Polymorphic Reticulosis (Lethal Midline Granuloma) and Lymphomatoid Granulomatosis: Identical or Distinct Entities?

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Summary. Two cases of polymorphic reticulosis were studied. Both cases had a fatal clinical course, that of the second case being rapid and progressive and ending 6 months after the onset of the disease with little demonstrable effect of steroid therapy. Biopsy material was obtained in both patients, and both were submitted to a post-mortem examination. The first case showed typical angiocentric, angiodestructive, polymorphous lymphoreticular infiltrates, involving the pharyngeal region and the tongue. The second case demonstrated these same lesions in the midfacial region, the lungs and the skin. A possible identity between polymorphic reticulosis and lymphomatoid granulomatosis is discussed (because of the coexistence of identical lesions in the midfacial region and in the lung parenchyma in the second case). Wegener's granulomatosis in limited and disseminated forms and malignant lymphoma are considered in the differential diagnosis.

Key words: Midfacial – Lung – Lethal granuloma – Lymphomatoid granulomatosis – Reticulosis.

Introduction

Lethal midline granuloma is a clinical term used to label midfacial, infiltrative and destructive granulomatous lesions with a malignant evolution. It includes three different entities, i.e., Wegener's granulomatosis, polymorphic reticulosis and malignant lymphoma of the midfacial region (Kassel et al. 1969; DeRemee et al. 1978).

Liebow et al. (1972) first described lymphomatoid granulomatosis of the lung (LYG). This newly introduced clinicopathological entity was then included among the group of conditions known as pulmonary angiitis and granulomatosis (Liebow 1973). The disease is characterized by angiocentric, angiodestructive,
lymphoreticular proliferative and granulomatous lesions predominantly involving the lungs, commonly sparing lymph nodes and the bone marrow, but often with associated lesions in the skin, central nervous system, the kidneys, adrenal glands and sometimes the digestive tract. Cases have been reported of involvement of extrapulmonary tissues in the absence of lung disease (Chen 1977; DeRemee et al. 1978).

Striking histopathological similarities between polymorphic reticulosis and lymphomatoid granulomatosis have led Douglas et al. (1976), DeRemee et al. (1978), Crissman (1979) and others to the conclusion that they are the same disease process.

Two cases are presented in this report, one with lesions involving the midfacial region only, and the other having an association of identical lesions in the midfacial region with lung and skin lesions.

Case Reports

Case No. 1

Clinical Findings

A 73 year-old woman was admitted to the neurology ward at the University of Geneva Cantonal Hospital because of pain along the course of the right trigeminal ophthalmic nerve branch and right hypoacusia. She had no significant medical history, but complained of palpitations and dyspnoea for several years. Five months prior to admission, she developed chronic hiccups, dysphagia, anorexia and weight loss. Physical examination revealed an elderly woman in good general condition. The temperature was 37°C, the pulse 80, blood pressure 140/80 and respiratory rate 18. Cardiovascular, pulmonary and abdominal examinations were negative. Neurologic examination revealed compression of the Vth, VIth, VIIth, VIIIth, IXth, Xth, XIth and XIIth right cranial nerves by a tumour infiltrating the entire right base of the skull. Skull X-rays disclosed proliferative and sclerosing infiltrates involving the petrous part of the temporal bone, the greater wing of the sphenoidal bone, the foramen ovale and the foramen spinosum on the right (Fig. 1).

A right temporal trepanation was performed and an exploration disclosed no tumour mass. The epipharynx disclosed necrotizing ulcerated lesions, and a biopsy within this region showed polymorphocellular, infiltrative lesions, and the diagnosis of polymorphic reticulosis was made. Radiation therapy was administered at a dose of 4,000 rads of telecobalt. The treatment was well tolerated and, as a result, considerable regression of the neurologic symptoms was observed, with improvement of deglutition, the palatine and pharyngeal reflexes becoming almost symmetrical.

The patient was discharged and was admitted 7 months later for pyrosis, dysphagia, vocal changes, weight loss and hypotension. Physical examination revealed a woman in poor general condition with facial telangiectasias and right conjunctival hyperaemia. Cardiovascular examination disclosed compensated congestive heart failure. Pulmonary and abdominal examinations are negative. Laryngoscopy showed extension of the pharyngeal lesions which were necrotizing, ulcerated and infected. The C.T. scan of the brain demonstrated an extension of the lesions seen previously, toward the right. The patient developed bronchopneumonia and died 5 months later, despite a second course of radiation therapy and antibiotics.

Pathology

Biopsies. The two initial fragments from the epipharynx were embedded in paraffin-wax and stained by H & E. They revealed a squamous mucosa with an ulcerated epithelium beneath which there was a nodular, granulomatous lesion. This lesion was composed of a mixture of lymphocytes, plasma cells,