Is EMLA Effective in Dercum's disease?

Sir,

Adiposis dolorosa or Dercum's disease (DD) is characterized by overgrowth of skin in some body areas with tenderness at first, then pain: the pain can be continuous or cyclic. It predominantly occurs in postmenopausal women with a tendency to fatness, fatigue and psychic disturbances; sometimes a dominant inheritance can be proved (1, 2). Histopathologically DD is indistinguishable from ordinary lipomas, normal fat cells like those of the subcutaneous tissue. Occasionally the histological appearance is that of an angiolipoma, or granulomas of the foreign body type have been noted within the fatty tissue (3). Right from the beginning there is tenderness, later followed by pain: it lasts for hours and worsens with movement, compelling the patient to immobility during attack.

CASE REPORT

The patient was a 50-year-old woman, a Catholic nun. Her medical history was uneventful apart from a surgical menopause caused by uterine fibromata at the age of 43. Since the age of 30 the patient had suffered from migraine treated with NSAIDs.

A limp and aching swelling, covered with a lightly erythematous skin, occurred on the patient's right hip: it was removed by a surgeon, without result. Four months later another swelling, similar to the first and symmetrically disposed, occurred on the left hip.

Later on, analogous lesions developed on the shoulder girdles and arms.

All examinations (laboratory tests, X-ray, echography, electromyogram) were normal.

A skin biopsy showed the histological picture of lipoma; electron microscopy, too, indicated no difference from the ultrastructural appearance of lipoma.

This together with chemical features, their length, the patient's weight increase (from 52 to 60 kg) and psychic weakness that appeared during the stay in hospital led to the diagnosis of DD.

The following treatments were tried without any success: intravenous lidocaine, mexiletine orally given and methylprednisolone i.m.

Psychotherapy carried out in the same time was of no benefit. Liposuction was suggested: the patient, after carefully weighing up the benefits and risks, refused it.

A eutectic mixture of local anaesthetics (acronym EMLA) brought relief to the patient even if for just a short time. EMLA emulsion (Astra), applied under occlusion for half an hour (and afterwards washed away) only on those areas where pain was more severe (no more than 2,000 cm^2 in any case), secured the patient 3 h of analgesia.

EMLA produced this effect every time (almost daily) the cream was applied without lowering of analgesia time during the days of observation as inpatient.

The level of analgesia achieved after EMLA removal was enough for pin-prick anaesthesia. In the follow-up, 3 months later, the EMLA effect on our patient's pain was unchanged. In this period EMLA was discontinued and a placebo cream was applied like EMLA for 10 days, during which the patient had no benefit: under pin-prick test the patient could determine where (right-left) EMLA-placebo had been applied.

DISCUSSION

The effectiveness of intravenous lidocaine in delaying the frequency of DD pain fits, as well as lowering it, has not been proved in our patient because of the adverse psychic side-effects (sexual dither, awkward especially for a nun, even after the infusion of 200 mg) that added to cardiocirculatory ones peculiar to the drug.

Cushing syndrome appeared shortly after the beginning of the treatment with corticosteroid, and the poor effectiveness of the treatment itself led to its interruption. Only application under occlusion of EMLA brought transitory (about 3 h) relief to the patient.

The risk of sensitization to prolonged application of a drug on the skin in a chronic disease, the width of the area under treatment and the following hazard of methaemoglobinemia (6, 7) meant that, in this particular case, use of this anaesthetic emulsion was restricted to the moments of maximum pain intensity.

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