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AJNR Am J Neuroradiol 1995, 16 (4) 790-792

<http://www.ajnr.org/content/16/4/790>

This information is current as
of June 6, 2025.

Cerebellomedullary Ganglioglioma: CT and MR Findings

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Summary: We present a case of cerebellomedullary ganglioglioma in a young child with indolent clinical symptoms. CT demonstrated a region of hypodensity with central contrast enhancement in the right cerebellar hemisphere and the inferior peduncle. On MR the lesion was isointense on T1-weighted images with fairly homogeneous enhancement with gadolinium, and of high signal intensity on T2-weighted images.

Index terms: Ganglioglioma; Children, neoplasms; Brain stem, neoplasms

Ganglioglioma is an uncommon primary lesion of the central nervous system, representing less than 2% of all intracranial tumors (1). Gangliogliomas of the brain stem are rare (2). They appear more commonly in children and young adults and tend to occur in the medullary region (3). The mixture of glial and differentiated nerve cells distinguishes this neoplasm from other intrinsic brain stem tumors. The favorable prognosis associated with ganglioglioma makes early recognition important for treatment and patient counseling. We present a case of cerebellomedullary ganglioglioma with computed tomography (CT) and magnetic resonance (MR) findings.

Case Report

A 4-year-old white boy presented with a lifelong history of delayed and awkward motor skills, associated with normal cognitive development. His clumsy, stiff gait had erroneously been attributed to Charcot-Marie-Tooth disease, which afflicted his maternal grandfather and great-grandfather. The patient's parents reported appropriate progression in motor development for his age.

His examination revealed a mild, bilateral lower motor neuron facial paresis; a high-pitched nasal voice with vocal tremor (reportedly unchanged); a slight head tilt to the left; diffuse hyperreflexia; a mild right hemiparesis; ataxia in all extremities; and downward plantar responses. Sensory examination was within normal limits.

CT scan of the head showed a region of hypodensity without calcification in the right cerebellar hemisphere and inferior peduncle with central contrast enhancement (Fig 1A and B). MR imaging (650/20/1 [repetition time/echo time/excitations]) revealed a low-signal-intensity lesion within the posterior medulla, upper cervical cord, and cerebellum (Fig 1C). With intravenous infusion of gadolinium (500/20/2), there was fairly homogeneous enhancement of the tumor. Below the region of enhancement, the cervical cord appeared widened and of low signal intensity. On T2-weighted images (2200/90/1), the lesion had a uniform high signal intensity. Exophytic extension of the tumor into the fourth ventricle and foramen of Luschka (Fig 1D and E) with associated lateral and third ventricular enlargement was observed.

A ventriculoperitoneal shunt was placed for hydrocephalus. Five days later, a posterior fossa craniotomy with removal of the lamina of C-1 and C-2 was performed. At surgery, a bulbous medulla oblongata with enlargement of the right inferior cerebellar peduncle and exophytic extension of tumor into the fourth ventricle was encountered. Biopsy of the medullary lesion and excision of the exophytic mass within the fourth ventricle was performed. However, total resection of the tumor was not possible.

Tumor biopsy demonstrated predominantly a low-grade astrocytoma with haphazard clusters of ganglion cells showing occasional bizarre and binucleate forms (Fig 1F). These findings were consistent with the histologic diagnosis of ganglioglioma.

Discussion

Ganglioglioma is a rare primary neoplasm with an incidence of 0.4% to 1.7% of all central nervous system tumors (3, 4). It is more commonly diagnosed in children and young adults; both sexes are equally affected (3, 5). The common sites of presentation, in decreasing frequency, are cerebral hemispheres, particularly the temporal lobes; region of the third ventricle; cerebellum; brain stem; and spinal cord (6). In children, the tumor has a proportionally greater

Received February 15, 1991; accepted after revision June 4, 1993.

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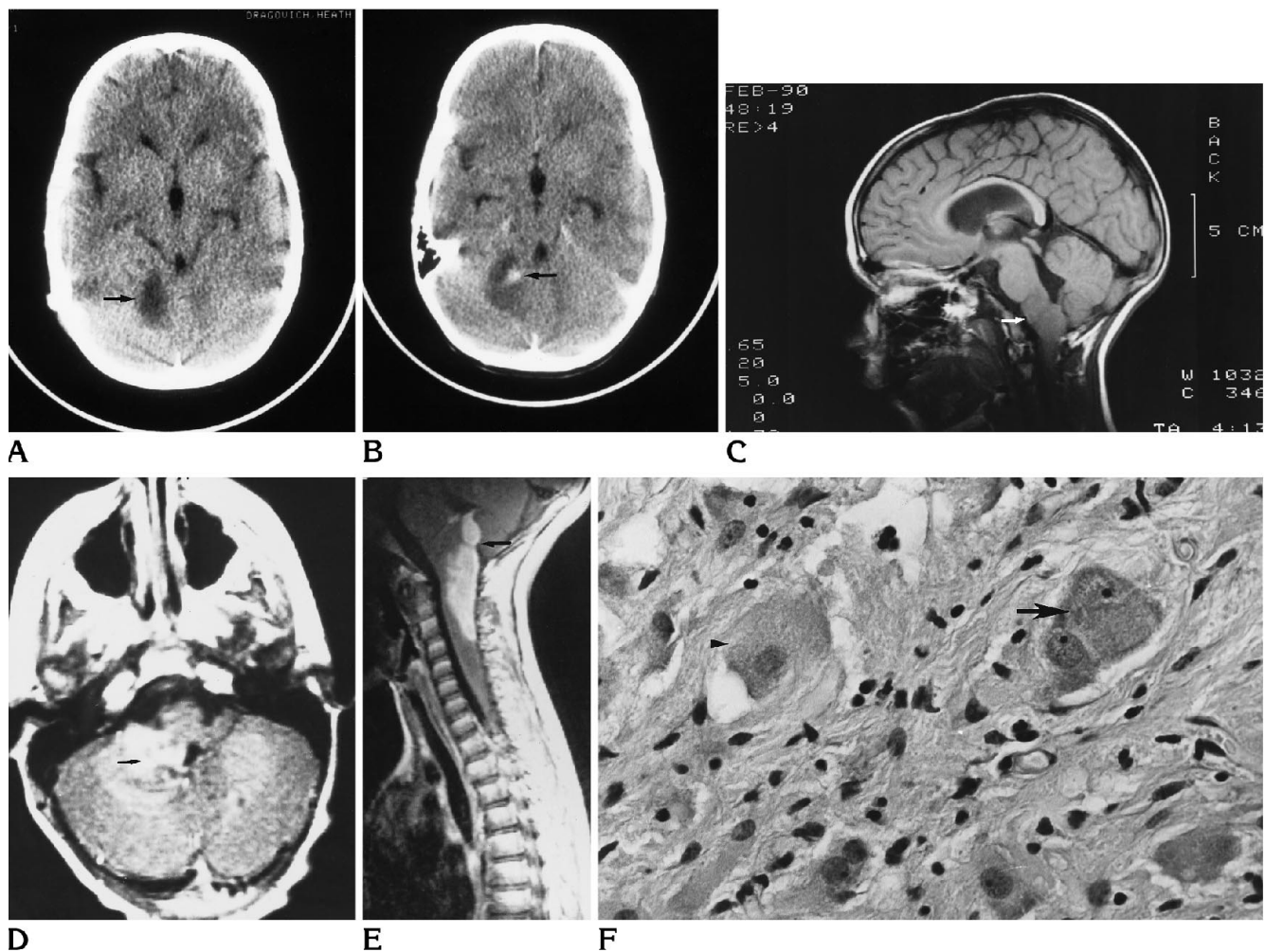


Fig 1. A, Precontrast CT scan demonstrates a region of hypodensity (arrow) in the right cerebellum.

B, Postcontrast scan shows an area of central enhancement (arrow).

C, MR imaging (650/20/1) with body coil shows hypointensity (arrow) in the medullary region and cerebellum.

D and E, Gadolinium-enhanced axial and sagittal images (500/20/2) demonstrate exophytic tumor extending into the fourth ventricle and out the foramen of Luschka (arrow).

F, Ganglioglioma: representative field of tumor depicting low-grade astrocytoma with clusters of ganglion cells (arrowhead) and binucleate ganglion cells (arrow). Hematoxylin-eosin stain, light microscopy $\times 100$.

incidence in the region of the fourth ventricle, medulla oblongata, and spinal cord (3). Ganglioglioma was named for its histologic components of differentiated nerve cells, in the form of ganglion cells, with a glial background that is typically astrocytic (7).

Ganglioglioma is thought to be a tumor of low malignancy potential with a benign clinical course (2, 8). Whereas these neoplasms are generally not known for accelerated growth, Johansson et al suggest a less promising outlook for tumors affecting the midline (5). However, most reported cases of brain stem ganglio-

glioma have a more indolent course than other intrinsic brain stem tumors (9).

CT and MR characteristics of supratentorial and cerebellar ganglioglioma were reviewed recently (10, 11). The tumors proved to have a 38% to 44% incidence of a cystic component with the majority being solid tumors. On CT, the tumors showed varying degrees of density with most having a region of low density or isodensity compared with normal brain stem. Most tumors showed areas of contrast enhancement; calcification was seen only occasionally. With MR, T1-weighted images showed a spectrum of

signal ranging from hypointense to hyperintense, relative to surrounding neural tissue. T2-weighted images showed high signal intensity in all but 1 of the 14 lesions. Gadolinium was given to one patient and, unlike our case, the tumor failed to enhance.

We found the lesion to be hypodense on CT with central contrast enhancement. No tumor calcifications were identified. With MR, the medulla oblongata and upper cervical cord were expanded and the tumor was isointense on T1-weighted images and uniformly enhanced with gadolinium. On T2-weighted images, the lesion had a high signal intensity relative to surrounding parenchyma. The tumor appeared to be infiltrating the medulla and upper cervical cord with an exophytic extension into the fourth ventricle and through the foramen of Luschka. The implied invasiveness of this lesion on radiographic studies does not necessarily correlate with the patient's clinical course. Involvement of the medulla and an exophytic extension have both been related to a more benign clinical course (2).

Histologically, gangliogliomas are mixed tumors composed of relatively mature neoplastic glial and ganglion cells in varying proportions. Both components usually exhibit low grades of malignancy, but it is the grade of the gliomatous element that predicts the biological behavior of the tumor. Histologic diagnosis may be complicated due to difficulty in determining whether the ganglion cells are truly neoplastic, or simply "trapped" neurons in an infiltrating glioma. Russell and Rubinstein (12) noted three features of neurons that aid in their identification as neoplastic: (a) heterotopic location of neurons; (b) distribution and spacing of neuron clusters, some with transitional neuronal forms; and (c) bizarre forms and binucleation.

Although the diagnosis of cerebellomedullary ganglioglioma is rarely made before surgery,

MR imaging makes visible lesions within the posterior fossa, brain stem, and upper cervical cord. A more precise histologic differential diagnosis may be possible. A lesion with a low intensity on T1-weighted images and high intensity on T2-weighted images involving the medullary region, particularly in a child with an indolent clinical course, should place ganglioglioma within the differential diagnoses. The distinct nature of this lesion makes early histologic diagnosis important for treatment and family counseling.

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