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Cerebellar ganglioglioma in a young woman

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Abstract

We describe a rare case of a cerebellar ganglioglioma in a 31-year-old woman discovered incidentally during a CT scan of the parotid glands. CT and MRI features are not specific, with mixed cystic and solid components with calcifications. Although rare, our case demonstrates that cerebellar ganglioglioma must be considered in the differential diagnosis of infratentorial tumors in young adult patients.

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Introduction

Ganglioglioma is a very uncommon primary neoplasm of the central nervous system, most commonly found in the supratentorial compartment. Occurrence in the posterior fossa is extremely rare. Moreover most reported cases concern children. No previous reports have compared the imaging semiology of pediatric and adult infratentorial gangliogliomas [1]. Here, we report an adult patient with cerebellar ganglioglioma.

Case report

A 31-year-old woman presented with intermittent left parotid pain. The neurological examination was normal.

A CT-scan showed normal parotid glands, but incidentally revealed a 3.3 cm cystic mass in the left cerebellum with a 1.8 cm calcified solid nodule with minimal mass effect (Figure 1). After IV injection of contrast medium there was some slight enhancement. Brain magnetic resonance (MR) confirmed the tumor with a large cystic component, some intralesional septa and an anterior and inferior solid component with calcifications (Figure 2). The solid component and the septa showed slight and inhomogeneous enhancement.

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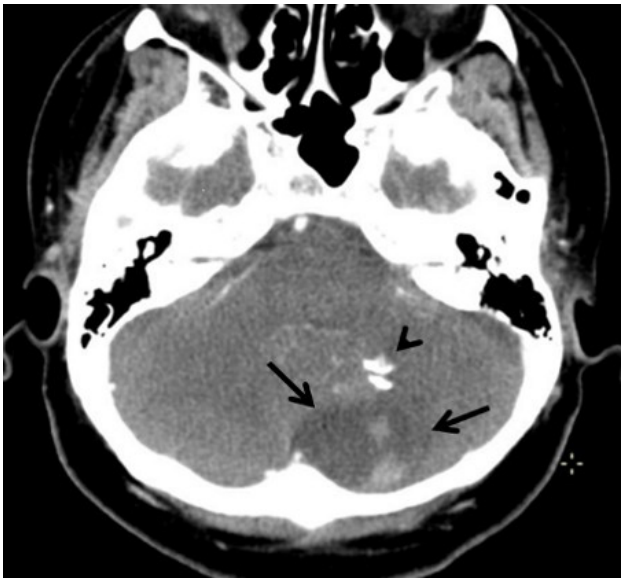

Figure 1: The computed tomography showed a heterogeneous cystic mass (arrows) in the left cerebellum with a calcified solid nodule (head arrow).

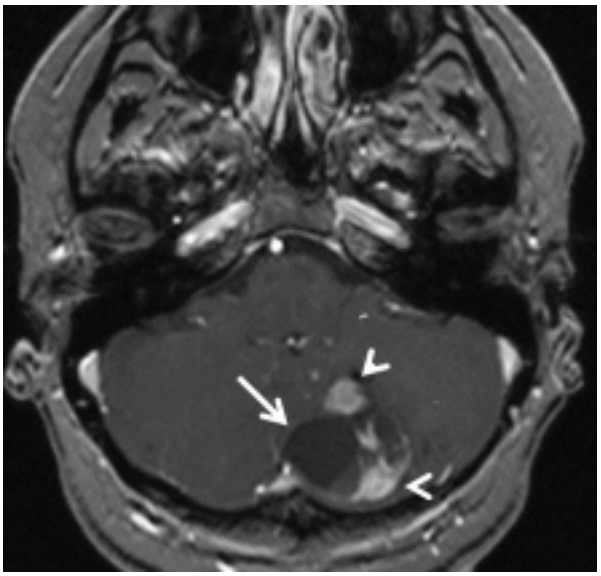

Figure 2: Enhanced T1 magnetic resonance image showing a heterogeneous cystic masse (arrow) in the left cerebellum with an enhanced solid nodule in its anterior and posterior parts (head arrows).

Discussion

Ganglioglioma (GG) is a rare primary tumor of the central nervous system, representing 0.5–1.7 % of total neuroepithelial tumors. They are composed of both neuronal (ganglion cells) and glial elements [2]. This tumor is usually seen in children and young adults with a peak age of incidence between 10 and 20 years and without gender preponderance [3]. They are often located in the cerebral hemispheres with the temporal lobe as the most common location. Other less common locations include frontal lobe, occipital lobe, thalamus, optic chiasm, pineal region ventricles, cerebellum, brainstem, and spinal cord [4]. In fact, only 15% of GGs are located in the posterior fossa (PF) [3].

Our case is unusual in that it was located in the left cerebellar hemisphere. Only 80 cases were reported in the literature: the majority of the lesions also involve the cerebellar peduncles; pure cerebellar GGs are rare [3].

A cerebellar GG can be an incidental finding as our patient. When symptomatic, the patient presents with slowly progressive cerebellar signs possibly associated with gait disturbance [1,3]. While the exact etiology and pathogenesis remains unclear, a few authors suggested that the subgranular zone of the dentate gyrus has a potential to produce the granular neurons postnatally. Those neurons have a greater susceptibility to a neuronal-glial neoplastic transformation resulting in a GG [3].

Radiologically, CT and MRI features of cerebellar GGs are not specific for supra or infra-tentorial GGs but almost all tumors appear as a cyst with an enhancing mural nodule variable mass effect and little or no surrounding edema [4]. The tumor is often isodense or hypo dense in the CT-scan, with calcification present in 6% to 30% of cases, as in our case [5]. On MRI imaging, the GGs are usually hyper intense on T2-weighted images and isointense to slightly hypo intense on T1-weighted images. The variable signal in the cystic portion depends on whether the contents are proteinaceous, hemorrhagic, or contain cerebrospinal fluid. Gadolinium enhancement of the solid components of the GGs is observed in about half of the cases, although the pattern varies from intensely homogenous to heterogeneous [5].

Although CT or MR imaging may suggest the diagnosis, the differential diagnosis of cerebellar GGs in children or young adults must include mainly pilocytic astrocytomas (PA), but also ependymomas, desmoplastic medulloblastomas, and hemangioblastomas. Each of these tumors may demonstrate cyst formation as well as a solid portion [5].

The PA is the most common primary cerebellar tumor in children and young adult, and cannot be distinguishable from cerebellar GG radiologically. The characteristic “cyst with mural nodule” appearance of PA in posterior fossa and somewhat heterogenous enhancement including wall of cyst, making us strongly suspect a PA preoperatively as in our case. Hemangioblastoma and metastasis can be excluded by the patient age and its association with systemic manifestation [5]. Medulloblastoma is characterized by an uniform hyperdensity on CT scans, midline location, and filling and expanding of the 4th ventricle. On the contrary, cerebellar GGs arise from hemisphere and compress the 4th ventricle. Ependymoma extends out the 4th ventricle foramina, and are usually heterogenous with calcification and hemorrhage, but those are not typical for GGs on CT and MR images.

Radical surgical resection is the mainstay of GGs treatment. Infratentorial gangliogliomas are considered to be more challenging than their supratentorial counterparts, except when they are confined to the cerebellum as in our case [3]. With regard to recurrent or progressive lesions and tumors with anaplastic features, radiation therapy has been recommended, but it remains uncertain whether or not this improves outcome [5]. Some reports suggest that radiatherapy may predispose to malignant transformation of GG and the role of chemotherapy is still debated [3]. Complete resection leads to a good long-term prognosis which results in a greater than 90 % of 5 year survival. Malignant transformation is unusual and when it occurs, most often involves gliobastoma (glial cells) and less frequently neuroblastoma (ganglion cells) [4].

In a recent article, immunohistochemical molecular analyses indicated that BRAF V600E mutation is present in 38-54 % of gangliogliomas without prognostic significance [3]. Anti-BRAF inhibitors can be used for targeted therapy in case of tumor regrowth. The status for BRAF-V600E mutation should be tested

for all infratentorial GG. A few cases of gangliogliomas treated by anti-BRAF therapy with promising results have been reported [3].

Conclusion

We present a rare case of a cerebellar ganglioglioma in an adult patient along with its radiological features and treatment. There is only a few cases observed and this case can contribute to our understanding of the characteristics of this rare posterior fossa tumor. Our case demonstrates that cerebellar ganglioglioma should be considered in the differential diagnosis of infratentorial tumors in young adult patients because the prognosis is better than other tumors in this location. The status for BRAF-V600E mutation should be tested for all infratentorial GG.

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