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Primary spinal primitive neuroectodermal tumor with extraneural metastases.

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Discussion

Cranial CT changes associated with metabolic and storage disorders have been well documented. However, until recently [1], CT findings in galactosemia had not been shown.

Previous neuropathologic autopsy findings in galactosemia have been described by Crome [2] as a "microencephaly caused by a mainly burnt out gliotic encephalomyelopathy." Hypothesized causes for this encephalopathy include hypoglycemia and competitive inhibition from hyperbilirubinemia or galactose-1-phosphate [3]. The toxic effect of galactose has been shown in animal models; it caused early and permanent injury to fetal nervous tissue and affected both growth and maturation [3]. Another study [4] showed the interference of galactose and galactitol in the metabolism of myelin constituents.

In our patient, the dramatic reversibility of diffuse white-matter low attenuation suggests the presence of an interstitial intracerebral edema that is secondary to the toxic effect of galactose, perhaps because of osmolar effects as suggested by Belman et al. [1]. Another possibility for the intracerebral edema is that, with a dysmyelinating process, the loss of myelin, which is normally hydrophobic, causes an influx of water. The reversibility of edema coincided with initiation of a lactose-free diet and correlated with significant improvement in affected end organs, specifically the liver, spleen, lens, and CNS.

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Primary Spinal Primitive Neuroectodermal Tumor with Extraneural Metastases

Primary spinal primitive neuroectodermal tumor (PNET) is rare [1]. Although extraneural metastases of CNS tumors once were considered rare, a recent review documented 282 cases, one of which was a primary spinal PNET [2]. We describe a case of a 26-year-old man with a primary spinal intradural PNET and subsequent metastases to bone, lymph nodes, pleura, and spinal extradural space.

Case Report

A 26-year-old man presented with a 6-month history of progressive lethargy and dyspnea. Three years before admission, he had presented with symptoms of cervical spinal cord compression, and a myelogram had shown an intradural, extramedullary mass at the C2 level, producing high-grade, incomplete obstruction to the flow of contrast (Fig. 1). Chest radiograph and CT scan of the head were normal. Subsequently, a large intradural extramedullary mass involving the C2–C3 level was excised surgically. The histologic diagnosis was PNET. Total neuraxis radiotherapy was administered. He was then lost to follow-up.

On admission, the patient was cachetic and had diffuse lymphadenopathy. Hematologic examination showed pancytopenia. A chest radiograph on admission showed diffuse sclerotic bone metastases, bilateral pleural effusions, and multiple pleural mass lesions (Fig. 2). The patient refused treatment and died 10 days after admission.

Autopsy showed spinal extradural and systemic metastases that were morphologically identical (by standard histologic techniques, immunoperoxidase studies, and electron microscopy) to the original surgically removed cervical intradural lesion (Fig. 3). Extradural tumor was present from C8 to T6. No residual intradural tumor was found. Extensive parietal pleural and paravertebral metastatic deposits and a large right apical visceral pleural metastasis were present. Widespread bone and lymph node metastases were found. The brain and intracranial leptomeninges were free of tumor. A small focus of extradural tumor was present in the posterior fossa.

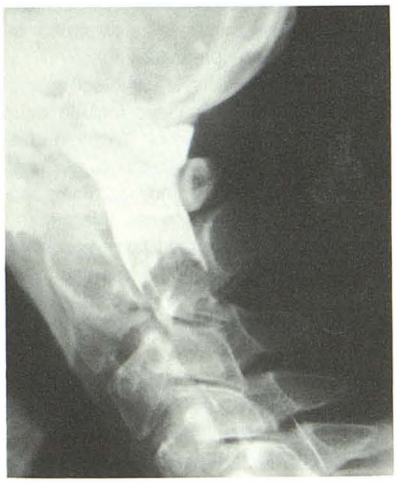

Fig. 1.—Cervical myelogram, lateral view, shows intradural extramedullary mass at C2.

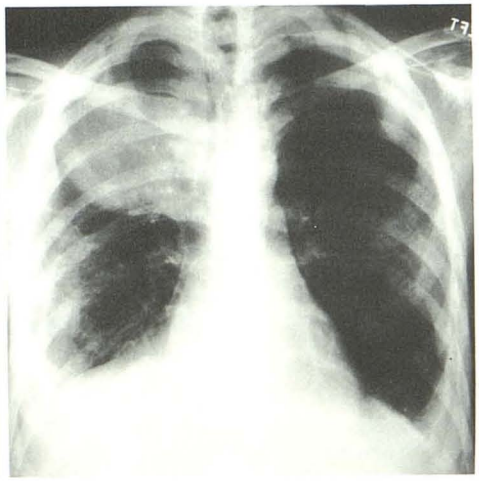

Fig. 2.—Posteroanterior chest radiograph shows sclerotic bone metastases, bilateral pleural effusions, and multiple pleural mass lesions.

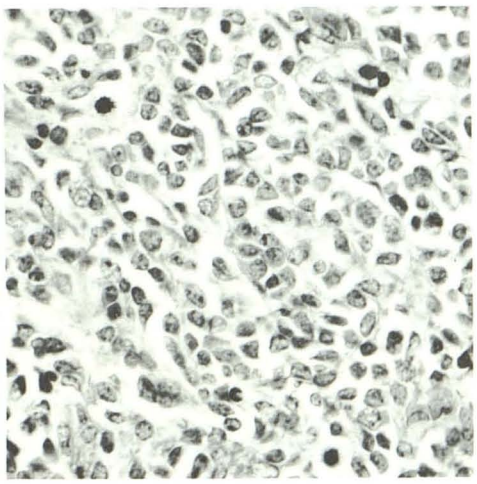

Fig. 3.—Photomicrograph of biopsy specimen from cervical extramedullary lesion shows "small dark cell" tumor. (H and E ×40)

Discussion

Primitive neuroectodermal tumor is a term used for morphologically similar tumors arising in both the central and peripheral nervous systems [3, 4]. PNETs occur preponderantly in children, and spread along cerebrospinal pathways is common. The most common PNET of the CNS occurs in the cerebellum (medulloblastoma) [4]. Extraneural metastases of medulloblastomas are well recognized [5]. Other than those arising in the cerebellum, primary PNETs in the CNS are infrequent and primary spinal intradural PNET is rare [1]. One such tumor with lung metastases has been reported [6]. In that case, metastatic spread was confined to the lungs; whether these were parenchymal metastases was not specified. In children with intracranial PNET, metastases to lung, lymph nodes, and liver have been reported [7].

Given the morphologic similarity of cerebellar PNET (medulloblastoma) and primary spinal PNET, it is not surprising that primary spinal PNET should give rise to similar radiologic metastatic manifestations. Sclerotic bone metastases, paravertebral soft-tissue mass, extrapleural mass, and pleural effusion have been reported with cerebellar PNET [8–12].

The prognosis for patients with metastatic CNS tumors is poor [2]. Although the tumor described in this report is rare, radiographic findings similar to this case are not unusual, and metastatic CNS tumors therefore should be included in the differential diagnosis.

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Parapharyngeal Angiofibroma

Juvenile angiofibromas are uncommon tumors that usually occur in adolescent males and account for approximately 0.5% of all neoplasms of the head and neck [1]. These tumors virtually always arise from the nasopharynx in the region of the sphenopalatine foramen and pterygopalatine fossa [1–6]. This report presents a unique case of an angiofibroma that involved the parapharyngeal space, without involvement of the sphenopalatine foramen or nasopharynx.

Case Report

A 25-year-old man presented with a parapharyngeal mass of 6-months duration. He had noticed an alteration in the quality of his voice, but he denied having pain, hemorrhage, or respiratory difficulty. Physical examination revealed a submucosal mass protruding into the left side of the oropharynx with displacement of the soft palate.

A CT scan showed an intensely contrast-enhancing mass in the left parapharyngeal space, with remodeling and anterior displacement of the left pterygoid plates (Figs. 1A and 1B). The pterygomaxillary fossa was normal. Angiography showed that the mass was hypervascular, with the primary blood supply from the internal maxillary artery (Fig. 1C).

After selective embolization of the internal maxillary artery, the mass was removed surgically. Pathologic examination revealed moderately large, stellate stromal cells distributed evenly within fibrous tissue and moderate-sized vascular channels, typical of a nasopharyngeal-type angiofibroma (Fig. 1D).

Discussion

Juvenile angiofibromas arise from the superolateral aspect of the nasopharynx near the sphenopalatine foramen. These lesions are usually histologically benign but tend to be locally invasive. Characteristically, expansion of the pterygomaxillary fossa occurs secondary to tumor growth [1–3].

Bryan et al. [2] have divided nasopharyngeal angiofibromas into three types according to which anatomic compartment the tumor occupies. Type 1 lesions extend medially from the sphenopalatine foramen and pterygomaxillary fossa to lie within the nasal cavity; type 2 lesions extend laterally into the infratemporal fossa; and type 3 tumors extend intracranially. Previous reports [1–7] have stressed that essentially all of these tumors have a component within the sphenopalatine foramen.

The present case is unusual because the location of the lesion in the parapharyngeal space was exclusive of involvement of the pterygomaxillary fossa. Also, the pterygoid plates were displaced anteriorly, unlike the usual posterior bowing seen with typical nasopharyngeal angiofibromas, which expand the pterygomaxillary fossa.

This case adds another lesion to the differential diagnosis of parapharyngeal masses. The more common masses include deep parotid tumors, schwannomas, glomus tumors, lymph nodes, metastases, and hemangiomas [1]. Although atypical in location, our case showed the characteristic CT and angiographic findings of a nasopharyngeal angiofibroma. In the parapharyngeal space, this appearance would be difficult to distinguish from a glomus tumor, hypervascular metastasis, or even a vascular tumor such as a hemangiopericytoma. However, the occurrence of a radiographically similar lesion in an adolescent male should raise the possibility of a juvenile nasopharyngeal angiofibroma.

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