Epidural involvement in Hodgkin’s disease

ÁRPÁD ILLÉS 1,*, ZSÓFIA MILTÉNYI 1, LÁSZLÓ MILTÉNYI 2, GYÖRGY CSÉCSEI 3 and GYULA SZEGEDI 1
1 3rd Department of Medicine, University of Debrecen, Medical and Health Science Center, Móricz Zs. krt. 22, H-4004 Debrecen, Hungary
2 Department of Radiology, University of Debrecen, Medical and Health Science Center, Nagyerdei krt. 98, H-4012 Debrecen, Hungary
3 Department of Neurosurgery, University of Debrecen, Medical and Health Science Center, Nagyerdei krt. 98, H-4012 Debrecen, Hungary

Abstract—Epidural involvement is analyzed retrospectively in 512 patients with primary treatment and follow up for Hodgkin’s disease (HD) between 1970 and 1999. In one case (0.2%) epidural manifestation was the first symptom and in six cases (1.2%) it occurred later, at a disseminated, advanced stage. All seven patients were male: three had mixed cellularity and four nodular sclerosis histological subtype. The thoracic segment was involved in four cases, the lumbar in two and the cervical segment in one case. The most frequent symptoms were back pain, limb weakness, paresis/plegia, incontinence. Computer tomography, magnetic resonance imaging and myelography were used as diagnostic procedures and in planning the treatment. Functional recovery was achieved by laminectomy, loco-regional irradiation and adjuvant polychemotherapy with remission of HD for 6-100 months. Later, however, six patients died due mainly to relapse/progression of HD. We emphasize the importance of an interdisciplinary approach in the treatment of HD with this relatively rare appearance, which requires close co-operation among oncohematologists, neurologists, radiologists, neurosurgeons, radiotherapists and physiotherapists.

Key words: Epidural involvement; Hodgkin’s disease; spinal cord compression; therapy.

INTRODUCTION

Epidural localization with extranodal involvement is relatively rare in malignant lymphomas: its rate of occurrence is 0.8–6% in all cases of lymphomas [1–4]. This form of appearance is three times more frequent in non-Hodgkin lymphomas (NHL) than in Hodgkin’s disease (HD) [3, 5, 6]: its incidence is 0.1–7.6% in the literature depending on whether it was diagnosed clinically or at autopsy, the latter being much higher [3, 5, 6]. It is usually diagnosed in the disseminated,
advanced stage of HD, after repeated treatment. Its primary appearance is extremely rare. Review articles on epidural involvement in HD including a great number of patients are scarce [6, 7], and due to this rare occurrence, case reports dominate the literature [1, 3–5, 8–11]. We present here an analysis on the frequency of epidural involvement, diagnostic possibilities, the forms and efficacy of therapy of our HD patients as compared to literary data.

PATIENTS AND METHODS

Based on clinicopathological, diagnostic and therapeutic findings, epidural involvement was analyzed retrospectively in 512 patients treated primarily for HD at our institution between 1970 and 1999. We speak of epidural involvement when HD includes the epidural region of the spinal cord and brain, which, as differentiated from subdural and intra-medullary involvement, through its growth and development, may later result in spinal cord compression. For the clinical staging of HD, the Ann Arbor nomenclature and Cotswolds revision were employed [12, 13]. Histological pictures were determined according to Lukes’ classification [14]. The exact location of the epidural tumor was determined by different imaging techniques — myelography, magnetic resonance imaging (MRI) or computer tomography (CT). Polychemotherapy (ChT) consisted of cyclophosphamide, vinblastine, procarbazine, prednisone (CVPP), or adriablastine, bleomycin, vinblastine, dacarbazine (ABVD), or dexamethasone, cytarabine, cisplatin (DHAP) and was administered as described in the literature [15–17]. Radiotherapy (RT) was delivered over 3–6 weeks using a cobalt-60 source with a dose of between 30 and 50 Gy for treated areas (2 Gy/day fraction). Therapeutic responses were assessed according to WHO recommendations [18].

RESULTS

Out of the 512 patients, epidural involvement was found as primary manifestation in 1 patient (0.2%), at late dissemination of HD in 6 patients (1.2%), and all the patients were male (Table 1). Out of the six previously treated patients, only one had local RT; in the rest of the cases, ChT or combined modality treatment (CMT) were given as systemic primary treatment. The mean age of the patients at epidural manifestation was 36.85 years (20–59 years), and an average of 4.1 years (0–9.8 years) elapsed from the diagnosis of HD. The characteristic symptoms were lower back pain, limb paresis/plegia, incontinence. In a symptom-free patient epidural involvement was detected during a control CT examination. The average time of diagnosing epidural localization after the initial symptoms was four months (range: 5 days to 17 months). The HD patient with primary manifestation was under rheumatologic treatment for lumboischialgia for 17 months, and besides hypersedimentation no other alterations were found. Epidural involvement was