Angioendotheliomatosis of the Central Nervous System

G. Th. A. M. Bots

Department of Neuropathology of the Pathology Laboratory of the University of Leyden, The Netherlands

Received August 6, 1973; Accepted November 5, 1973

Summary. A case of angioendotheliomatosis is described in which the neoplastic proliferation of the vascular endothelium was restricted to the central nervous system. Although the process resembled a malignant tumour, neoplastic growth did not occur outside the blood vessels. The process was accompanied by a chronic inflammatory reaction in and around the blood vessels.

Key words: Angioendotheliomatosis — Vascular Endothelium — Neoplastic Growth — Inflammatory Reaction.

Introduction

In 1959, Pfleger and Tappeiner described a patient with skin anomalies caused by neoplastic proliferation of the endothelial lining of blood vessels accompanied by a perivascular chronic inflammatory reaction. Since then, a small number of similar cases have been published (Braverman and Lerner, 1961; Strouth et al., 1965; Fievez et al., 1971). In a few of these cases the disease showed the behaviour of an inflammatory process, but in most of them it resembled a neoplastic process. In the latter there was virtually no growth of tumour cells extravascularly. The distinction between the two forms cannot always be made on morphological grounds alone (Lever, 1967). At present, the disease is known under the terms neoplastic angioendotheliomatosis and angioendotheliomatosis proliferans. In addition to the skin, the central nervous system and one or more visceral organs can be affected. The case reported here is the first to be published in which the process was limited to the central nervous system.

Case Report

The patient was a woman aged 73 who was admitted to the Neurology Department of the Leyden University Hospital at the end of 1968 because of dizziness and disturbance of memory, consciousness, and motor function as well as faecal and urinary incontinence. The complaints had started early in the same year and had gradually become worse.

The history included a partial gastrectomy for a gastric ulcer in 1942, rheumatoid arthritis since 1955, and a cholecystectomy in 1967 because of gallstones, after which she suffered a gradual loss of appetite that had finally become severe.

On admission, a partial transverse lesion of the spinal cord at the level of Th 1 was found. The sedimentation rate was 42 mm/h. The hematocrit was 39%; the leukocyte count was 5200; the differential count was: 4% eosinophils, no staff cells, 80% polymorphonuclear cells, 11% lymphocytes, and 5% monocytes. The electrocardiogram was normal. The cerebrospinal fluid showed 7 cells/mm³ and 90 mg-% protein, with a cytological picture of chronic inflammation. There was no fever. The electroencephalographic anomalies were consistent with cerebrovascular insufficiency of the left hemisphere of the brain.

The patient’s condition deteriorated gradually, the transverse lesion became total, and 2 months after admission she became comatose. Bronchopneumonia developed and she died.
Fig. 1. Neoplastic cells in the lumen and in the spongy thickened intima of an arachnoidal artery. H.-E. × 270

Fig. 2, 3 and 4. Proliferation and desquamation of endothelium cells of arterioles. The detached cells have the appearance of neoplastic cells. H.-E. × 425