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CE - MEDICAL ILLUSTRATION

Dupuytren's contracture… or is it?

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Clinical case

A 90-year-old previously healthy man presented with progressive thickening of the skin of his left hand over the prior 4 months. It had become severe enough to limit movements of the fingers of his hand. He had received localized steroid injections for a presumptive diagnosis of 'Dupuytren's contracture' with no symptomatic improvement. He reported no prior or current alcohol use, no cigarette smoking, and did not take any scheduled medications. He reported no trauma to the hands, occupational or accidental. Vital signs were unremarkable but examination of the left hand showed a 6 cm mass involving the thenar eminence partially obliterating the first webspace, with atrophic and shiny overlying skin (Fig. 1a). Basic laboratory studies including blood counts, renal, liver and thyroid functions were within normal limits. The patient was referred to a hand surgeon, and a subtotal resection of the mass was performed. Histopathology demonstrated a nodular proliferation of medium–large-sized, aberrant T-lymphoid cells, concerning for a lymphomatous process. A positron emission tomography (PET) scan showed diffuse uptake in the left hand without systemic uptake (Fig. 1b). Based on the microscopic features and PET findings of isolated hand involvement, a diagnosis of solitary acral CD4 T cell lymphoma (TCL) was made. Despite localized radiation therapy, oral vorinostat (histone deacetylase inhibitor) treatment and ultimately amputation of the hand due to gangrene, he died a few weeks later.

Our patient was initially thought to have a Dupuytren's contracture, which is a common condition usually affecting the medial hand digits. Palmar fascia fibrosis progressively results in the formation of nodules and bands that limit flexion [1]. Involvement of the thumbs and index fingers is rare and a clue to a potentially different etiology. It is commonly associated with alcohol use, cigarette smoking, diabetes mellitus, repetitive hand trauma and with other fibrosing conditions such as plantar fibromatosis and Peyronie's disease. Our patient had none of these associations; it is exceedingly rare in patients without history of heavy alcohol use or smoking [2].

Cutaneous TCL usually presents as non-resolving plaques, patches or erythroderma with variable distribution although very few cases isolated to the hand have been reported [3]. CD4 TCL is an extremely rare form with potential for aggressive disease [4]. It can mimic more common dermatologic conditions leading to a delay in diagnosis. Outcomes are typically poor despite aggressive therapy.

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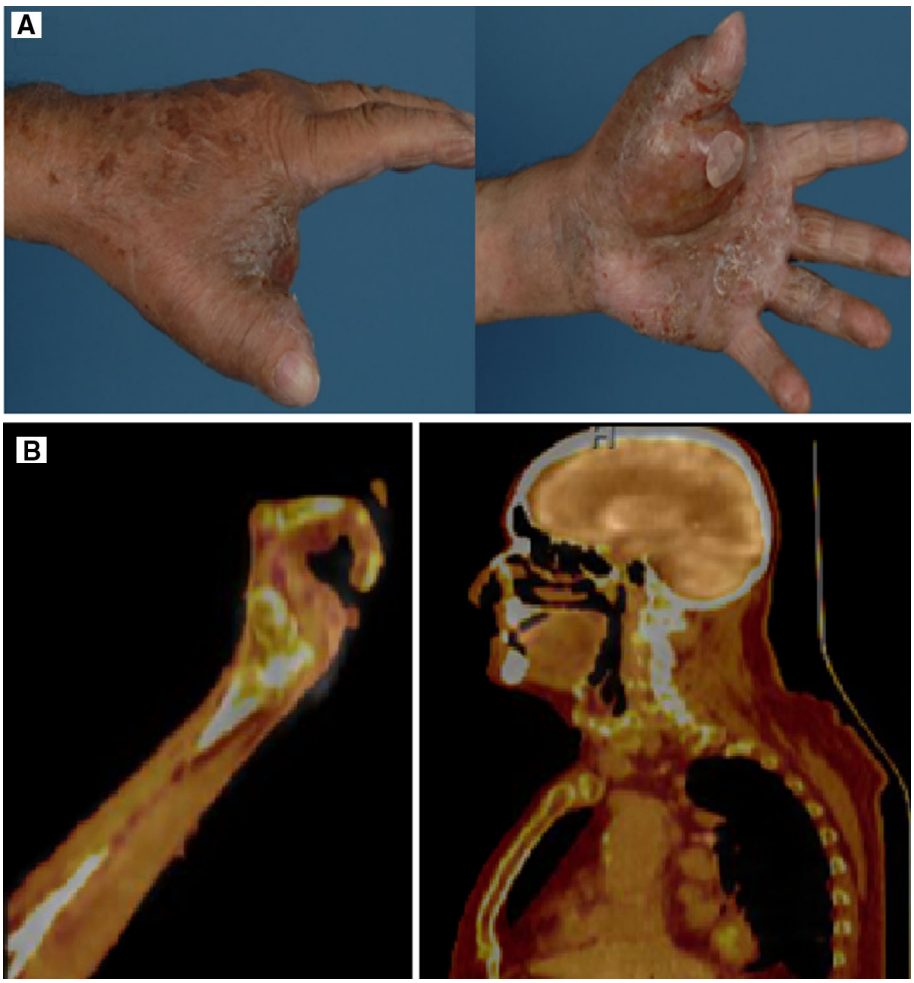

Fig. 1 **a** Physical examination revealed a large nodular mass in the first webspace of the left hand extending over the thenar eminence, with overlying erythema and atrophic skin. **b** PET imaging showing diffuse uptake in the left hand with absence of uptake throughout the rest of body suggesting localized disease within the hand

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Statement of human and animal rights All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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