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To the Editor:

We appreciate Dr. Dyer's comments regarding our review article. We apologize for not citing his abstract. We would agree that circulating neutrophils are not required for PMA-induced increased permeability pulmonary edema, and this would indeed change our conclusion regarding PMAs mechanism(s) of action. In fact, if it were to be shown that neither intrapulmonary nor circulating neutrophils are important, this would add another model where neutrophils are not of prime importance in increased permeability. We agree that it is important to demonstrate that there are no intrapulmonary neutrophils trapped in animals made leukenopic prior to PMA administration. However, we would like to caution Dr. Dyer that histologic detection of intrapulmonary neutrophil trapping is, in our hands, not a very sensitive technique. We have found that, in the ethchlovinyl model of increased permeability pulmonary edema, there is not histologic trapping of leukocytes, but these cells can be found in abundance when simultaneous BAL is performed (unpublished data). However, consistent with our conclusions in the review, these intra-alveolar leukocytes do not play a role in the increased permeability. Thus, Dr. Dyer may wish to look for leukocytes by histologic means and BAL, as well.

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Myocardial Telangiectases in Hereditary Hemorrhagic Telangiectasia

To the Editor:

Hereditary hemorrhagic telangiectasia (HHT) is frequently accompanied by visceral vascular malformations, but heart involvement is extremely rare, and to the best of our knowledge was not described before.

An 85-year-old man was admitted to our hospital with acute pulmonary edema. His medical history disclosed recurrent gastrointestinal bleeding of many years' duration, a barium meal examination done a few years earlier was interpreted as normal. Physical examination revealed pallor of the mucous membranes and the presence of multiple telangiectases on the buccal and nasal mucosa. Recurrent gastrointestinal bleeding occurred and endoscopy was performed, which revealed telangiectases in the gastric mucosa of the greater curvature and antrum and of the duodenum.

Six days following admission, the patient developed an extensive anterior myocardial infarction and died in intractable pulmonary edema. Postmortem examination confirmed the diagnosis of HHT and revealed telangiectases of the skin, oropharynx, stomach, small and large bowel, brain, lung, kidney, spleen and urinary bladder.

The heart was enlarged (540 g in weight). Right atrium was dilated; left and right ventricular walls showed mild hypertrophy. Microscopic examination of the myocardium showed many elongated irregular vascular channels lined by endothelial cells with thickened and hyalinized walls (Fig 1). Hyalinized papillary projections were observed protruding into the vessel lumens.

Subsequently, the patient's daughter was examined and multiple telangiectases were found in her buccal and nasal mucosa, and a diagnosis of HHT was established.

Since the first published case of HHT in 1862 by Babington, many cases were reported in the English literature. Telangiectases aneurisms and/or arteriovenous shunts have been found in the lung, liver, pancreas, gastrointestinal tract, spleen, urinary bladder, kidney, genital tract, brain, aorta, bones, conjunctiva, retina and in the adrenal gland. Cardiac failure without any explanation other than HHT has been described. High cardiac output states due to anemia and systemic arteriovenous shunting accounted for almost all cases of heart disease. No cardiac vascular malformation was found. Bird et al describe an elderly woman with HHT and heart decompensation. At autopsy, a small, irregular shaped subendocardial area of redness was found, probably a telangiectasia; no microscopic description was given. Dilated microvascular lesions can be seen in healed infarcts and in the subendocardial lesions associated with chronic subendocardial ischemia. In the present case the pattern of distribution and the abundance of the myocardial vascular malformations are highly suggestive of telangiectases found also in the patient's other viscera.

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