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Pineal gliosarcoma in a 5-year-old girl

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ABSTRACT

The purpose of this paper is to report a rare case of a pediatric pineal gliosarcoma. Gliomas on the pineal region are uncommon, representing 0.4%-1% of all brain tumors. Furthermore, pediatric gliosarcomas are a very rare entity. We present a case of a 5-year-old girl, with a history of headache, vomiting, diplopia, and gait disturbances. A pineal tumor was found with pathology results consistent with a gliosarcoma. A total of 25 cases of pediatric gliosarcomas have been reported, none of them in pineal topography. Only 3 gliosarcomas were found in the pineal region, but these were found in adults. To our knowledge, this is the first pediatric pineal gliosarcoma reported in the literature.

Introduction

The pineal region is limited superiorly by the corpus callosum and choroid plexus of the third ventricle; anteriorly by the third ventricle; inferiorly by the quadrigeminal plate and cerebellum; and laterally by the thalamus and cerebral hemispheres. Pineal tumors represent 0.4%-1% of all cerebral tumors, and germinal cell tumors are the most frequent type of neoplasm in this topography, accounting for 50%-75% of overall cases; gliomas are very rare in this area [1–3].

The gliosarcomas are a sarcomatous variant of glioblastomas, accounting for 1.8%-2.8% of all cases of high-grade glioma, with predominance in the fifth and sixth decades of life. A literature review by Mallick et al. found a total of 25 reported cases of nonpineal pediatric gliosarcoma [4].

We present a case of a 5-year-old girl with a history of headache, vomiting, diplopia, and gait disturbances. A pineal tumor was found with pathology results consistent with a gliosarcoma.

Case report

A 5-year-old girl was transferred to our institution with a history of 2 weeks of severe headache that woke her up during the night, associated with several episodes of vomiting, double vision, left gaze deviation, and gait disturbances. During the physical examination, she was alert, with 15/15 Glasgow scale, mydriatic 4-mm hyporeactive pupils, and with III and IV cranial nerves paralysis found and fundoscopy that revealed papilledema.

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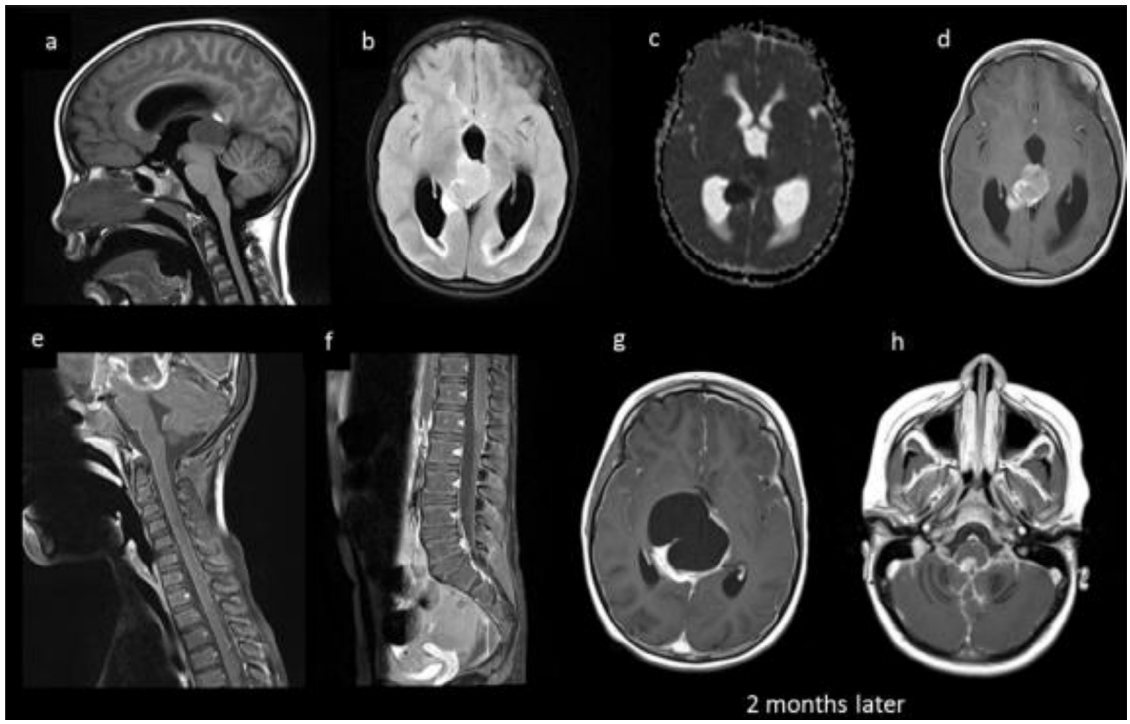

Fig. 1 – Brain MRI sagittal T1 weighted image (A), axial FLAIR image (B), axial DWI (C), axial T1 contrast-enhanced image (D), cervical sagittal T1 contrast-enchanced image (E), lumbar sagittal T1 contrast-enhanced image (F), and brain MRI axial T1 contrast-enhanced image (G and H). Brain MRI revealed a mass type lesion in the topography of pineal gland, hypointense on T1, restricted on DWI, and with heterogenous contrast enhancing. Cervical and lumbar MRI showed diffuse meningeal and filum terminale roots enhancement. Two months later, a brain MRI revealed an increase in the cystic component and a nodular lesion in the medulla. DWI, diffusion-weighted imaging; FLAIR, fluid-attenuated inversion recovery; MRI, magnetic resonance imaging.

A computed tomography (CT) scan showed a mass lesion in the pineal gland region, causing hydrocephalus and mesencephalic compression. Subsequently, a brain magnetic resonance imaging (MRI) demonstrated a mass lesion was found, hypointense on T1, hyperintense on T2 with strongly heterogeneous contrast enhancing. Associated with these findings, ventricular system dilatation and transependymal edema were also observed. A diagnosis of germinal cell tumor, followed by pineal parenchymal or glial cell tumor, was suspected (Fig. 1). An endoscopy biopsy through the third ventricle and external ventricular drain was made. Histopathologic results showed a high-grade glial cell lesion, immunohistochemistry revealed vimentin positive cells, Glial Fibrillary Acidic Protein positive, and KI67 of 90%, consistent with grade IV gliosarcoma.

The patient presented with an episode of acute elevated intracranial pressure with altered mental status and mydriasis with slow pupillary response despite a ventriculostomy. A follow-up CT scan revealed a bleeding mass in the pineal region with ventricular system dissemination and an increase in the residual tumor size. Based on this, an intratumoral mass resection with Sonoca ultrasonic aspirator via frontoparietal craniotomy with an anterior trans-splenic interhemispheric approach and a ventriculoperitoneal shunt was performed.

After this surgical approach, the patient improved her clinical status and an oncological evaluation was performed. Further assessment included a bone gammagraphy and follow-up brain and spine MRI; at this point no metastases were found. An implanted port was inserted to initiate chemotherapy along with adjuvant therapy with temozolomide and radiotherapy.

Two months later, neurologic symptoms such as nystagmus, hemiparesis, gait disturbance, tremor, and myoclonic type seizures persisted. A brain CT scan showed increased tumor size with ventricular system bleeding. A spine MRI was also taken, and meningeal enhancement of filum terminale roots was found, and metastatic dissemination was suspected. All these findings were taken into consideration for the therapeutic plan. Therefore, surgical resection was ruled out. Because of poor therapeutic response and prognosis, the oncology group decided to discontinue chemotherapy with concomitant radiotherapy and to start palliative management of symptoms.

Discussion

Pineal tumors are infrequent and correspond to 0.4%-1% of all cases of intracranial tumors [1,2], a diverse group of tumor types that originates from the pineal gland [3]. Germinal cell tumors are the most frequent type of pineal tumor; gliomas are very rare in this area [3]. During childhood, pineal tumors are more common, corresponding to 3%-11% of all pediatric central nervous system tumors. However, only 25 cases of pediatric nonpineal gliosarcoma had been reported, none of them in pineal topography [4,5].