Cutaneous manifestations of lung disease

Skin disease may be associated with pulmonary disease in a variety of ways. This article describes cutaneous manifestations of selected pulmonary conditions and other associations between the skin and lung.

Pulmonary disease can be associated with skin disease in a variety of ways, which can be subdivided as follows:

- Pulmonary disease as a direct consequence of a primary skin disease
- Skin abnormalities that occur as a direct consequence of respiratory pathology
- Skin and respiratory tract manifestations of the same disease process, i.e. as part of a multisystem disorder.

Pulmonary disease as a consequence of primary skin disease is rare and will not be discussed in this article. An example is lung metastases from a cutaneous malignant melanoma.

Skin abnormalities as a consequence of respiratory pathology are more often encountered in a general medical setting. These skin signs of internal disease include clubbing, cyanosis and superior vena cava (SVC) obstruction.

The most common scenarios in which skin and lung disease are associated are the multisystem diseases – in these cases, the skin and respiratory tract exhibit the same disease process. Examples of such multisystem diseases include sarcoidosis, tuberculosis (TB), the vasculitides and the connective tissue diseases.

This article concentrates on a selection of cutaneous manifestations associated with pulmonary conditions which the general internal physician may encounter in clinical practice. The various cutaneous manifestations associated with pulmonary disease are outlined in *Table 1*.

Skin abnormalities secondary to respiratory pathology

Clubbing

Nail clubbing describes the bulbous swelling of soft tissue of the distal portion of a digit with subsequent loss of the normal angle between the nail and the proximal nail fold (Lovibond angle) (*Figure 1*). Clubbing may be bilateral and symmetrical or unilateral or even involve a single digit. Bilateral clubbing produces a positive 'Schamroth sign', which is the obliteration of the normally diamond-shaped space formed when dorsal sides of the distal phalanges of corresponding right and left digits are opposed.

Clubbing may be a marker of underlying systemic disease (secondary clubbing) or it may occur without evident underlying disease (primary or idiopathic clubbing, or familial clubbing). Systemic causes of finger clubbing are listed in *Table 2*. Clubbing has been reported in 29% of patients with lung cancer and is seen more commonly in non-small cell lung carcinoma (35%) than in small cell lung carcinoma (4%) (Sridhar et al, 1998). In idiopathic pulmonary fibrosis, approximately 65% of patients have finger clubbing (Kanematsu et al, 1994).

Cyanosis

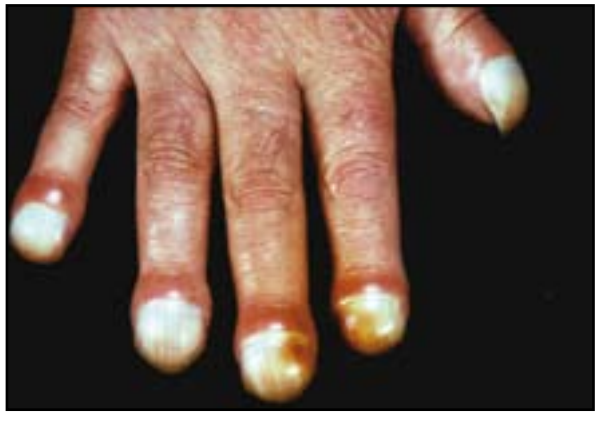
Cyanosis is a distinctive dusky bluish or purplish discoloration of the skin and mucous membranes.

Central cyanosis involves the entire skin surface but may be best appreciated in the tongue and oral mucosa – it signifies hypoxaemia, and reveals the presence of elevated amounts (>5 g/dl) of desaturated (unoxygenated) haemoglobin. Severely anaemic patients are therefore unable to manifest central cyanosis. It is a manifestation of congenital heart disease with right-to-left pulmonary shunts, pulmonary hypertension, as well as most severe pulmonary diseases (e.g. chronic obstructive pulmonary disease, severe pneumonia).

Peripheral cyanosis spares the mucosal surfaces and may be best seen on the peripheries, such as the fingertips. It is a manifestation of diminished tissue perfusion, occurring, for example, in shock, cardiac failure or peripheral vascular disease.

Clinical distinction between the two types of cyanosis is notoriously difficult. Further, rarer conditions, such as

Figure 1. This man with pulmonary fibrosis has finger clubbing, with prominent swelling of the distal portions of all digits. Loss of the Lovibond angle is seen on his thumb.



Dr Ai-Lean Chew *is Specialist Registrar in Dermatology and*
Dr Daniel Creamer *is Consultant Dermatologist, Department of Dermatology, Kings College Hospital, London SE5 9RS*

Correspondence to: Dr D Creamer

methaemoglobinaemia and sulfhaemoglobinaemia, as well as staining from topical or systemic ingestion of heavy metals, may also impart a bluish tinge to the skin, often misdiagnosed as cyanosis.

Superior vena cava obstruction

SVC obstruction (SVC syndrome) is the interruption of blood flow through the SVC – the main drain for venous blood from the head, neck, upper thorax and arms.

Table 1. Conditions that may affect the skin and respiratory system

	Disease	Cutaneous features	Respiratory features
Skin abnormalities secondary to respiratory pathology	Clubbing	Clubbing	Bronchial carcinoma Chronic obstructive pulmonary disease Suppurative chronic lung disease
	Cyanosis	Cyanosis	Hypoxaemia (e.g. congenital heart disease with right-to-left shunts, pulmonary hypertension)
	SVC obstruction	Facial oedema, plethora, cyanosis Cheimosis	Dyspnoea, orthopnoea Coughing, stridor
Skin and respiratory tract manifestations of multisystemic disease	Sarcoidosis	Specific skin lesions (papules, plaques, nodules) Reactive skin lesions: erythema nodosum	Bilateral hilar lymphadenopathy* Pulmonary fibrosis Pulmonary infiltrates*
	Tuberculosis	Specific skin lesions: lupus vulgaris, scrofuloderma Reactive skin lesions: erythema nodosum	Cavitating lung infiltrates* Hilar lymphadenopathy*
	Atopic disease	Eczema	Asthma, hayfever
	Systemic lupus erythematosus	Discoid lupus Butterfly rash Mouth ulcers Photosensitivity Livedo reticularis Palpable purpura	Pleuritis Pleural effusions Pulmonary fibrosis Interstitial lung disease Pulmonary hypertension
	Systemic sclerosis	Scleroderma Raynaud's phenomenon Nail fold inflammation	Pulmonary fibrosis Pneumothorax Pulmonary hypertension
	Dermatomyositis	Gottron's papules Heliotrope rash Periorbital oedema Nail fold inflammation	Pulmonary fibrosis Muscular weakness Interstitial lung disease
	Ehlers–Danlos syndrome	Skin fragility and hyperextensibility	Pneumothorax
	Wegener's granulomatosis	Palpable purpura Subcutaneous nodules Pyoderma gangrenosum-like ulcers Oral ulcers Gingival hyperplasia	Vasculitis Cavitating pulmonary nodules* Upper respiratory tract inflammation (destructive sinusitis, epistaxis, saddle nose deformity)
	Churg–Strauss syndrome	Palpable purpura Urticarial weals Subcutaneous nodules Livedo reticularis	Asthma Migratory pulmonary infiltrates*
	Behcet's disease	Oral and genital ulcers Papules, pustules, plaques Erythema nodosum-like lesions Thrombophlebitis	Pleurisy Perihilar opacities* Pulmonary arterial aneurysm
	Microscopic polyangiitis	Nodules Palpable purpura	Nasopharyngeal involvement Alveolar haemorrhage
	Yellow nail syndrome	Yellow, thickened nails	Pleural effusions Bronchiectasis Recurrent respiratory infections

* denotes chest X-ray findings

The mechanism by which SVC obstruction occurs may be by neoplastic invasion of the venous wall associated with intravascular thrombosis, or by extrinsic pressure of a tumour against the SVC. Lung cancer is seen in the majority of cases of SVC obstruction, while less common causes include lymphomas and thymus malignancies. Non-malignant causes of SVC obstruction include benign mediastinal tumours, mediastinal fibrosis, aortic aneurysm and infections (such as syphilis or tuberculosis).

Facial, periorbital, neck or arm oedema may be evident on examination, as may plethora or cyanosis. Cheimosis (conjunctival oedema) may also be seen. Jugular venous pressure will be elevated and non-pulsatile, and engorged veins may also be seen on the upper chest. Bending forward or lying down will exaggerate the clinical findings. As the condition progresses, the patient may develop respiratory symptoms of dyspnoea, orthopnoea, coughing or stridor. Headache and a feeling of facial or upper body fullness may be present.

Multisystemic diseases with skin and lung manifestations

Sarcoidosis

The hallmark of sarcoidosis is the presence of non-caseating granulomas in tissues; the organs most commonly involved are the lungs, lymph nodes, eyes and skin.

The respiratory system is affected in 90% of patients with sarcoidosis. Often, this may consist of the incidental finding of hilar lymphadenopathy on chest X-ray; the patient may not have any respiratory symptoms. If the disease progresses, the patient may develop symptoms of cough, dyspnoea on exertion and haemoptysis. Approximately 10% of patients will eventually develop pulmonary fibrosis.

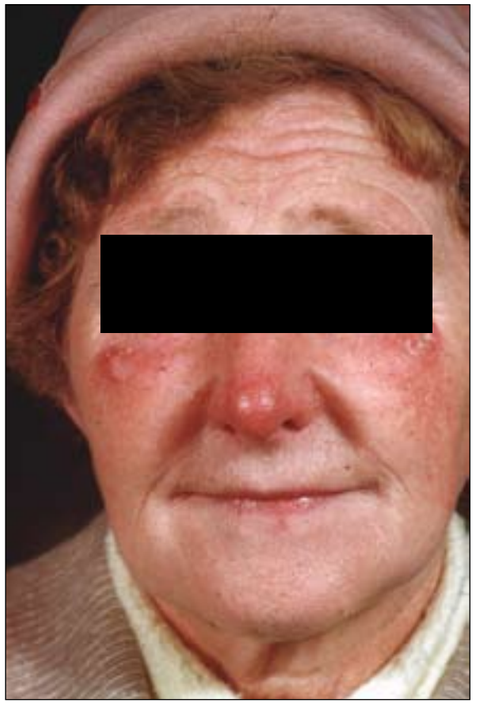
Skin involvement occurs in approximately 25% of patients with systemic sarcoidosis, but cutaneous sarcoidosis can also occur in isolation (Kerdel and Moschella 1984). The cutaneous manifestations of sarcoidosis are legion. Papules, plaques and cutaneous and subcutaneous nodules are common presentations. Papular sarcoidosis usually consists of small papules, typically seen on the head and neck, periorbitally and adjacent to the nasolabial folds. They may be flesh-coloured, reddish, purple, or brown in colour. Plaques in sarcoidosis are thick and may be of various colours. Lupus pernio refers to distinctive plaques of sarcoidosis classically seen on the nose (*Figure 2*). Less common presentations of sarcoidosis include annular, ulcerative, psoriasiform, hypopigmented, ichthyosiform and verrucous lesions, as well as erythroderma and scarring alopecia. Granulomas can also form on old scars ('scar sarcoidosis') or on tattoos.

If granulomas are present on skin biopsy, then the lesions are termed 'specific' lesions of sarcoidosis. Lesions that are associated with sarcoidosis but are not granulomatous are non-specific. The most common non-

Table 2. Systemic associations of finger clubbing

Organ system	Disease
Pulmonary	Bronchogenic carcinoma
	Mesothelioma
	Asbestosis
	Idiopathic pulmonary fibrosis
	Chronic bronchitis
	Empyema
	Lung abscess
	Bronchiectasis
	Cystic fibrosis
	Tuberculosis
Cardiovascular	Cyanotic congenital heart disease
	Infective endocarditis
	Atrioventricular malformations
	Atrial myxoma
Gastrointestinal	Inflammatory bowel disease
	Liver cirrhosis
	Malabsorption, e.g. coeliac disease
	Gastrointestinal lymphoma

Figure 2. This lady with sarcoidosis has lupus pernio, seen here as a smooth red indurated plaque on the tip of her nose.



specific cutaneous lesion seen in association with sarcoidosis is erythema nodosum, which manifests as tender, red subcutaneous nodules, most often found on the shins. It is usually an early feature and associated with a favourable prognosis. The association of erythema nodosum with bilateral hilar lymphadenopathy, fever, arthritis and uveitis is known as Lofgren's syndrome.

Tuberculosis

Mycobacterium tuberculosis, the causative organism for TB, is predominantly spread by airborne droplets. TB manifests in the lungs in 85% of cases. Extrapulmonary TB can affect a variety of organs, including the skin. Cutaneous TB can be caused by three main routes – direct inoculation, contiguous spread and haematogenous spread. Also, there are skin reactions which can occur in response to TB elsewhere in the body – 'tuberculid reactions'. Non-specific reaction patterns, such as erythema nodosum and erythema multiforme, may also be associated with TB.

Direct inoculation

A tuberculous chancre from direct inoculation usually follows an injury causing skin disruption, as mycobacteria poorly penetrate intact skin. A tuberculous chancre develops in those who have no prior exposure to *M. tuberculosis*: 2–4 weeks after exposure, an inflammatory papule develops at the inoculation site. This breaks down into a painless ulcer, usually in association with regional lymphadenopathy. The ulceroglandular complex is the result of a primary exposure and is therefore the equivalent of the pulmonary Ghon complex. Depending on the host immune response, the primary lesion may heal with scarring, or progress into more chronic forms of cutaneous tuberculosis or lead to disseminated spread.

Tuberculosis verrucosa cutis is the result of direct inoculation in patients who have had previous exposure to *M. tuberculosis* or *M. bovis*. Clinically, an indolent warty plaque develops, classically seen on the hands of people in high-risk groups such as mortuary technicians and pathologists. Other sites affected include the feet and buttocks. This slow-growing lesion can be mistaken for a viral wart initially, but gradually enlarges and persists for many years. Spontaneous resolution with scarring can occur.

Contiguous spread

Scrofuloderma results from breakdown of the skin overlying a tuberculous focus, usually at a lymph node, but occasionally over infected bones or joints. A firm, painless subcutaneous nodule develops that gradually enlarges and becomes suppurative, eventually breaking down to form an ulcer or a sinus. Scrofuloderma is more common in children.

Oroficial TB is a rare condition that describes TB of the mucous membranes and skin surrounding the orifices (mouth, anus, vulva) caused by autoinoculation into these areas, in patients with TB affecting the internal organs. Lesions usually consist of a painful red nodule which ulcerates.

Haematogenous spread

Lupus vulgaris is a chronic, progressive form of cutaneous TB in previously sensitized individuals. Lesions usually develop on the head and neck and are usually solitary. Typically, a soft brown nodule forms on the nose or cheek, then extends peripherally to form a larger plaque, which progresses relentlessly, destroying any underlying cartilage. Marked disfiguration may occur, and the mucous membranes of the mouth, nose and conjunctiva may be involved.

Miliary TB of the skin is a rare manifestation of miliary TB resulting from haematogenous spread to multiple organs, including the skin. The initial site of infection is usually pulmonary or meningeal. Miliary TB tends to occur in children or immunosuppressed patients. Disseminated small maculopapular lesions are seen, often with purpura, vesicles or central necrosis. These patients are usually very ill, and prognosis is poor.

Tuberculids

Tuberculids are a group of exanthems now thought to be the result of haematogenous spread of bacilli in patients with tuberculin immunity. The underlying focus of infection is not always apparent clinically and the affected tissue may be negative for mycobacteria on microscopy and culture. This group includes erythema induratum, lichen scrofulosorum and papulonecrotic tuberculid. Erythema induratum (Bazin's disease) manifests as tender, erythematous nodules, usually seen on the lower legs of young females.

Atopy

Asthma, eczema (atopic dermatitis; AD) and hayfever form the atopic triad. The association between the three conditions is well-known but the link still remains poorly understood. Infants with AD are at increased risk of developing asthma and hayfever in later life, but there seems to be a difference in prevalence of each component. Most patients develop only one or two components of the atopic diathesis.

The main symptom of AD is incessant itching. Primary skin lesions seen in AD include xerosis (dryness), and eczematous inflammation. In the acute phase, excoriations (scratch marks) may lead to open, exudative, lesions, which become secondarily infected. In chronic eczema, repeated rubbing and scratching leads to lichenification, in which the skin becomes thickened with prominent skin markings.

The distribution of the rash varies with the age of the patient. In infancy, the rash usually involves the face, scalp and extensor surfaces of the limbs. Around 2 years of age, children develop flexural involvement and the face becomes less frequently involved. The chest, back, abdomen and buttocks are also commonly involved in children. In adulthood, AD tends to occur in the same distribution as in late childhood, mainly in the neck and flexural parts of the limbs. Hands are also a common site for adult AD, often exacerbated by irritant factors, such as wet work.

Other clinical findings which are associated with AD include xerosis (dry skin), ichthyosis vulgaris, hyperlinearity of the palms and soles, and keratosis pilaris. Keratosis pilaris is seen as hyperkeratotic follicular papules found on upper arms, thighs or buttocks. Pigmentary changes, such as post-inflammatory hypo- or hyperpigmentation are sometimes seen after the active eczema has resolved. Pityriasis alba, characterized by hypopigmented scaly patches on the face and shoulders, is thought to be a manifestation of AD.

Connective tissue diseases

The connective tissue diseases are a group of multisystemic autoimmune disorders, which may have respiratory and cutaneous manifestations. This group includes systemic lupus erythematosus (SLE), systemic sclerosis (SSc), dermatomyositis, Sjögren's syndrome and mixed connective tissue disease.

Systemic lupus erythematosus

SLE is a multisystem autoimmune disease of unknown aetiology which can affect almost any organ system. Antinuclear antibodies (ANA) are seen in almost all patients with SLE, while anti-dsDNA antibodies are highly specific.

The most well-known respiratory manifestation of SLE is pleurisy, which may present as pleuritic chest pain or dyspnoea. Pleural effusions may be seen on chest radiograph. Less common respiratory manifestations of SLE include lupus pneumonitis, interstitial lung disease or pulmonary fibrosis, and pulmonary hypertension.

Mucocutaneous manifestations are common in SLE and in fact constitute four of the eleven diagnostic criteria set by the American College of Rheumatology (Tan et al, 1982), namely oral ulcers, malar (butterfly) rash, photosensitivity and discoid lupus. The butterfly rash of SLE consists of diffuse patches or plaques of erythema on the cheeks. Photosensitivity consists of a macular or maculopapular rash in sun-exposed sites, such as the face, 'V' of the neck or dorsal surfaces of the hands, precipitated by sun exposure. In subacute cutaneous lupus, patients have a non-scarring annular rash, which is photosensitive. Discoid lupus presents as round, disc-like lesions, with follicular plugging, usually seen on the face, neck, ears and scalp. When discoid lupus occurs on the scalp, it can result in scarring alopecia. Other skin manifestations seen in SLE (but not specific to SLE) include panniculitis (lupus profundus), bullous lesions, vasculitic purpura, urticaria, chilblains, Raynaud's phenomenon and livedo reticularis.

Systemic sclerosis

SSc is characterized by fibrosis of skin, subcutaneous tissue and internal organs, in association with abnormalities of the microvasculature and immune system. The hallmark of the disease is scleroderma, which is tight, indurated, thickened skin caused by excess collagen deposition. SSc can affect any organ in the body, including the lungs. Pulmonary manifestations of SSc include interstitial fibrosis, which affects at least 50% of patients and presents with symptoms of exertional dyspnoea and cough. Pneumothorax, pleural effusion, pulmonary hypertension, respiratory muscle involvement and 'splinting' of the chest by sclerotic skin may also occur. Oesophageal dysmotility in CREST syndrome (Calcinosis, Raynaud's, oesophageal dysmotility, Scleroderma, Telangectasiae) may cause aspiration pneumonia.

By definition, skin involvement is seen in 100% of patients with scleroderma. Three phases of skin involvement may be distinguished:

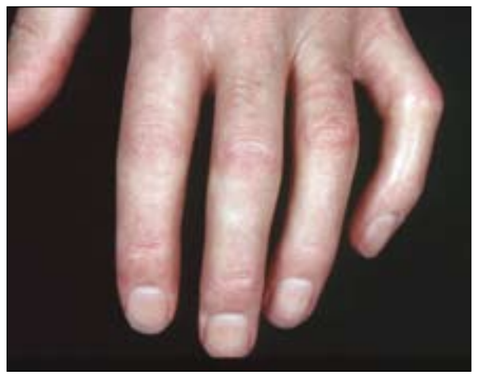
1. Oedematous phase (diffuse puffiness of the fingers and feet)
2. Indurative phase (hard, taut, 'hidebound' – skin becomes adherent to underlying structures)
3. Atrophic phase (softened skin, burnt out).

Sclerodermatous involvement of the hands may lead to a limited ability to extend the fingers and a resultant claw-like appearance, termed sclerodactyly (*Figure 3*). Fingertip ulceration may be seen in severe cases as a result of ischaemia or extrusion of calcinotic lesions. Involvement of the mouth results in microstomia. Facial and digital telangectasiae may be a prominent feature in some patients.

Scleroderma associated with internal involvement is divided into limited cutaneous SSc and diffuse cutaneous SSc.

In limited cutaneous SSc, skin sclerosis is limited to the acral sites (hands and feet), face and forearms. Raynaud's phenomenon is usually associated and may be present for years before sclerodermatous change. Dilated nailfold

Figure 3. Sclerodactyly – a claw-like appearance of the hands caused by tight, thickened skin in patients with systemic sclerosis.



capillary loops may be present – these may be visualized with nailfold capillaroscopy. The entity known as CREST syndrome is a subset of limited SSc, and is associated with anticentromere antibodies in approximately 80% of patients. Patients with limited SSc have a more benign course, with a lower incidence of systemic involvement, such as renal or pulmonary involvement.

In contrast, diffuse cutaneous SSc is characterized by more widespread scleroderma, involving truncal as well as acral sites. There is early and higher incidence of interstitial lung disease, oliguric renal failure, diffuse gastrointestinal disease and myocardial involvement. Anti-Scl-70 (anti-topoisomerase) antibodies may be present.

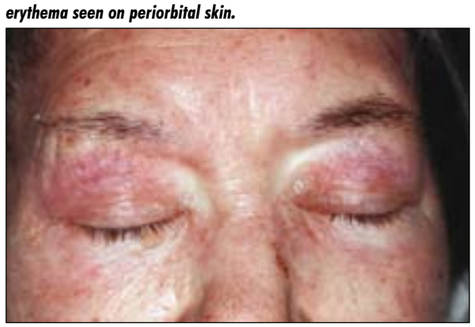
Dermatomyositis

Dermatomyositis is an inflammatory myopathy with characteristic cutaneous features. The five criteria proposed for the diagnosis of dermatomyositis are: proximal muscle weakness, elevated creatine kinase (CK), electromyography consistent with a myopathy, muscle biopsy evidence of myositis and compatible cutaneous disease.

The characteristic cutaneous features of dermatomyositis are the heliotrope rash and Gottron's papules. The heliotrope rash is a violaceous or dusky erythematous rash usually involving the periorbital skin (*Figure 4*). The rash is symmetrical and is commonly associated with oedema. A similar rash may be seen in a photosensitive distribution, as well as the extensor surfaces. Gottron's papules are elevated violaceous papules and plaques over bony prominences, particularly on the dorsa of the hands. As with other connective tissue diseases, nail fold inflammation and telangectasiae are also seen.

In dermatomyositis, three main mechanisms provide a link to respiratory disease. First, dermatomyositis may be a paraneoplastic phenomenon – bronchial carcinoma is the most common associated malignancy in males. Second, muscular weakness as a result of myositis may affect the intercostal and thoracic musculature – this leads to difficulty breathing and may cause aspiration pneumonia. Finally, interstitial lung disease may occur as a result of dermatomyositis. This lung disease is typically associated with the presence of antiaminoacyl-tRNA antibodies, notably anti-Jo-1 and with anti-tRNA antibodies such as anti-tRNA.

Figure 4. The 'heliotrope rash' of dermatomyositis – violaceous erythema seen on periorbital skin.



The antisynthetase syndrome consists of dermatomyositis in association with interstitial lung disease, arthritis, Raynaud's phenomenon and mechanic's hands, caused by antibodies against histidyl-transfer ribonucleic acid synthetase (Jo-1). Mechanic's hands denotes callous-like hyperkeratotic change on the radial surfaces of hands or feet, and is thought to be a specific marker for myositis (Mitra et al, 1994).

Ehlers–Danlos syndrome

Ehlers–Danlos syndrome (EDS) is a group of inherited connective tissue disorders unified by fragility of the skin and blood vessels, hyperextensibility of the skin and joint hypermobility. There are currently nine phenotypic subtypes of EDS (subtypes I–VIII and X). The basic defect in EDS predominantly involves the synthesis, structure or function of one of the fibrillar collagens.

Pneuomothoraces are seen in vascular EDS and, less commonly, in classical EDS. Vascular (type IV) EDS is a severe form characterized by spontaneous rupture of the large arteries, colon and gravid uterus. Dissecting aortic aneurysm is a common cause of sudden death in this form. Classical EDS (types I and II) is more common; patients have cutaneous manifestations of soft, hyperextensible skin which bruises easily. Trivial trauma to skin may result in atrophic 'cigarette paper' scarring and spongy 'molluscoid pseudotumours'. Joint hypermobility is seen in classical EDS, but is associated more with type III (hypermobile) EDS which has minimal cutaneous features.

Vasculitides

The vasculitides are a diverse group of disorders characterized by inflammation of the blood vessels. The systemic vasculitides may involve vasculature of different sizes, hence the lungs and skin may be concurrently affected. The most common vasculitides with lung and skin involvement are Wegener's granulomatosis, Churg–Strauss syndrome (CSS), microscopic polyangiitis and Behcet's disease.

Wegener's granulomatosis

This is a rare disease characterized by necrotizing granulomatous vasculitis of the upper and lower respiratory tracts, and kidneys. The antineutrophil cytoplasmic antibody, c-ANCA, with PR3 specificity is very specific for Wegener's granulomatosis.

Symptoms involving the upper or lower respiratory tract are present in up to 80% of patients and at presentation more than 70% of patients will have nasal, sinus, tracheal or ear involvement. Otitis, epistaxis, rhinorrhoea and sinusitis are common upper respiratory presentations of Wegener's granulomatosis. A saddle-nose deformity may result from necrotizing granulomas of the nasal mucosa. Lower respiratory presentations include cough, dyspnoea, chest pain and haemoptysis.

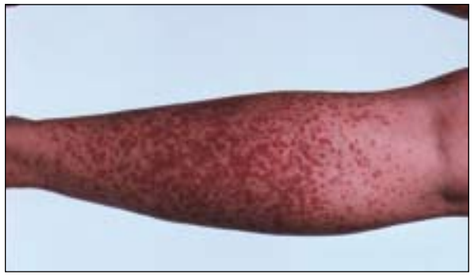


Figure 5. The classic 'palpable purpura' of vasculitis.

Cutaneous lesions will develop in approximately 40% of patients, but are not usually the presenting symptom. Common skin lesions seen in Wegener's granulomatosis include cutaneous vasculitis with palpable purpura, subcutaneous nodules, papules and ulcers (*Figure 5*). The necrotic ulcers seen in Wegener's granulomatosis often have a heaped-up border and be mistaken for pyoderma gangrenosum. The ulcers are usually on the lower extremities but cases of genital ulceration have been reported. Oral mucosal ulceration may also be seen, as well as gingival hyperplasia with petechiae. Less common skin findings reported include, vesicles, pustules, haemorrhagic bullae, petechiae, livedo reticularis and subungual splinter haemorrhages.

Churg–Strauss syndrome

CSS (allergic granulomatous angiitis) is a rare multisystemic disease characterized by asthma, eosinophilia and vasculitis.

CSS has three clinical phases, which may or may not be sequential: the prodromal or allergic phase is characterized by asthma, which may be accompanied by rhinitis. The second (eosinophilic) phase is marked by peripheral blood eosinophilia and eosinophilic tissue infiltration. The third (vasculitic) phase manifests as a vasculitis, which can involve any organ. Constitutional symptoms such as weight loss, fever and malaise are non-specific and may be seen in any phase.

Skin involvement occurs in more than two thirds of patients. The clinical picture is variable, and different morphologies may be seen simultaneously. Urticarial weals, tender subcutaneous nodules, palpable purpura, livedo reticularis, erythematous macules and papules resembling erythema multiforme, and bullous lesions are some of the morphologies that may be present in CSS.

Yellow nail syndrome

The yellow nail syndrome comprises a classic triad of yellow nail discoloration in association with lymphoedema and pulmonary features.

The nails in the yellow nail syndrome are thickened, slowly growing and have an increased lateral curvature. The lunulae and cuticles may be absent. Onycholysis (separation of the nail from the nail bed) and transverse ridging may occur. The nail colour varies from pale yellow to green. These nail changes are secondary to congenitally hypoplastic lymphatics – this is also the cause of the lymphoedema seen in this syndrome.

The lymphoedema is usually symmetrical, non-pitting and involves the lower extremities.

Yellow nail syndrome was first described in association with pleural effusions. Typically, the effusions are exudates (protein concentration >3.0 g/dl) and may be unilateral or bilateral. They often recur after drainage and may require pleurodesis. A variety of other pulmonary manifestations have been described, including bronchiectasis, recurrent respiratory infections, sinusitis and fibrosis.

The nail changes often are the earliest presentation of yellow nail changes and may precede the lymphoedema or lung manifestations by years. For this reason, patients who present with these nail changes should have long-term follow up.

Conclusions

Cutaneous signs may provide invaluable diagnostic clues to an underlying systemic diagnosis. This article has illustrated several cutaneous abnormalities secondary to respiratory pathology, as well as skin and lung manifestations of multisystemic disease. It highlights the importance of recognizing cutaneous manifestations of pulmonary disease, as this may impact on the diagnosis and management of a patient. BJHM

Conflict of interest: none.

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KEY POINTS

- Recognizing cutaneous signs may provide invaluable diagnostic clues to underlying respiratory pathology.
- Skin abnormalities may occur as a consequence of respiratory pathology. Examples include clubbing, cyanosis and superior vena cava obstruction.
- Skin and respiratory manifestations may also occur as part of a multisystem disorder, such as sarcoidosis, tuberculosis, connective tissue diseases and vasculitides.
- Sarcoidosis probably represents the most common multisystem disease with both skin and lung manifestations. Cutaneous sarcoidosis may present as papules, plaques, nodules, lupus pernio and erythema nodosum, while the most common lung abnormality is hilar lymphadenopathy.