Erythromelalgia as a Paraneoplastic Syndrome in a Patient with Abdominal Cancer

Sir,

Erythromelalgia (EM) is a symptom-diagnosis characterized by red, warm and painful extremities. Symptomatic relief is achieved by cooling and sometimes by elevation of the affected limbs. Attacks are initiated or aggravated by exposure to heat and often by exercise and standing. Microvascular arterio-venous shunting combined with tissue hypoxia is thought to cause the symptoms (1). EM can be a primary condition, or secondary to a primary disease. We report here on a patient with EM of the lower extremities who was not previously described.

CASE REPORT

A 37-year-old woman with EM presented at our clinic in December 1989. The patient had had symptoms consistent with irritable colon from the age of 16 years. Constipation had been the predominant symptom and she used laxatives regularly. During pregnancies in 1973 and 1978 she easily developed lower limb oedema with extreme swelling. She had had moderate to severe anaemia during the years 1981–1989, which was thought to be caused by iron deficiency, and was treated with iron supplements. From 1986 she suffered from intermittent burning pain, warmth, redness and swelling of the feet, ankles and legs, which was worsened by standing, walking, running, exposure to the sun, and warm shoes. The symptoms and signs occurred more often during warm weather and in the evening and were relieved by elevation of the feet. The symptoms gradually worsened from 1988 to 1990. She started active cooling by walking barefoot on cold floors and in 1989 the symptoms exacerbated markedly and she cooled the feet in ice-cold water for up to 1 h/day. Symptoms often got worse during her work, where she was standing for 8 h/day. Fatigue and poorly localized abdominal pain. She had noticed increasing constipation with mucous and fresh blood in the stools. She had had symptoms consistent with irritable colon from the age of 16 years. Constipation had been the predominant symptom and she used laxatives regularly. During pregnancies in 1973 and 1978 she easily developed lower limb oedema with extreme swelling. She had had moderate to severe anaemia during the years 1981–1989, which was thought to be caused by iron deficiency, and was treated with iron supplements. From 1986 she suffered from intermittent burning pain, warmth, redness and swelling of the feet, ankles and legs, which was worsened by standing, walking, running, exposure to the sun, and warm shoes. The symptoms and signs occurred more often during warm weather and in the evening and were relieved by elevation of the feet. The symptoms gradually worsened from 1988 to 1990. She started active cooling by walking barefoot on cold floors and in 1989 the symptoms exacerbated markedly and she cooled the feet in ice-cold water for up to 1 h/day. Symptoms often got worse during her work, where she was standing for 8 h/day. Fatigue and poorly localized abdominal pain. She had noticed increasing constipation with mucous and fresh blood in the stools. She had noticed increasing constipation with mucous and fresh blood in the stools.

On clinical examination in November 1989, she had no symptoms and signs of EM. A complete blood count was normal. Inspection of the peripheral blood smear and bone marrow revealed no pathology. Electrolytes, creatinine and urea, albumin and s-protein, glucose, IgE, lactate dehydrogenase, urine examination and liver function tests were all normal. Antinuclear antibodies and rheumatoid factor were negative. Histology from affected areas was normal and evaluation of the same biopsy with immunofluorescence showed non-specific changes with fluorescence against IgM, complement C3, terminal complement complex and fibrinogen in some dermal vessels.

Treatment with aspirin, metoprolol and propranolol during 1989 had no effect. Prostaglandin E1 infusions were tried in March 1990 with excellent relief of the symptoms for 4 months. At this time the patient experienced periods of nausea, reduced appetite, weakness, fatigue and poorly localized abdominal pain. She had noticed increasing constipation with mucous and fresh blood in the stools. Two weeks following prostaglandin E1 treatment she was hospitalized with vomiting and increased abdominal size. During laparotomy 12 l ascites fluid and large tumour masses of the left ovary, sigmoid colon, peritoneum and liver was found. Adnexa, omentum majus, parts of sigmoid colon and metastases of fossa Douglas were extirpated. Histological examination showed middle-differentiated adenocarcinoma compatible with a primary colonic cancer. No irradiation or further surgery was indicated. Intermittent treatment with 5-fluouracil intravenously was tried for 3 months without effect. She then received palliative treatment until she died in November 1990.

This patient is one of 124 cases of EM collected over a period of 15 years (2, 3). In this patient colonic cancer was diagnosed after 4 years of suffering from EM, but the adenocarcinoma was probably undiagnosed for several years. The symptoms of EM gradually worsened when the cancer progressed, consistent with a parallel course of the conditions. It has been reported that myeloproliferative disorders may be preceded by EM (4, 5). In this study 1 patient developed chronic myeloid leukaemia and 4 polycythaemia vera.

REFERENCES


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Cato Mørk1, Ole M. Kalgaard2 and Knut Kvernebo2
1Department of Dermatology, The National Hospital and 2Department of Surgery, Ullevaal Hospital, University of Oslo, Norway.
E-mail cato.mork@rh.nio.no