Letter to the Editor

Aspirin Responsive Thrombotic Complications in Thrombocythemia Vera. A Novel Platelet-Mediated Arterial Thrombophilia

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About erythromelalgia

To the Editor,

I have read with enthusiasm and surprised with Dr. Michiels' paper entitled "Aspirin responsive thrombotic complications in thrombocythemia vera. A novel platelet-mediated arterial thrombophilia" in the recent issue of the Journal (2002;19:225-233).

I believe "There is no treatment available" for the idiopathic erythromelalgia/erythermalgia should be deleted from the Table 2 in Dr. Michiels' article, since sodium nitroprusside treatment of ours, was also brought to the attention by Drenth, Michiels and Ozsoylu (19th reference in the article). The success of sodium nitroprusside treatment was also mentioned by Prof. Hans-Gerd Lenard (correspondence).

I can not agree with Dr Michiels' statement about idiopathic erythromelalgia as "an incurable congenital disorder" either. It is definitely treatable and not a congenital disorder. It is hereditary origin is also not evident despite Dr. Michiels statement in item 7 of Table 2; "a hereditary basis of idiopathic erytromelalgia/erythermalgia has become evident". I have seen or consulted most cases of idiopathic erythromelalgia. I would like to see evidence that it is a hereditary disorder.

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Response

Drenth, Michiels and Özsoylu described the clinical symptoms in 9 children with severe but transient acute secondary erythromelalgia/erythermalgia associated with hypertension[1]. Intravenous sodium nitroprusside was effective in ameliorating the erythromelalgic/erythermalgic symptoms with a drop in blood pressure to normal in 5 of these 9 patients[1]. The episodes of transient erythromelalgia/erythermalgia did not recur during follow-up[1].

I have clearly defined the hereditary entity of idiopathic erythromelalgia/erythermalgia, which is very rare and refractory to any treatment including nitroprusside[2,3]. Secondary erythromelagia is an acquired condition and may occur in association with a wide variety of a detectable underlying disorder and usually respond to appropriate treatment of the underlying disease when treatable[4,5]. I fully agree with Dr. Özsoylu that there are a lot of isolated case reports on idiopathic erythromelalgia/erythermalgia in the complete absence of an underlying disease[4,5]. According to my restricted experiences in about 10 to 15 adult patients with persistent life-long erythromelalgia/eryhermalgia, such cases usually did not respond to any treatment including nitroprusside. The literature on the latter group of idiopathic erythromelalgia/erythermalgia in children or adults with no family history and without any evidence of an underlying disorder is very confusing and difficult to interpret what is fiction, fact, imagination or true observation[6,7].

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