Hand Lesions: An Unusual Presentation to the Acute Medical Take

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Abstract

A 66 year old gentleman presented with painful hand lesions, fevers and a new pansystolic murmur. The initial working diagnosis of infective endocarditis was ruled out by multiple negative blood cultures and a normal transoesophageal echocardiogram. Skin biopsies of the lesions were highly suggestive of cutaneous T cell lymphoma. This is a rare disease but should be considered as part of the differential diagnosis of hand lesions. It is incurable but often has a long indolent course. Management depends on the stage and the subtype and the treatment is wide ranging from topical to systemic therapies.

Keywords: Cutaneous lymphoma; Lesions hand blisters

Key Points

- The differential diagnosis of hand lesions is wide ranging
- A new murmur with a fever is Infective Endocarditis until proven otherwise
- Cutaneous T cell lymphomas are a group of heterogeneous Non-Hodgkin’s lymphomas
- The presentation of cutaneous T cell lymphoma is wide ranging and often the diagnosis is not obvious
- There is no cure, however, most primary disorders have a long protracted and indolent course
- A multidisciplinary approach should be taken with involvement of haematologists, dermatologists and pathologists
- Management of cutaneous T cell lymphoma depends on the stage and the subtype

Case History

A 66 year old gentleman was referred to the acute medical take with a one month history of painful lesions on his hands. They began as white papules, which over time developed into larger necrotic lesions. He described no other symptoms and was normally fit and well. He was a retired carpenter with a 30 pack-year smoking history. Observations revealed multiple swinging pyrexias of 38 degrees, heart rate of 60 bpm, respiratory rate of 20, blood pressure of 127/93 mmHg and oxygen saturations of 98% on air.

Clinical examination revealed violaceous nodules on the tip of the right index finger and the base of the right thumb with a central area of necrosis. There was also a small macule over the proximal inter-phalangeal joint of the middle finger as depicted in figure 1. On auscultation a pansystolic murmur was noted. The remainder of the examination was unremarkable. 12 lead ECG showed sinus rhythm with a prolonged PR interval. Urine dip showed no microscopic haematuria and 3 sets of blood cultures from 3 different sites were sent for MC&S. Blood tests revealed a normal white cell count and CRP.

The initial working diagnosis was Infective Endocarditis as the hand findings were suggestive of Janeway lesions. This was also in combination with a new murmur and first degree heart block. Transthoracic echocardiogram showed no evidence of endocarditis so a transoesophageal echocardiogram was performed which was also normal.

The differential diagnoses were then widened to include a cutaneous vasculitis, a neoplastic phenomenon or a cutaneous presentation of a sexually transmitted infection.

To further narrow the differential blood tests including HIV and Syphilis were sent, all of which were negative. A vasculitic screen including ANA, ANCA and ESR was requested which showed no abnormality. Hepatitis B and C serology were also taken, as a leucocytoclastic vasculitis could be a cutaneous manifestation of hepatitis C, secondary to cryoglobulinaemia.

A CT scan of his thorax, abdomen and pelvis was performed to look for any evidence of an occult malignancy, which was entirely normal. A skin biopsy showed a heavy lymphoid infiltrate which extended into the superficial epidermis. The remainder of the epidermis was hyperplastic with massive infiltration of lymphoid cells. On immunohistochemistry...
the majority of the cells were T cells with loss of the CD5 T cell marker, which is highly suggestive of cutaneous T cell lymphoma.

This gentleman was then seen by the haematology team who started him on topical Fucibet cream. He was subsequently discharged home. In follow up clinic the lesions were responding well to treatment. The haematologists suggested a very low threshold for future CT scanning to look for any evidence of lymphadenopathy and advised frequent monitoring and follow up.

Discussion

Cutaneous T cell lymphomas (CTCL) are a group of heterogeneous Non-Hodgkin’s lymphomas. There are many different subtypes however Mycosis Fungoides and Sézary Syndrome are the most common variants [1]. The condition was first described in 1806 by Jean-Louis-Marc Alibert, a French dermatologist who named it Mycosis Fungoides as the tumours were similar in appearance to mushrooms [2].

Incidence and aetiology

CTCL is an uncommon malignancy with an incidence of 0.4 per 100 000 per year [3]. The prevalence is, however, much higher as the malignancy generally has a long indolent course with long survival rates. The disease is more common in Afro-Caribbean’s and about twice as common in men [3].

The aetiology of this condition is not clear. Genetic, infectious agents and environmental factors have all been proposed as contributing factors; however none of these claims have been proven.

The presentation of CTCL can be highly variable. Mycosis Fungoides, which accounts for nearly 50% of all cases, is most often found on sun-exposed areas [4]. The lesion commonly starts as an erythematous annular macule. This is most often a very slowly progressive lesion before turning into an eczematous area. This is known as the patch stage and may be difficult to distinguish from eczema or psoriatic conditions. In the tumour stage these patches become increasingly irregular and develop a more lump-like appearance and may metastasize to other organs.

Sézary syndrome is a leukaemic variant and is much less common than Mycosis Fungoides. It affects the whole skin and presents as a generalized erythroderma with lymphadenopathy. Extracutaneous disease is more common in Sézary syndrome. Any organ may be involved and hepatosplenomegaly is well recognised [5].

Diagnosis of CTCL must be made with histological and immunophenotypic evidence. Multiple ellipse biopsies are advised to confirm the diagnosis [3]. Classically the papillary epidermis is involved with a band-like infiltrate [1]. Blood samples should be taken for white cell count, renal function and more specifically LDH, Sézary cells, CD4/CD8 ratio, human T cell lymphotropic virus and TCR gene analysis. This aids with staging. Bone Marrow aspirates or Trephine biopsies may also be taken to look for other variants of CTCL. Patients should also have a staging CT thorax abdomen and pelvis, which can then help with monitoring disease progression. An important factor in the diagnosis and management of this condition is the early involvement of a multidisciplinary team.

Stage

The most commonly used classification system is the Tumour, Node, Metastasis (TNM) system. This system was adopted by Bunn

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Table 1: Tumour, Node, Metastasis (TNM) system [14].
and Lamberg in 1979 and uses both clinical and histopathological information (Table 1) [6]. Blood staging is a recent addition as blood involvement has been shown to be a major prognostic factor [7]. A revision of this whole system has, however, recently been proposed as it has been shown that other variants of CTCL do not display the same clinical course or prognosis as Mycosis Fungoides or Sézary syndrome [8].

**Management**

A multidisciplinary approach should be used for all cases of CTCL with involvement of haematologists, dermatologists, pathologists and specialist nurses. Repeat ellipse biopsies should be taken in order to confirm the diagnosis before any treatment is started [3].

In early stages of the disease topical therapy alone is sufficient. Photochemotherapy (PUVA) has shown good response rates, especially in earlier stages of the disease [3,9]. Thick plaques can be treated with radiotherapy, as these are very radiosensitive and is often combined with PUVA therapy. Immunotherapy with alpha interferon has shown response rates of up to 80% in early stages of the disease [10].

Chemotherapy should not be used in early stages of the disease as it is relatively chemo-resistant, and results are not long lasting [11]. Newer agents such as monoclonal antibodies, retinoids and extracorporeal photophoresis treatments have also been used but there are no randomised control trials to assess response rates.

**Prognosis**

Most cases of Mycosis Fungoides have a long protracted and indolent course, however prognosis depends on the type, the stage and the presence of extra-cutaneous manifestations. In patients who remain at patch only stage have similar survival rates to the general population with 10 year survival rates of around 98% [12]. If there is lymph node involvement this drops to 20%. Sézary syndrome is much more aggressive and the prognosis is therefore poorer. One study found that the median survival was 31 months and the five year survival was 33.5% [13]. Patients with this leukaemic variant generally die of opportunistic infections [4].

**Summary**

The differential diagnosis of hand lesions is wide ranging, and therefore presentation to the medical take provides a diagnostic challenge. CTCL, although rare, should be considered in the differential diagnosis once more common pathology has been ruled out. Once a diagnosis has been made care should shared by a multidisciplinary team with regular observation and follow up.