Posters

EXPLORATORY THERAPEUTIC APPROACH IN DERCUM'S DISEASE: IMMUNOMODULATORS AND GLP-1 RECEPTOR AGONISTS

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Dercum's disease (DD) is a rare chronic disorder characterized by painful subcutaneous lipomas, predominantly affecting overweight or obese middle-aged women. According to lesion distribution and extent, Hansson et al. classified DD into four types: generalized diffuse (I), generalized nodular (II), localized nodular (III), and juxta-articular (IV), although mixed forms may occur [1]. The aetiology remains unclear, and evidence for medical treatments is limited. Surgical approaches may reduce pain but are often followed by relapse and are difficult to apply in extensive disease. We report a patient with DD treated with multiple medical strategies. A 66-year-old woman with a mixed type II+IV DD has been followed since 2023 at the Rare Disease Unit, Section of Dermatology, University of Florence. Visual Analogue Scale (VAS), Dermatology Life Quality Index (DLQI), and BMI were assessed at each visit. She presented with painful subcutaneous masses, first noted 15 years earlier and recently worsened, associated with a 20 kg weight gain. She also reported memory and gait impairment. Examination revealed multiple discrete, painful lipomas (ultrasound-confirmed) in the right popliteal region, thighs, and trunk, along with diffuse cutaneous hyperalgesia. Diagnosis of mixed type II+IV DD was made. Baseline DLQI, VAS, and BMI were 8, 6, and 27.6 kg/m^2, respectively. Infliximab (5 mg/kg i.v at weeks 0, 2, 6, and 15, followed by 120mg s.c. every two weeks starting at week 24) was initiated and was associated with clinical improvement, lasting approximately two years. At clinical worsening, methotrexate (12.5 mg s.c. weekly) was introduced. Given metabolic comorbidities (overweight and type 2 diabetes), semaglutide (3 mg daily, increased to 7 mg after 4 weeks) was added. The patient experienced progressive pain reduction, together with a total weight loss of 6 kg over 15 months. Given the favorable clinical response, infliximab was discontinued at T30, with no relapse at T36. Infliximab was apparently associated with a stable improvement in quality of life, VAS and BMI (DLQI −88%, VAS −100% and BMI −4.3%) over 21 months. The combination therapy methotrexate + semaglutide was also associated with improvements (DLQI -83%, VAS -80%, BMI -10%) lasting for 15 months, ongoing. Immunomodulators and incretin-based therapies may be a promising strategy in DD patients refractory to conventional therapies and with extensive disease, where surgery is not feasible.

References

1. Hansson, E.; Svensson, H.; Brorson, H. Review of Dercum's Disease and Proposal of Diagnostic Criteria, Diagnostic Methods, Classification and Management. Orphanet J Rare Dis 2012, 7, 23, doi:10.1186/1750-1172-7-23.