

## Desmoplastic infantile ganglioglioma: case report

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Desmoplastic infantile gangliogliomas are very rarely encountered, large supratentorial masses, derived from neuroepithelial origin, which have cystic and solid components and contain cells with astrocytic and ganglionic differentiation. These tumors are benign tumors of childhood that become symptomatic when they reach giant sizes. Sixty cases of desmoplastic ganglioglioma have been reported to date. In the present study, a case of giant desmoplastic infantile ganglioglioma in a 22-month-old patient is presented, which had an aggressive radiological appearance in the midline and presented with atypical symptoms.

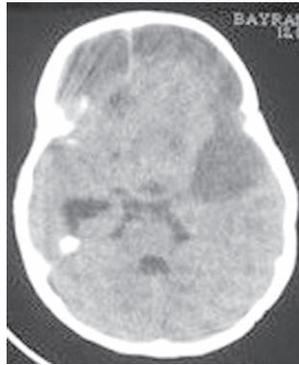
**Key words:** desmoplastic infantile ganglioglioma, magnetic resonance imaging, pediatric patient.

Desmoplastic infantile gangliogliomas (DIGs) are brain tumors with unknown malignant potential, which are rarely seen and usually affect children. They are frequently observed in the frontal and parietal lobes. They are localized in the vicinity of the meninges and involve the cerebral cortex and the leptomeninges. In spite of their aggressive radiological appearances, these tumors have good prognosis. They have cystic and solid components, and the cystic component is the dominant and indispensable component of the tumor<sup>1-4</sup>. When they reach huge sizes, these tumors present with clinical symptoms that include headache, seizures, bulging in the anterior fontanelle, increase in the head circumference and paresis. In the present study, we present a case of giant DIG in a 22-month-old pediatric patient, who was followed-up for the provisional diagnosis of meningitis.

### Case Report

A 22-month-old male patient with nausea, vomiting and fever was hospitalized at the pediatrics clinic with the provisional diagnosis of meningitis. An intracranial mass was detected on the cranial computerized tomography (CT) that

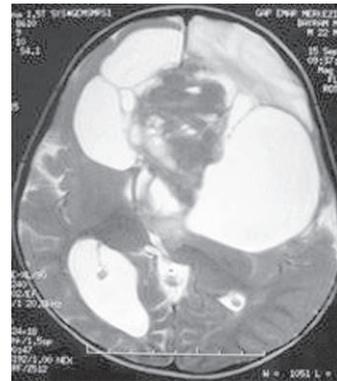
was performed after the patient failed to improve clinically, after which he was referred to the Department of Neurosurgery. On physical examination, he had a mild somnolence and a subfebrile temperature (Glasgow Coma Scale was 13; Motor: 6, Verbal: 4, Eye: 3). The head circumference of the patient was 54 cm. On the brain CT without contrast material, a mass with a solid component was observed in the anterior and middle fossa, with heterogeneous density and accompanying cysts at the periphery, in a hypodense appearance (Fig. 1). The mass was also detected on a cranial magnetic resonance imaging (MRI), which was performed to determine the features of the mass and to plan the pre-operative surgical approach. On T1-weighted MRI (Fig. 2A), a lobulated mass with a heterogeneous hypointense center and a homogeneous prominent hypointense periphery was observed, whereas on T2-weighted MRI (Fig. 2B), the same mass was observed with a heterogeneous, hypointense, solid central portion and a hyperintense, cystic periphery. On axial (Fig. 3A) and sagittal (Fig. 3B) T1AG with contrast material, a mass was observed with irregular borders, extending posteriorly and superiorly to the pineal gland region, obliterating the sellar



**Fig. 1.** On the brain CT without contrast material, a mass with a solid component is observed in the anterior and middle fossa, with heterogeneous density, and accompanying cysts at the periphery, in a hypodense appearance.

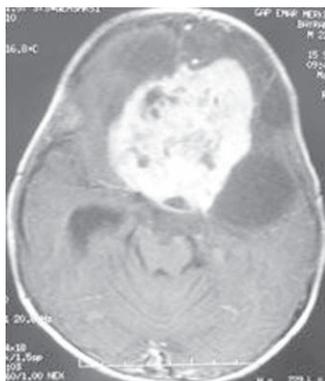


(a)

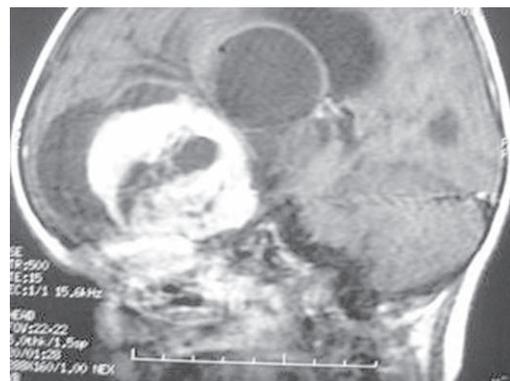


(b)

**Fig. 2A and 2B.** On T1-weighted MRI (2A), a lobulated mass with a heterogeneous hypointense center and a homogeneous prominent hypointense periphery is observed, whereas on T2-weighted MRI (2B), the same mass is observed with a heterogeneous, hypointense, solid central portion and a hyperintense, cystic periphery. The mass obliterates the lateral ventricle on the left. The right lateral ventricle is enlarged due to the pressure of the mass on foramen Monro. In the left frontal lobe, there is enhancement of signals on T2 due to edema and/or gliosis, which is a result of the pressure created by the mass.



(a)



(b)

**Fig. 3A and 3B.** On axial (3A) and sagittal (3B) T1AG with contrast material, a mass is observed with irregular borders, extending posteriorly and superiorly to the pineal gland region, obliterating the sellar and suprasellar structures, and with heterogeneous contrast enhancement, whereas the surrounding cystic components do not show contrast enhancement. There are cystic/necrotic areas in the center of the mass that do not display contrast enhancement.

and suprasellar structures, which displayed heterogeneous contrast enhancement, whereas the surrounding cystic components did not show contrast enhancement.

The mass with solid and cystic components was subtotally excised through an interhemispheric approach following bifrontal craniotomy. During operation, it was observed that the solid component of the mass had tightly adhered to the base of the frontal bone in the anterior aspect, and the mass was firm and hemorrhagic. Because of the texture of the mass, the excision was performed using an ultrasonic aspirator. The histopathological evaluation of the specimen revealed DIG. In the light microscopic evaluation, a lesion composed of pleomorphic spindle astrocytic cells forming short bundles and arranged in a storiform pattern was observed. Large ganglionic cells with abundant eosinophilic cytoplasm and round-oval vesicular nucleus were randomly distributed throughout the lesion. Mitotic activity, microvascular proliferation and necrosis were absent (Figs. 4, 5). The patient was discharged on the 10<sup>th</sup> post-operative day with no additional neurological deficits.

### Discussion

Desmoplastic infantile tumors were first described by Taratuto et al.<sup>2</sup> in 1984. However, DIG was accepted as a distinct clinic pathological entity by Vandenberg et al.<sup>5</sup> in 1987. There have been only 60 reported cases of DIGs in the literature up to 2004<sup>6</sup>.

Desmoplastic infantile tumors are divided into two histopathological groups as desmoplastic infantile astrocytomas (DIA) and DIG<sup>5</sup>. The

difference between these two groups is that DIGs have a neuronal component, whereas DIAs do not<sup>3</sup>. DIG and DIA have been classified by the World Health Organization as low grade, superficially localized desmoplastic neuroepithelial tumors of childhood<sup>7-8</sup>.

Desmoplastic infantile gangliogliomas constitute 0.5-1.0% of all intracranial tumors<sup>6</sup>. They are more commonly observed in male children. DIG is seen in the childhood period and early adulthood. These tumors are generally observed in the first 18 months of life, especially in the first 4 months. When they are detected in the perinatal or natal period, they are huge in size<sup>5</sup>.

Desmoplastic infantile gangliogliomas are large masses with solid and cystic components, most frequently involving the frontal and parietal lobes. Pure solid DIG has only been reported in two cases to date. On the other hand, cases located in the temporal lobe, suprasellar region and the hypothalamus have also been reported<sup>9-10</sup>. More than one lobe is affected in over 60% of cases. In our case, a giant mass with an aggressive radiological appearance and a cystic component that exhibited marked contrast uptake was seen to have extended from the cribriform plate to the sellar and suprasellar regions, hypothalamus and the temporal lobe.

Despite reaching large sizes, these tumors become symptomatic in the late stages. Clinical symptoms include seizures, paresis, bulging in the anterior fontanelle, and an increase in head circumference<sup>11</sup>. Although the mass was

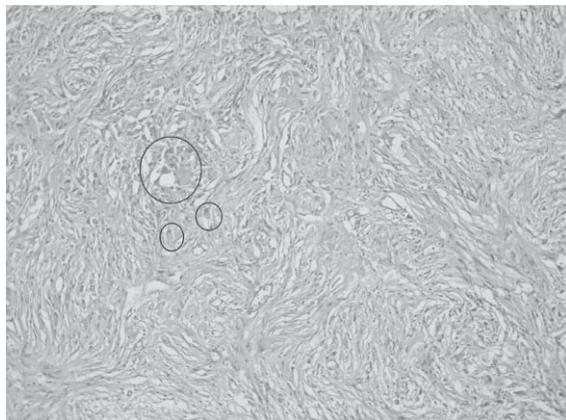


Fig. 4. Ganglionic cells (in circle) and storiform pattern of spindle astrocytic cells (Hematoxylin and eosin [HE], 100X).

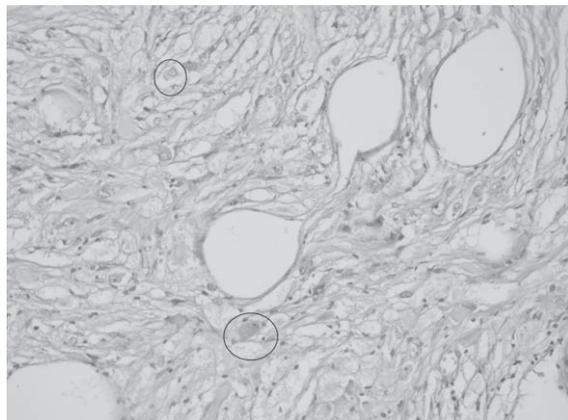


Fig. 5. Cystic degeneration areas, and ganglionic cells (in circle) (HE, 200X).

gigantic in our patient, it had not resulted in distinct neurological deficits. He was provisionally diagnosed with meningitis due to the presence of subfebrile temperature, mild tendency to sleep and vomiting.

The radiological images of DIGs in children and young adults are similar. CT reveals the solid component that is isodense with the cortex or slightly hyperdense. There are usually no calcifications or hemorrhagic foci. The cystic component is hypointense in T1-weighted MRI and hyperintense in T2-weighted MRI. The solid component is isointense both in T1- and T2-weighted MRIs<sup>12</sup>. Atypical appearance has been reported in tumors that are located in the ventral diencephalon, suprasellar region and the hypothalamus<sup>13</sup>. In our case, it was observed that the giant lesion with multiple cysts had initiated from the frontal and temporal lobes, filled the sellar and suprasellar regions, and then extended to the diencephalon.

Histologically, these tumors are low grade benign tumors with glial and ganglionic differentiations. The striking feature of DIGs is the stromal desmoplasia, as well as the variations in neuroepithelial and fibroblastic elements. Four types of neoplastic cells are observed in the cellular areas of the tumor: astrocytic cell type, neuronal cells in different stages, primitive cells, which are counterparts of the neuroepithelial cell type, and Schwann cells. The mitotic activity and the Ki-67 proliferation index of these tumors are said to be low<sup>13,14</sup>.

The differential diagnoses of DIGs include dysembryoplastic neuroepithelial tumor, primitive neuroepithelial tumor (PNET), ependymoma, ganglioglioma, cerebral astrocytoma, malignant teratoma, and sarcomas of childhood<sup>15</sup>. Dysembryoplastic neuroepithelial tumors are supratentorial cortical lesions with a small cystic component and peri-lesional edema and they do not create a mass effect. Primitive neuroectodermal tumors are composed of both cystic and solid components localized in the deep white matter, and may present with calcifications. Ependymomas are localized in the para-ventricular areas along the parietal white matter. Meningocerebral astrocytomas, which are rarely observed, are attached to the dura and are localized superficially. Typical gangliogliomas are usually localized in the temporal lobe and although they are small cystic tumors, they may have a solid component as well<sup>13</sup>.

Desmoplastic infantile gangliogliomas have good prognosis when totally removed<sup>2,5</sup>. Complete surgical resection provides good results even in tumors with atypical histological features. Total excision could be performed in 70% of all the tumors reported to date. Radiotherapy or chemotherapy is recommended in cases in which total excision cannot be performed. In our case, only a subtotal excision could be performed because of the marked adhesion between the part of the mass extending to the diencephalon and the surrounding tissues.

In conclusion, DIGs are low grade supratentorial masses that are seen in the pediatric age group. They have cystic and solid components and have a heterogeneous density radiologically. These very rarely encountered tumors may not be detected until they reach gigantic sizes. DIG should be considered in tumors with a cystic component and an aggressive appearance, which may cause symptoms of increased intracranial pressure in late stages in the pediatric age group.

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