Eponyms in medicine revisited

Martorell’s ulcer

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Summary
This paper reports a rare form of ulceration of the lower leg and, as a result of subsequent investigations and literature review, re-addresses a recent debate regarding the legitimate classification of these ulcers as a separate disease entity

Keywords: Martorell’s ulcer, hypertensive ischaemic ulcer, microvascular changes

Potted life history of Martorell

Otzet Fernando Martorell, Spanish-born cardiologist (1906–84). First to report ‘hypertensive ischaemic ulcers’ in 1945 (four female cases). Also reported ‘pulseless disease’ and aortic arch syndrome.

Box 1

Figure 1 Medium-sized artery with intimal and medial hypertrophy

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Painful ulceration of the lower leg in association with diastolic arterial hypertension was first identified by Otzet Fernando Martorell¹ who referred to them as ‘hypertensive ischaemic ulcers’. Whilst histology of these ulcers has traditionally indicated characteristic changes, in recent years the specificity of microvascular changes described by Martorell (and associates) has been questioned, shedding doubt upon the legitimate classification of these ulcers as a separate disease entity.

We have recently identified a patient who presented with ulceration consistent with the clinical appearance of Martorell’s ulcer and considered it necessary to re-address the specificity debate.

Case report

A 46-year-old woman referred to the Wound Healing Research Unit by her general practitioner, presented with painful ulceration of the anterolateral aspect of her left lower leg. It appeared that the episode began some months previously with a painful red blister in the skin. The blister increased in size over subsequent weeks and following treatment with simple dressings a small area of the skin broke down. This area increased in size to the point of examination when the indurated area was of approximately 3 cm in diameter with a uniform depth of 1.5 cm. There had been little in the way of exudate and there was no evidence of slough. It was also noted that there was a small red spot on the same area of the right shin.

The patient’s systemic blood pressure was 179/95 mmHg (just within hypertensive parameters) and at the ankle her systolic pressure was 220 mmHg giving an Ankle Pressure Index of 1.29. The differential diagnosis was vasculitic ulcer, an ulcerated erythema nodosum, or perhaps Martorell’s ulcer.

The primary objective was to eliminate other causes before labelling it Martorell’s ulcer. It was therefore arranged for her to have full blood count, liver, thyroid and renal function tests, C-reactive protein, erythrocyte sedimentation rate, antinuclear factor, rheumatoid factor, anti-mitochondrial antibodies, anti-thyroid antibodies, glucose and chest X-ray. The wound was dressed with an alginate and arrangements were made for weekly review.

At her next appointment no significant wound changes were noted and a 6-mm punch biopsy was taken from the wound edge. Results of previous tests were unremarkable. Histology report from the punch biopsy supported a diagnosis of Martorell’s ulcer. The report revealed no evidence of vasculitis or neoplastic disease and showed a medium-sized artery with intimal and medial hypertrophy (figure 1). On the basis of the report the patient’s ulcer, during her third visit, was dressed with an alginate soaked in Brilliant Green (1% aqueous solution).

By the patient’s fourth visit wound dimensions were found to be half the original, the ulcer was reported to be less painful and the patient stated that she could sleep at night without being disturbed by pain. Further consultation was arranged on a fortnightly basis until complete wound closure had been achieved (week 14: figure 2). It was during this period that the literature was reviewed and the specificity debate re-addressed.

Literature review

The ulcer has been thought to be caused by obliterating lesions of small arterioles.² These lesions were held to be consistent with lesions found in other localities in essential hypertension (eg, narrowing of retina and renal arterioles). Whilst slight local trauma was felt to be significant to the development of arteriolar lesion and subsequent ulceration it has been acknowledged that the ulcers also develop in the absence of trauma.

Histology² of these ulcers show an increase in the size of the arteriolar wall (hypertrophy of media musculature; enlargement of intima; figure 1) and a decrease in the diameter of the lumen tending to luminal stenosis. Occasionally hyalinosis of the media (figure 3), obliteration of the lumen by thrombi or

Figure 2 Wound closure

Figure 3 Hyalinosis of media musculature

Martorell’s ulcer: diagnos-
tic criteria

- hypertension
- palpable pulses in the legs
- absence of disturbance in venous circulation
- superficial necrosis of anterolateral aspect at junction of mid and lower leg
- symmetry of lesions
- absence of arterial calcification

Box 2

proliferating intima and periarteritis of small arteries and arterioles has been found.

Signs and symptoms are as detailed in the case report but may vary slightly. The first symptom consistently reported in the literature is that of a painful red blister in the skin which soon becomes blue and purpuric. Superficial necrosis develops and circular ulceration appears. Typically ulcers have been bilateral and symmetrical. They locate on the anterolateral aspect of the leg at the union of the lower and middle thirds. When ulceration is unilateral then a simple pigmented spot has usually been present on the corresponding site of the opposite leg. Ulcers have varied between 1 and 11 cm in diameter. Previous reports note that the patients’ pain is not relieved by rest or elevation. The traditional diagnostic criteria are shown in box 2.¹

There is no record of the ulcer’s incidence or prevalence, although a number of cases have been reported in the literature (table). Both medical and surgical treatment options have been pursued: excision has produced rapid healing; excision and grafting achieves ‘quick healing’; control of blood pressure by lumbar sympathectomy has been known to bring about complete healing in 10 days⁷; and, the administration of antihypertensives and vasodilators with the topical application of Brilliant Green (a triphenylmethane dye) has effected healing within approximately two months.⁶

Specificity debate

The characteristic clinical appearance of these ulcers has not been disputed. However the underlying arterial/arteriolar pathology has been contended by two authors in particular. Kuiper¹⁰ reasoned that if hypertension is a common disorder of the elderly population then the (presumably) low incidence and prevalence of Martorell’s ulcer would suggest that hypertension alone is rarely responsible for the development of skin necrosis. However, hypertension is commonly a disease of the male population and Martorell’s ulcer is predominantly female, a possible explanation for infrequent reports of the ulcer’s apparent prevalence. Furthermore control of blood pressure has played a significant role in the treatment of these ulcers, thus establishing the correlation.

Kuiper also argued that the microcirculatory lesions associated with the ulcers could be attributed to a variety of conditions (including chronic venous insufficiency and diabetes) and in old age in the absence of hypertension, disputing the specificity of the ulcer’s claimed aetiology. However, whilst these arterial and arteriolar lesions have been reported at various sites (eg, retina, spleen, kidneys, pancreas, liver and adrenal glands) lesions proximal to the skin have rarely been documented outside of hypertension.¹¹ Furthermore lesions found at least common sites (normotensively) are related to a younger population with comparatively more rapid disease onset and progression, namely acute microbial infection and inflammatory disease.¹²

Leu⁹ compared the histological findings in Martorell’s ulcer with those in and around ulcers due to chronic venous insufficiency. Biopsy or excision of the entire ulcer was performed from seven patients bearing the clinical diagnosis of Martorell’s ulcer. The findings were compared with those obtained from 27 patients with ulcers allegedly of venous origin. In all cases the results with Martorell’s ulcers showed, amongst other things, numerous small arteries and arterioles with thickened media consisting of hyperplastic smooth musculature or hyaline tissue. Twenty-five of the 27 venous ulcers did not reveal similar changes of the small arteries and arterioles. Of the two which were claimed to have shown arterial and arteriolar changes identical to Martorell’s ulcers, the first patient was known to be hypertensive and the second was known to have arterial disease. It was later recorded that the ‘identical’ changes claimed in the two referred to were of a ‘lesser’ degree to that of patients with Martorell’s ulcer.

Table Cases reported in the literature

Year	Author	No. of cases	Comments
1945	Martorell ¹	4 female	first report
1946	Valls-Serra ³	1 male	first male case
1946	Hines and Farber ⁴	11 female	variation in the site of the ulcer
1948	Wright ⁵	1 male	2nd male case
1951	Alonso (cited in ⁶)	1 case	
1954	Alonso ⁶	1 female	
1958	Monserat ⁷	1 female	
1966	Schnier <i>et al</i> ⁸	40 cases	variability of the selection criteria used
1992	Leu ⁹	7 cases	histological changes described

Learning points

- presently insufficient grounds on which to question the traditional classification of Martorell's ulcer
- large scale studies involving more comprehensive investigation of skin vasculature necessary.

Box 3

Conclusion

On the basis of the evidence presented there does not appear to be sufficient grounds to question the specificity of the histological (in particular small artery and arteriole) changes associated with Martorell's ulcer. Nevertheless additional research is required to finalise whether or not the ulcer may be legitimately classified as a separate disease entity. Clearly this will necessitate strict inclusion/exclusion criteria and large sample populations. It would also be advisable to perform more comprehensive physiological investigation of skin vasculature. Tests could include *in vivo* skin surface microscopy and laser doppler flowmetry. The presence/absence or magnitude of the reflux drop in blood pressure and flow upon postural change again could be investigated. Hence local circulatory problems, possibly an additional critical factor in those few individuals with hypertension who are prone to Martorell's ulcer, could be identified or ruled out. Likewise it would be prudent to test for more generalised arteriopathy.

Given the rarity of Martorell's ulcer it would be necessary before any large scale studies may be performed to first identify prevalent cases. To assist identification it is proposed that an interactive clinical database be established to which wound care practitioners may provide or extract information pertaining to this condition. The network advocated would provide an invaluable resource serving future investigation and may also be further developed to incorporate additional forms of rare ulceration.

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