Review Article

Dermatological signs of chest diseases

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A wide spectrum of pulmonary diseases displays cutaneous manifestations, which may be the presenting signs and symptoms of the underlying respiratory diseases. This review focuses on the clinical features of such diseases affecting both skin and lungs with appropriate management. A heightened awareness of these conditions may facilitate early diagnosis and a better approach to treatment of such diseases.

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Key words **: Scrofula, Scrofuloderma, erythema nodosum, wegener's granulomatosis.**

A variety of pulmonary disorders such as noninfectious inflammatory diseases (sarcoidosis), heredi-tary diseases (Birt-Hogg-Dube syndrome), and malignancies (lung cancer)—may affect both the skin and the lungs. The recognition and appropriate elucidation of these cutaneous findings helps in limiting differential diagnosis and prognosis estimation. This article aims at recapitulating the dis-tinguishing cutaneous manifestation of pulmonary diseases which may help in early diagnosis and treatment.

Cyanosis :

The bluish or purple tinge of skin and/or mucous membrane[1] becoming evident on the presence of at least 5 g/dL of deoxyhemoglobin is known as cyanosis. According to the cause of haemoglobin desaturation, cyanosis may be central or peripheral.

Central cyanosis is due to circulatory or ventilatory complications leading to decreased oxygenation of blood in the pulmonary circulation as seen in congenital heart disease with right-to-left shunts and is best elicited on tongue, oral mucosa and conjunctiva.

Peripheral cyanosis occurs as a result of increased local oxygen extraction and decreased perfusion of tissues leading to high levels of desaturated hemoglobin, mostly seen in peripheral vascular diseases.

Clubbing :

Clubbing refers to the focal bulbous uniform enlargement of the soft tissue of the terminal phalanx of a digit with obliteration of the normal angle between the nail and the nail bed. Clubbing may be idiopathic, hereditary or more commonly acquired. It is associated with a variety of lung diseases[2] such as lung cancer, tuberculosis, lung abscess, bronchiectasis, cystic fibrosis, idiopathic pulmonary fibrosis[3]. The underlying pathogenesis is not clearly understood but platelet derived cytokines is hypothesised to play a key role. Lodging of the megakaryocytic in the digital microvasculature leads to release of platelet derived cytokines causing increased vascular permeability and connective tissue proliferation. Clubbing can be easily clinically identified by measurement of Lovibond angle, obliteration of diamond shaped window on opposing the corresponding fingers (Schamroth sign) and phalangeal depth ratio (ratio of distal phalangeal to interphalangeal depth >1)(Figs 1&2).

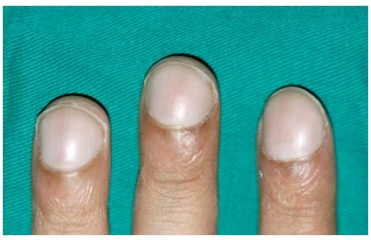

Fig 1 — A 54 years old male patient of bronchiectasis showing marked clubbing in index, middle and ring finger of right hand

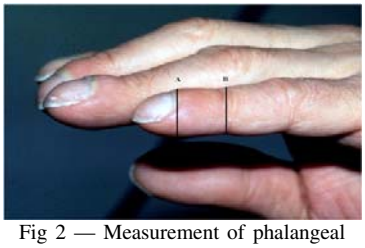

Fig 2 — Measurement of phalangeal depth ratio. Ratio of Distal phalangeal depth (A) and inter pha-langeal depth (B) is more than 1 is seen in clubbing

Hypertrophic Osteoarthropathy :

The triad of clubbing with periostitis of long bones and arthritis is known as hypertrophic osteoarthropathy (HOA). It may be primary (hereditary or idiopathic) or secondary.

Primary HOA :

Primary HOA or pachydermoperiostosis, characterised by thickening of facial skin or pachydermia, cutis verticis gyrata, and severe seborrhea is association with periostitis

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and digital clubbing[4]. Joint pain is usually absent in this condition.

Secondary HOA :

Secondary HOA is mostly associated with pulmonary malignancies[5], non-small cell lung cancer being the most frequent. It may be seen rarely in other conditions such as lung abscess, mesothelioma, primary biliary cirrhosis, ulcerative colitis, and Hodgkin disease.

Treatment :

HOA responds rapidly to treatment of the primary tumour, especially on curative resection. Aspirin or nonsteroidal anti-inflammatory drugs help in alleviation of arthritic or periostitic pain[6].

Scrofuloderma :

Cervical tuberculous lymph adenitis is known as scrofula (Fig 3). Cutaneous extension of tuberculous focus of lymph node is called scrofuloderma which is characterised by cutaneous red brown lesions. Firm painless, discrete lymph nodes later become matted. Lymph nodes then enlarge andeventually become fluctuant which rupture draining caseous material and form ulcers and sinuses. Diagnosis is confirmed by FNAC (Find Needle Aspiration Cytology). Treatment is by standard anti-tuberculous drugs.

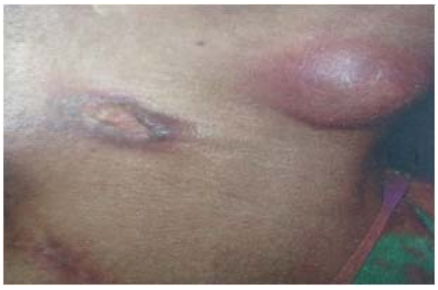

Fig 3 — A fluctuant, necrotic lymph node ready to rupture with scarred lesions

Lung Cancer :

Dermatological manifestation of lung cancer, although rare, occurs in form of cutaneous, subcuta-neous, or superficial lymph node metastases (Fig 4). The nodules are usually single, painless, firm and fast growing usually seen on chest, abdominal wall, neck, and scalp. Superficial lymph nodes mostly supraclavicular or anterior cervical lymph node groups, become firm and may be adherent to the adjacent skin. Adenocarcinomas seem to have the highest predisposition to metastasise to the skin[7-9].

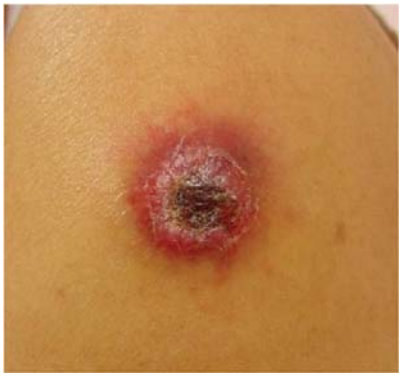

Fig 4 — A hard single nodular lesion seen on shoulder of a 59 years old male patient of small cell lung carcinoma

Superior Vena Cava Syndrome :

Superior vena cava syndrome is a life threatening condition caused due to obstruction of blood flow through the superior vena cava. It is mostly caused due to malignancies of lung. Other causes being syphilitic aneurysms or tuberculous mediastinitis.

Obstruction of superior vena cava results in increased collateral flow through the subcutaneous vessels which is seen as parallel clusters of markedly dilated venules on the chest wall (Fig 5). These disappear with the relief of obstruction. Other signs due to venous congestion are facial edema, con-junctival congestion and proptosis[10].

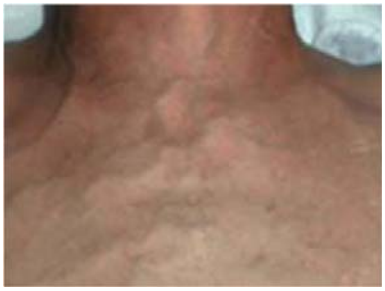

Fig 5 — Markedly dilated collaterals seen on the chest wall of a 45 years old male patient of lung cancer

Symptomatic treatment can be obtained by elevation of head end of the bed with oxygen supple-mentation. Intraluminal stenting provides rapid relief. Definitive treatment is resection of tumour or other underlying causes.

Sarcoidosis

Sarcoidosis is chronic multisystem inflammatory disease characterized by non caseating epitheliod granuloma formation in affected tissues most commonly lungs. Cutaneous involvement is seen in 25-30% of the patients with sarcoidosis which may be specific, revealing granuloma on biopsy or non-specific which is mainly reactive such as erythema nodosum[11] (Fig 6).

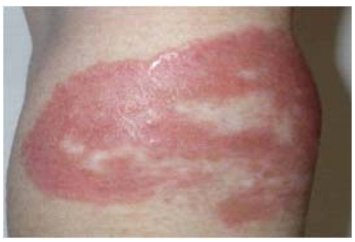

Fig 6 — Annular plaque lesions seen on back of left leg in a female patient of sarcoidosis. Biopsy showed non caseating granuloma

Specific lesions of sarcoidosis on skin manifest as Lupus pernio, maculopapular, nodular, scar, plaque, angiolupoid, ichthyosiform, lichenoid, annular, verrucous, psoriasiform, subcutaneous no-dules and ulcerations. These lesions tend to involve the entire dermis with mild discolouration of the overlying epidermis[11].

Erythema nodosum is most common form of nonspecific cutaneous finding in sarcoidosis. It presents as tender erythematous eruptions over the anterior aspect of lower extremities seen more frequently in young females. Erythema nodosum is associated with good prognosis[11].

Corticosteroids are the mainstay of treatment in patients with pulmonary involvement.

Wegeners Granulomatosis (Granulomatosis with Polyangitis) :

Wegener granulomatosis, is a rare idiopathic multisystem autoimmune disease consisting of triad of necrotizing granulomatous inflammation of upper and lower respiratory tract with renal involvement. Cutaneous manifestations are seen in 10% of cases[12-14]. Lesions include palpable purpura and necrotising ulcers seen on lower extremities. Nonspecific oropharyngeal ulcers are also seen. Gingival hyperplasia with pain and bleeding gums is a pathognomic feature. Skin lesions correlate with renal involvement therefore helps in prognosis. Treatment with corticosteroids and cyclophosphamide shows dramatic improvement[12-14].

Churg Strauss Syndrome (Eosinophilicgranulomatosis with Polyangitis) :

Churg Strauss syndrome is a chronic disease affecting multiple systems affecting lungs, peripheral nervous system and skin. It is characterized by early onset allergic rhinitis,late onset asthma and mono neuritis multiplex affecting peroneal nerve leading to foot drop. Skin lesions are seen in 70% of patients presenting as palpable purpura, subcutaneous nodules (may or may not be tender), urticarial rashes and livido reticularis[15]. Biopsy shows eosinophilic granuloma with a necrotic centre. High grade peripheral eosinophilia with counts more than 1.5 X 10^9 cells/L is striking lab finding. It usually responds well to high dose corticosteroids.

Relapsing Polychondritis :

Recurrent, progressive inflammation of cartilaginous structures of the body is referred to as relapsing polychondritis[16,17]. Ear cartilage is most frequently affected. Erythema nodosum, erythema multiform and saddle nose deformity may also be seen. Immunosuppressive therapies are found to be responsive.

Fat Embolism Syndrome :

Fat embolism syndrome is a rare complication following trauma and fractures of the long bones or pelvis. The fat emboli reaching the microcirculation induce an intense systemic inflammatory cascade leading to respiratory distress, altered mental sensorium, and thrombocytopenia, occurring within 24-72 hours of trauma. The characteristic cutaneous finding are transient petechial rashes on nondependent portions of the body (ie, upper chest, neck, axillae, conjunctivae)[18]. Mortality rate is high and supportive therapy with high flow rate oxygen remains the mainstay of treatment.

Yellow Nail Syndrome :

Yellow nail syndrome consists of the triad of yellow malformed toenails and fingernails, lymphe-dema, and pleural effusions[19] (Fig 7). Thick, yellow, dystrophic nails with transverse ridging and a growth rate of less than 0.5mm/week are hallmark features. Lymphedema usually involves lower extremities. Pleural effusion develops at last and is recurrent. Treatment is supportive and symptomatic as the etiology is unknown.

Scleroderma :

Cutaneous fibrosis of skin (sclerosis) and Raynaud's phenomenon are the characteristics features of scleroderma. It is associated with pulmonary fibrosis and pulmonary hypertension. Cyclophospha-mide and penicillamine are drugs used for treatment of scleroderma.

Dermatomyositis :

Gottron's papules, heliotrope and poikiloderma are cutaneous manifestation of dermatomyositis which is accompanied with pulmonary fibrosis. Treatment is corticosteroids and immunosuppressive drugs.

Birt-Hogg Dube Syndrome :

It is a rare autosomal dominant condition caused by mutations in the gene coding for protein folliculin responsible for limiting cell growth and division. The disease is characterized by pneumothorax and lung cysts, multiple benign skin tumours, and kidney tumours[20].

Most frequent dermatological features are fibrofolliculomas (pale tumours in hair follicles), tricho-

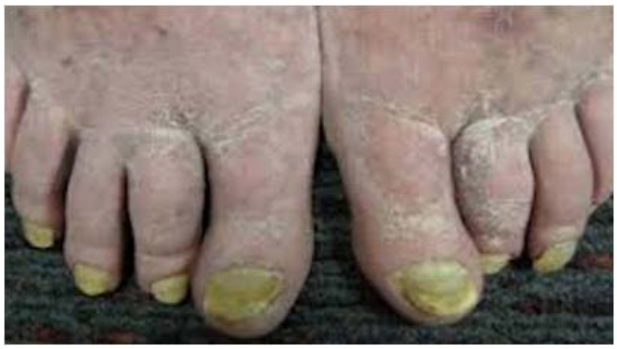

Fig 7 — A 48 years old male patient presenting with lymphedema and yellow, thickened, dystrophic toe nails

discomas (skin coloured tumours of hair follicles), and acrochordons (skin tags) seen on the face, neck, and upper trunk. Early diagnosis includes screening for germline mutation of folliculin gene in family members.

Conclusion :

The skin frequently serves as a marker for underlying internal chest diseases. A greater awareness of these markers can lead to appropriate diagnosis and comprehensive management and ultimately contribute to enhanced patient care.

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