[2] pulmonary diseases can be life threatening and early detection and treatment may have impact on patient's quality of life. Better knowledge and understanding of both common and rare cutaneous manifestations of pulmonary diseases enables physician to enhance their clinical competency.

The common dermatological manifestations noted in COPD patients are xerosis, senile purpura, onychorrhasis, onychomycosis, tobacco nail stains and hyperkeratosis of both elbows. It has been seen that a non-invasive determinant of skin elasticity is independently associated with pulmonary emphysema and tissue proteolysis in tobacco exposed individuals. Loss of skin elasticity is a novel observation that link common pathology in skin and lung in emphysema.[3] More recent reports suggest that an association between facial wrinkling and airflow obstruction exists, which may explain common susceptibility to deleterious effects of smoking.[4]

Nicotine containing cigarette smoke induces alteration in sebum composition as it causes increased oxidation stress, lipid peroxidation and reduced levels of alpha-tocopherol.[1] There is high grade of lipid peroxidation in sebum of smokers which leads to comedones and acne. Squalene, which is particularly important per-oxidated lipid in human sebum has hyperproliferative effect on keratinocytes and is therefore "comedogenic".[5,6]

Favre-Racouchot syndrome is a very peculiar type of dermatosis which is characterised by multiple large open and closed comedones on actinically damaged skin.[7] Though mostly reported in Caucasian men (prevalence 6%), cases have been rarely reported in dark skinned population including South Asians.

The exact mechanism by which smoking contributes to this syndrome is poorly understood but some proposed theories include:

- ultraviolet activated phototoxic properties of tobacco smoke,
- increased reactive oxygen species,
- increased matrix metalloproteinases.

Conclusion

To conclude, the importance of this skin manifestation in a chronic smoker lies in the fact that early identification of this condition and confirming it with histopathology, we might alert these patients particularly who are asymptomatic or mildly symptomatic regarding the development of chronic obstructive airway disease because of similar proposed pathogenesis. Positive association between smoking and this condition definitely exists and cessation of smoking along with sun protective measures are required for prevention of recurrence of lesions.

Declarations

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Author contributions

Conceptualization, M.I.S. and H.I.S.; Investigation, A.C.; Resources, Writing – Original Draft Preparation, M.I.S; Writing – Review & Editing, S.D.; Visualization, A.M., Ş.K.E., Ş.Ü.A.; Supervision, S.D.

Conflicts of interest

The authors have no conflict of interest.

Data availability

The data have not been made public, but are kept with the authors, if necessary.

Ethics approval

All subjects gave informed consent to the inclusion prior to participating in the study.

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