alcohol sponge, the needle is inserted into the mass under palpating guidance. When the needle is secured in the mass, the cylinder is retracted, establishing negative pressure. Then, while the aspirating negative pressure is constantly maintained, multiple short, quick insertions of the needle are performed in various angulations, but the direction of the needle is maintained. Before withdrawal, the negative pressure is discontinued. The needle is disconnected and pulled out separately. The cylinder is further retracted, the needle is remounted, and its contents are expelled on the glass slides to prepare a thin, even smear from the aspirated material.

3. We disagree with Dr. Ackerman that “unguided, direct FNA is not only not indicated, but is unsafe.” As opposed to his comment, we do recommend that this safe procedure should be routinely performed as the initial bedside diagnostic method in patients with palpable abdominal masses. Only when direct and repeated aspirations fail to obtain adequate material should imaging-guided FNA be considered.

4. Intestinal penetration and aspiration of its material should be avoided whenever possible. However, we have clinical and cytologic evidence that such an event is not dangerous as long as the negative pressure is discontinued before withdrawal. Abolition of the negative pressure before withdrawing the needle is essential in order to prevent spread of any material, possibly even of tumor cells.

5. The fatal infection after direct FNA performed by the inexperienced pathologist that Dr. Ackerman described just emphasizes what we have stated above. This complication could have occurred with an improper CT-guided FNA procedure, and could have been avoided had proper direct FNA been performed.

YEOUNDA EDOUTE, M.D., PH.D.
SHLOMO AVRAM BEN-HAIM, M.D.
EHUD MALBERGER, D.M.D., F.I.A.C.
Rambam Medical Center and Bruce Rappaport Faculty of Medicine
Technion-Israel Institute of Technology
Haifa, Israel


CLINICAL CHARACTERISTICS AND PATHOPHYSIOLOGY OF ERYTHROMELALGIA AND ERYTHRITEMALGIA

To the Editor:
In their review of the literature on erythromelalgia and allied disorders, Kurzrock and Cohen [1] attempt to divide erythromelalgia into an early-onset (childhood) and an adult-onset form, but also refer to a secondary form. According to them, adult-onset erythromelalgia is idiopathic or secondary to either myeloproliferative disorders with thrombocythemia or other conditions like systemic lupus erythematosus (SLE), hypertension, vasculitis, and so forth. Early-onset erythromelalgia arises in childhood and does not usually have an apparent underlying illness. The basis for this grouping lacks a sound scientific foundation and warrants analysis. In doing so, they namely ignore earlier classifying works on the differentiation between erythromelalgia and erythermalgia [2-4]. There are several noteworthy discrepancies between their newly proposed and other available taxonomies. We would like to draw the readers’ attention to this topic and address the main inconsistencies in their article.

The greatest flaw in their classification is the categorization of different subtypes regardless of their cause. Moreover, their published scheme is not sufficiently supported by original clinical, laboratory, and pathologic data. Our major objection concerns the category “adult-onset erythromelalgia,” since they combine several forms with separate pathophysiology. This leads to unnecessary confusion.

We categorize patients with red, warm, and painful extremities according to their etiology. We discovered erythromelalgia to be causally related to thrombocythemia and separated this entity, since it has been shown that this entity constitutes a distinct disorder with clear diagnostic, pathologic, and therapeutic parameters [5,6]. Erythromelalgia in thrombocythemia is caused by abnormal platelet-mediated arteriolar inflammation and thrombosis [7]. Aspirin provides...
rapid and long-lasting clinical relief due to its irreversible inhibition of platelet cyclooxygenase activity, and can be used as a reliable therapeutic test. Other forms of red, warm, and burning extremities not characterized by thrombocythemia are classified as erythermalgia in order to denote the important aspect of warmth, and can be divided into a primary and secondary form. Primary erythermalgia is a completely separate entity that develops mainly at a young age. These patients presumably have an as yet unknown common pathophysiology distinct from that in patients with erythromelalgia. We have established six diagnostic criteria for the diagnosis. In brief: (1) episodes of local vasodilatation and congestion accompany the increased skin temperature and burning pain; (2) the symptoms are symmetric, bilateral, and persistent; (3) the episodes may be easily provoked and aggravated by exercise and heat; (4) cold, rest, and elevating the affected limbs afford relief; (5) there is no primary or associated disease; and finally, (6) the condition is refractory to any pharmacologic measure [2-4].

Other forms of red, warm, swollen, and burning extremities, regardless of age, in conjunction with several conditions are labeled as secondary erythermalgia, which either originates from associated disorders or develops as a consequence of side effects of drugs. Secondary erythermalgia is basically dissimilar from thrombocythemic erythromelalgia, since in none of these conditions has any platelet dysfunction been shown [2,3]. Consequently, aspirin is not effective, and treatment of the underlying disorder or withdrawal of the incriminating drug leads to resolution of the clinical symptoms. We have evaluated a patient with secondary erythermalgia caused by SLE. Skin punch biopsies from this patient did not show intravascular arteriolar platelet plugging but an inflammatory process compatible with vasculitis. Aspirin was not helpful but immunosuppressive treatment of the SLE resulted in abatement of the symptoms [9].

We agree that there is an imperative need in medicine to discard unwieldy and confusing terms and start again with a rational and simple classification. A careful delineation of patients with red, warm, and painful extremities facilitates treatment. Our categorization reflects original research and has a logical basis [2-4]. The classification by Kurzrock and Cohen [1], however, only confuses matters more, since the categorization into an early-onset and adult-onset form is rather artificial and is not supported by scientific evidence.

J. P. H. Drenth, M.D.
University Hospital Radboud
Nijmegen, The Netherlands
J. I. Michiels, M.D., Ph.D.
University Hospital Dijkzigt
Rotterdam, The Netherlands


The Reply:
Drenth and Michiels believe that our classification of erythromelalgia is confusing. In particular, they have suggested that the variant of red, warm, burning extremities associated with myeloproliferative disorder-related thrombocythemia be termed erythromelalgia, whereas all other forms be termed primary or secondary erythermalgia. In our review [1], we designated as erythromelalgia all forms of the clinical constellation of red, burning, warm extremities in which symptoms are exacerbated by heat or dependency and alleviated by elevation or cooling. We then suggested that this disorder could be subdivided into an early-onset (childhood) form that is generally idiopathic in nature and refractory to aspirin therapy, and an adult-onset form that may be either idiopathic or secondary to a variety of illnesses, the most common one being the thrombocytoysis of a myeloproliferative disease. The latter variant is caused by abnormal platelet-mediated arteriolar inflammation and thrombosis and is rapidly relieved by aspirin.

Drenth and Michiels have made several important contributions to the literature on erythromelalgia and allied disorders by pointing out that the early-onset form of this disorder is resistant to aspirin therapy [2] and probably has a pathophysiologic basis distinct from that of erythromelalgia in adults with increased platelet counts and an associated hematologic neoplasm [3]. Although their group is in general agreement with us in regard to the signs and symptoms of the various forms of this disorder, they raise several objections.