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Primary spinal medulloblastomas?

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Abstract
Two cases with spinal medulloblastoma are presented in which even modern neuroradiological methods (computed tomography – CT – and nuclear magnetic resonance imaging – NMR –) did not demonstrate intracranial lesions. These cases should be considered to be primary spinal medulloblastomas, even if until now the existence of real primary spinal medulloblastomas has been doubted.

Keywords: Medulloblastomas, spinal tumours.

1 Introduction

Medulloblastomas are typical tumors of the inferior cerebellar vermis, into which they spread from the roof of the fourth ventricle [12]. There are rare cases in the pons, and, als Zulch points out: "medulloblastomas in the supratentorial space are absolutely exceptional and histological examination requires checking". In the opinion of RUBINSTEIN so-called "primary spinal medulloblastomas" are metastases whose cerebellar origin is too small to diagnose. This statement by a well-known neuropathologist is based on autopsy and older neuroradiological investigation methods such as ventriculography and pneumencephalography.

Two cases are presented in which even modern diagnostical methods (CT and NMR) could detect no cerebellar origin. Therefore, the existence of real primary spinal medulloblastomas should be accepted.

2 Case reports

Case 1. J. D., male 26 years old. This patient developed signs of increased intracranial pressure. CT revealed hydrocephalus internus. Even after contrast enhancement and special investigation of the posterior fossa no space-occupying intracranial lesion could be detected (Figure 1). Results of ventriculography and CSF scintigraphy were also negative, shunting reduced the signs of increased intracranial pressure. Amipaque myelography was quite normal in the cranio-cervical and thoracical region, but in the lumbar region at L 3 and L 4, there were some round contrast deficits (Figure 2). Cytopathology of the cerebro-spinal fluid did not show any pathological cells.
A further CT control, which became necessary because of shunt insufficiency, was also negative except for the reenlarged ventricles.

Three months later the patient developed paraparesis. Control myelography showed a space-occupying lesion from D 1 to D 5 at this time.

At operation a partially extrapartially intramedullary, brown soft tumor was found and partly removed. Histologically a medulloblastoma of the desmoplastic type was diagnosed and therefore a radiation therapy of the neuroaxis and chemotherapy were carried out.

One-and-a-half years later, again before a necessary correction of the shunt, CT revealed, in addition to hydrocephalus, two small, round hyperdense masses with a diameter of 1 cm, one at the roof of the fourth ventricle, the other one between two frontal horns. Another cranial radiation with 3000 rads normalised CT findings.

During the next 7 months respiration disturbance developed. While the cranial CT remained normal, the spinal CT showed a slowly ascending growth of the thoracic tumor. After three years the patient died. Autopsy was refused.

Histological findings (HE coloring, Gomori silver stain): a tumor with isomorphic cells with some cytoplasm and without differentiation. The nuclei are rather large, round to oval with much chromatin and mitoses. Homer-Wright rosettes are not present. Between the varying amounts of tumor cell aggregations lies a silverfiber net of uneven width (Figure 3).

Immunohistochemical findings (PAP): The tumor expresses in some regions neuron-specific enolase and punctional, even acidic, glia fiber protein (Figure 4).

Case 2. B. A., a fifteen year old female patient, was hospitalised under suspicion of polyradiculitis. CSF investigation showed 58/3 cells and a markedly elevated protein

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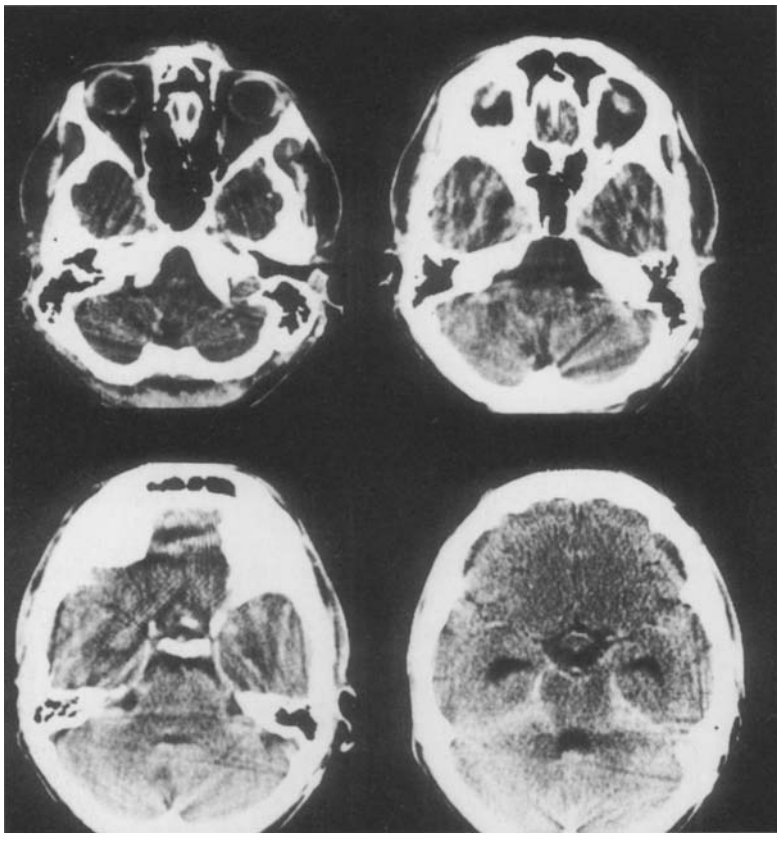

Figure 1. Posterior fossa with contrast medium

level of 4 380 mg/cl. An increase in CSF monocytes was considered to be a sign of inflammation.

Cortison therapy resulted in the disappearance of clinical symptoms. Fourteen days later acutely progressive paraparesis developed. Myelography showed a space-occupying lesion from D 8 to L 2.

Operation revealed a hard grey tumor (Figure 5) which involved the spinal roots and extended intramedullary. The histological diagnosis was medulloblastoma of the desmoplastic type.

CT and NMR (Figure 6) with special regard to the posterior fossa revealed no intracranial tumor. Radiation therapy of the neuroaxis combined with cytostatics (Metotrexat, Vincristin, CCNU and Cis-Platin) did not, unfortunately, influence the tumor effectively enough, and the girl died within 18 months. Unfortunately autopsy was also refused in this case.

Histological findings (HE and Gomori silver staining): Isomorphic tumor with a large number of cells. Cytoplasm is thin, nuclei are round to oval with much chromatin and mitoses. Tumor cell necroses are rare and there are no Home-Wright rosettes. The tumor contains regions in which the cell amount is reduced, these regions are free of agrophile fibers, while the other regions show a net of reticuline fibers.

Immunohistochemical findings (PAP): Some cells express acidic glia fiber proteins. Most of these cells are situated within bright cells. Also, neuron-specific enolase can be found in great cell connections which are also situated in the bright cells.

3 Discussion

Spinal tumors with small cells are difficult to classify. In childhood these tumors may be metastases of medulloblastomas or neuroblastomas or malignant lymphoma, while in elder patients they may be metastases of small-cell bronchogenic carcinoma. Immunhistological investigations are necessary for exact classification. According to Rubinstein medulloblastomas consist of cytogenetically undifferentiated bipotential cells, which are able to differentiate into glial and/or neuronal cells. In both of our tumors immunohistologically proven glia protein and neuron-specific enolase were found. There-

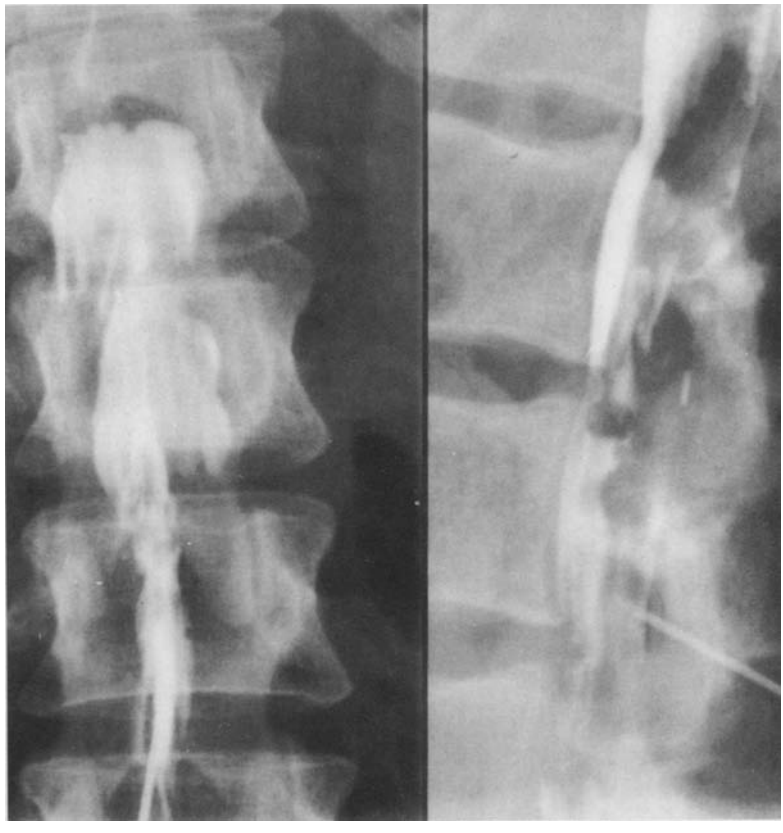

Figure 2. Panmyelography with round deficits in the lumbar region

fore there is no doubt that these tumors were medulloblastomas*.

Our classification of these tumors as primary spinal medulloblastomas could be criticized since autopsy was refused. But in the first case CT controls over nearly three years did not demonstrate a primary intracranial medulloblastoma. The two small lesions between both frontal horns of the lateral ventricles and at the roof of the fourth ventricle, which developed only 1½ years later, were probably metastases. In the second case even NMR demonstrated no intracranial mass.

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In the literature there is no data relevant to the relation between brain tumor growth and metastasis size, but we know from general pathology, especially the pathology of small cell lung cancer, that there are very small primary tumors with expanding metastases. There are no reports of such tumors in the brain because metastases of brain tumors are rare.

Striking is the fact that the medulloblastomas were of the desmoplastic type in both cases. This type is more typical for older children and is more often located in the cerebellar hemispheres.

Because in both cases even CT and NMR investigations detected no primary intracranial tumors, we feel that the classification of these tumors as primary spinal medulloblastomas is justified.

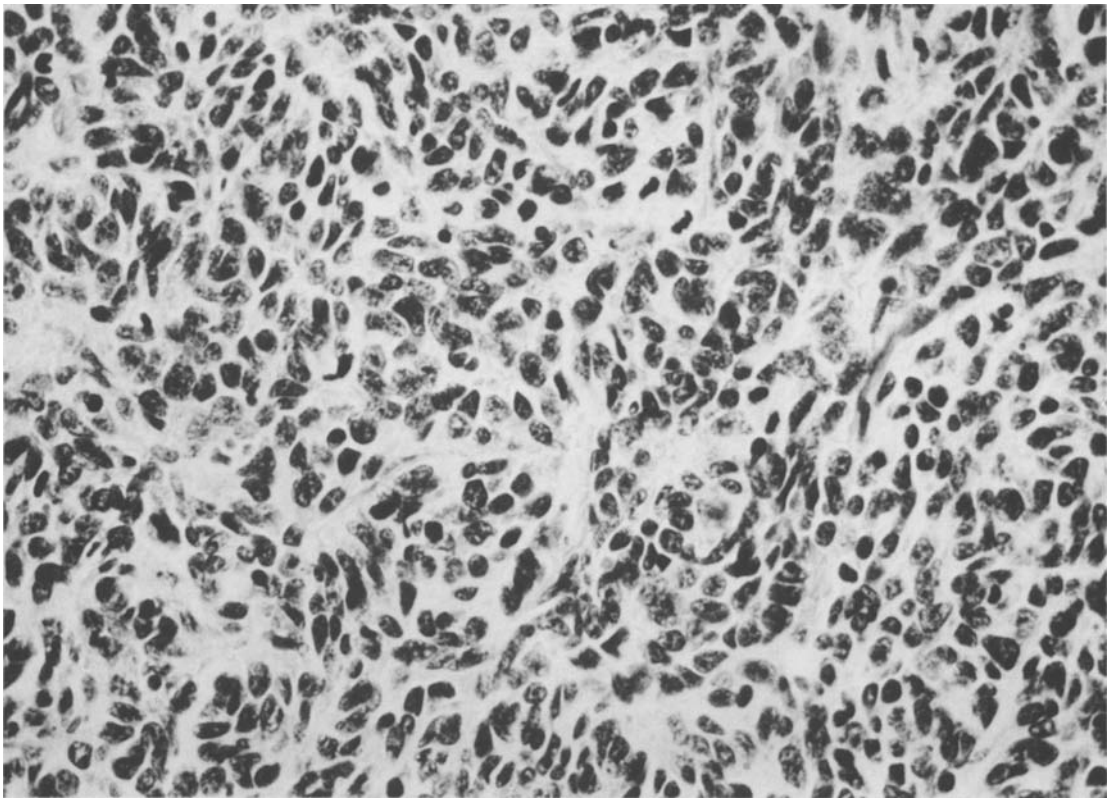

Figure 3. HE coloring of the medulloblastoma

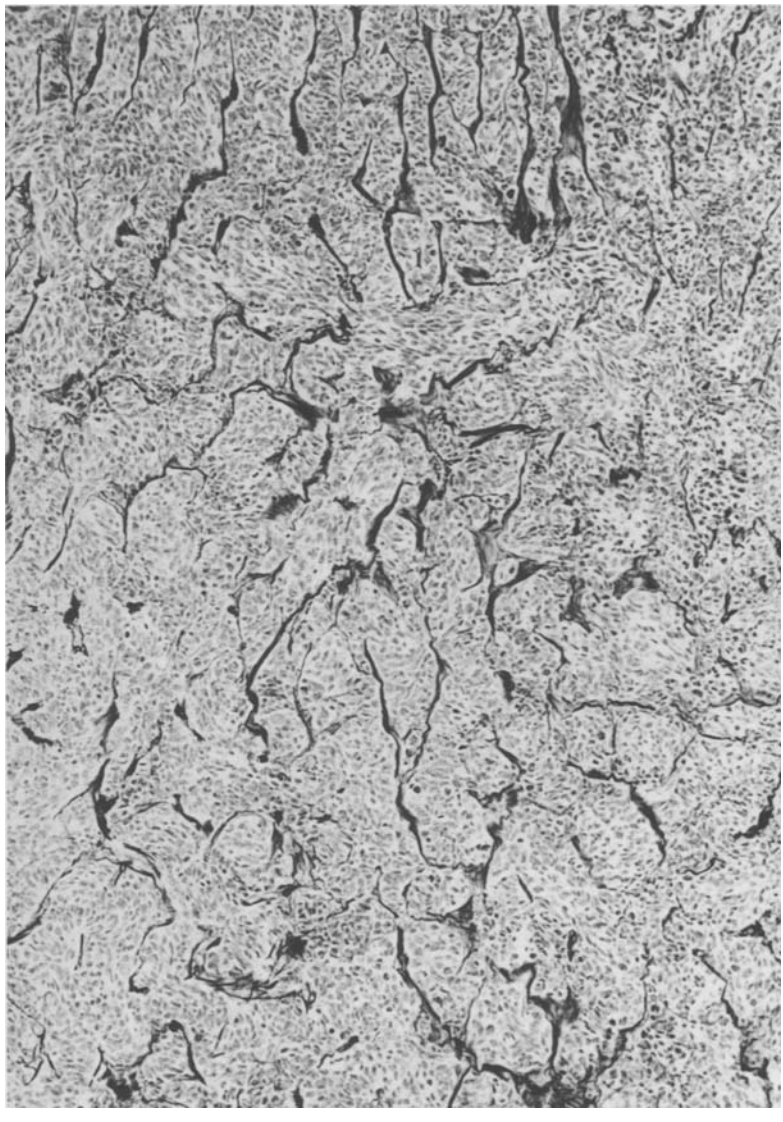

Figure 4. Gomori silver staining

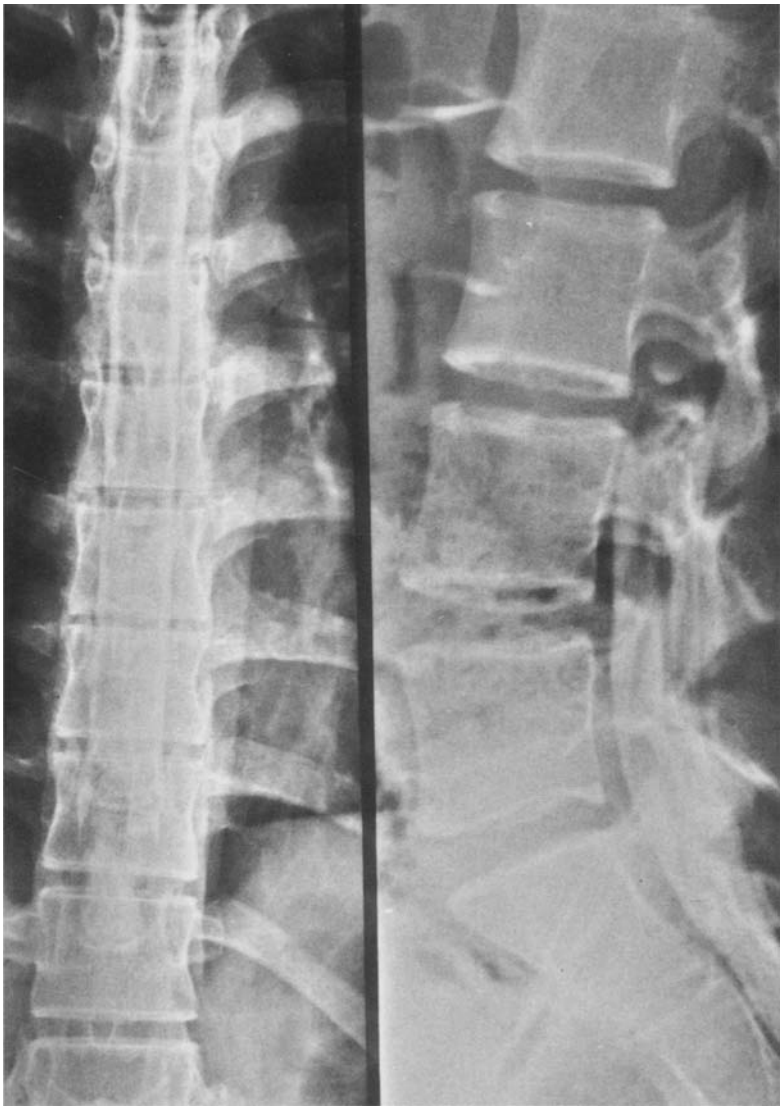

Figure 5. Myelography up to the level conus

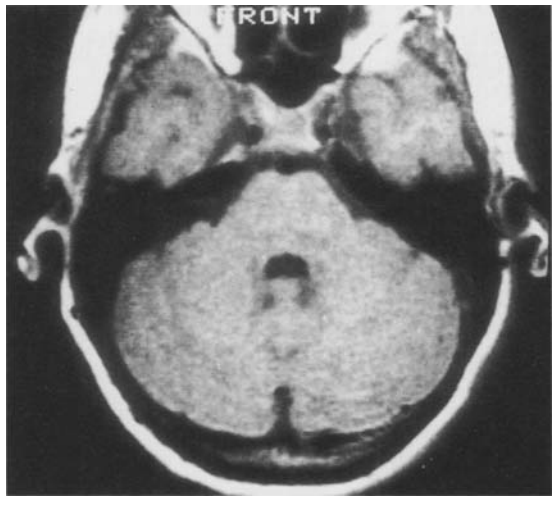

Figure 6. NMR of the posterior fossa

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