Glioblastoma of the brainstem associated with Hodgkin lymphoma

Ederli A., Lo Russo F., Vesentini G.
Divisione Neurologica, Ospedale S. Filippo Neri, Roma

This case of Hodgkin lymphoma associated with glioblastoma multiforme is, as far as we know, unique. The possible implications of this association and of HL associated in 3 known cases with meningioma are discussed.

Key-Words: Hodgkin lymphoma — Glioblastoma multiforme

Case report

This 50 year old woman was admitted to a neurological ward in February 1973 because of headache, complex oculomotor paralysis, truncal ataxia and a psycho-organic syndrome. Nothing noteworthy in the personal or family history. A few weeks before admission she had complained of febricula, loss of appetite and marked weight loss followed by the ataxia for which she was referred to us. No general physical signs apart from very poor nutrition and febricula (37-38 °C).

Neurological examination: head bent and rotated to the left; neck somewhat stiff but no pain; paresis of upward and rightward gaze with nystagmus on attempted rightward gaze; eye and head-turning reflexes spared. Pupils equal and normoreactive; fundus oculi within normal limits. The right arm tended to fall when stretched forward and the legs fell rapidly when stretched forward; weak left foot dorsiflexion. Deep reflexes brisk, especially left biceps and left knee; left Babinski. Marked hypokinesia on initiating movement or speech; sluggishness and poverty of intentional movements. Mentally the patient was torpid with extensive memory gaps (the history was taken from relatives).

Laboratory tests: blood glucose, BUN, SGOT and electrolytes within normal limits. WBC: 11 700 with relative lymphopenia (18%); other blood counts normal. ESR increased: KI 37.5 CSF: proteins 0.10 g/1000 ml; cells 1/mm³; immunodiffusion: gamma-G 4.3 mg/100 ml (N.V. 1.76 mg/100 ml± 0.412). Chest X-ray: broadening of the superior mediastinum towards the right, which on tomography proved to be due to walnut-sized lymph nodes in the paratracheal region, the lowest of which pressed slightly on the superior lobar bronchus; calcifications in the left inferior paramediastinal region with the equatorial plane 8 cm from the diaphragmatic plane. Biopsy of the prescalene lymph nodes revealed a Hodgkin lymphoma.

Neuroradiological investigations were refused by the relatives.

Cyclophosphamide + vincristine therapy was started. Radiological regression of the mediastinal mass was accompanied by deterioration of the neurological status with onset of akinetic mutism. Apparently alert, the patient would obey commands sometimes and sometimes not. After a few months the deep reflexes disappeared and EMG revealed a concomitant polyneuropathy, presumably due to vincristine. Seizures occurred in the preterminal stage followed by deep drowsiness; bilateral papilledema. The patient died on 1/1/74 about a year after onset of symptoms.

In short, ataxic and pyramidal signs with supranuclear oculomotor disturbances followed by intracranial hypertension and preterminal akinetic mutism; Hodgkin lymphoma.
Necropsy: Hodgkin lymphoma confirmed; mediastinal and cervical lymph node involvement; bronchopneumonia with confluent patches in lower lobes; brown atrophy of the myocardium; acute swelling of the spleen; diffuse atrophy of the liver; atrophy of gastric mucosa; pancreas and adrenals intact; turbid swelling of kidneys; nodular goiter. Extensive decubitus ulcers of the sacrococcygeal and iliac regions bilaterally. Brain: hemispheres symmetrical; cerebral edema; gyri diffusely enlarged and flattened; no thickening or granularity of the meninges; bilateral hernia of the uncus; pons transverse diameter increased; vessels of the base moderately and focally thickened. On dissection: marked dilatation of the lateral ventricles and third ventricle; foramina of Monro dilated (diameter 1 cm). Posterior section 1 cm from the mammillary bodies revealed a space-occupying lesion 2×4 cm infiltrating the posterior wall of the third ventricle, the thalamus bilaterally in the region of the pulvinar, the posterior portion of the striatum and of the brainstem as far as the roof of the midbrain. Pons intact. The cerebral aqueduct was infiltrated and completely occluded. Nothing noteworthy in the cerebellum apart from modest hernia of the cerebellar tonsils.

Histology: a) the prescalene lymph node (biopsy) and paratracheal lymph nodes (necropsy) (Figs. 1-2) presented quite similar histological patterns with total disappearance of the lymphatic structure, which was replaced by a great variety of cell types including the diagnostic Reed-Sternberg cells. The similarity of pattern between the biopsy and necropsy specimens rules out a change of cellularity and hence of typing in the course of the disease, definable as of “mixed cellularity”. b) All the other organs examined were broadly normal, especially spleen and liver, which were unaffected. c) Brain: the tumor consisted here and there of fusiform cells but mostly of small, roundish, lymphocytelike cells. The characteristics of the gliomatous growth were: high cell density, cellular polymorphism, considerable structural disorder, presence of atypical mitoses and of giant cells, considerable vascular development with structural abnormalities of individual ves-