Disseminated Juvenile Pilomyxoid Astrocytoma of the Hypothalamic-Chiasmatic Region

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Abstract- Pilomyxoid astrocytoma (PMA) is a recently described histological type of pilocytic astrocytoma (PA), but the tumors show histological differences. PMA has more aggressive malignant behavior than PA. Magnetic Resonance Imaging (MRI) may play a crucial role in the preoperative setting and also help to establish an appropriate therapeutic regimen. In this case report, we illustrated MRI findings of a hypothalamic-chiasmatic PMA in a 15-year-old female patient presenting with extensive leptomeningeal seeding. The patient was operated for total tumor resection, but could not survive the second post-operative day. We have comprehensively discussed the clinical, imaging, and histopathological features of these relatively rare tumors and also reviewed the recent literature.

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Keywords: Pilocytic astrocytoma; Pilomyxoid astrocytoma; Leptomeningeal; Dissemination

Introduction

Pilomyxoid astrocytoma (PMA) is a recently described rare astrocytic tumor recognized as a histological variant of pilocytic astrocytoma (PA) (1). These tumors are known to exhibit different clinical and histopathologic features. The most striking characteristics of PMA that help to differentiate them from typical PA are leptomeningeal dissemination and higher rates of recurrence (2). Therefore, enabling a differential diagnosis between these two separate entities plays a crucial role in terms of patient management. More aggressive treatment regimens are needed for PMA due to its more malignant behavior and shorter survival rates (3). Although pathognomonic imaging characteristics have not yet been identified in order to discriminate PMA from PA, magnetic resonance imaging (MRI) may play an important role in the preoperative assessment of these tumors and enables an appropriate therapeutic algorithm. Besides, it can reveal some imaging features typical of these tumors and might suggest a correct preoperative diagnosis without needing histopathological proof. In this case report, we described a hypothalamic-chiasmatic PMA in an adolescent female patient who presented with extensive leptomeningeal mass-like dissemination and has also reviewed the recent literature.

Case Report

A 15-year-old female patient was admitted to our hospital complaining of nausea, vomiting, headache, and pain in the cervical region. She also had decreased visual acuity for one year. She was referred to physical therapy after plain cervical x-ray films being obtained and underwent conservative therapy. Due to the persistence of her complaints, a brain and cervical MRI were ordered. These examinations were performed in our clinic by a 1.5 Tesla MR scanner using a 12-channel phased-array head and cervical coils (Avanto-SQ Engine; Siemens, Erlangen, Germany). During these examinations, axial and sagittal T1 weighted, axial and coronal T2 weighted, axial and coronal FSE IR (flair), axial SWI (susceptibility weighted) and finally following intravenous gadolinium DTPA administration (0.1 mmol/kg) axial and coronal post-contrast T1 weighted images for brain and sagittal and axial T1, T2 and post-contrast sagittal and axial T1 weighted images for the cervical region were obtained. On these images, a 30 x 20 mm diameter solid mass lesion in the hypothalamic region invading the optic chiasm posteriorly and extending into the suprasellar and interpeduncular cisterns was detected. It was hypointense on T1 weighted images (T1WI) and

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hyperintense on T2 weighted images (T2WI) and showed minimal contrast enhancement (Figure 1). Besides, on post-contrast T1WI, intensely enhancing mass lesions were detected in the left cerebellar region consistent with leptomeningeal tumor seeding (Figure 2). On contrast-enhanced cervical spine imaging, we found a huge mass with 80 x 50 mm diameters in the level of the foramen magnum, extending inferiorly through the anterior cervical compartment up to the C 4 vertebra region. It was located outside the cervical spine but severely compressed it posteriorly. It had low signal intensity on T1WI and high signal intensity on T2WI and showed heterogenous contrast enhancement (Figure 3). These findings were re-interpreted as leptomeningeal seeding metastases like those of the left cerebellar ones. The patient underwent a biopsy procedure via a transsphenoidal approach. Histopathological analysis of the biopsy specimen revealed a glial tumor consisting of monomorphic and piloid cells lying in a striking myxoid background. Immunohistochemical staining for glial fibrillary acid protein was found positive. Eosinophilic granular bodies and Rosenthal fibers were absent. Based on these findings, the patient was diagnosed as having PMA. The treatment was planned to remove the tumor by total excision, followed by adjuvant chemotherapy. But unfortunately, after total tumor resection, the patient died on the second post-operative day in the intensive care unit.

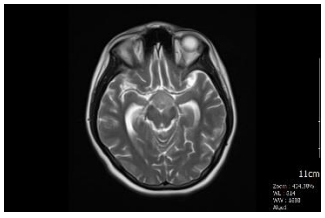

Figure 1. Axial FSE T 2 weighted image, a slightly hyperintense solid mass lesion is seen in the suprasellar cistern compressing the left cerebral peduncle

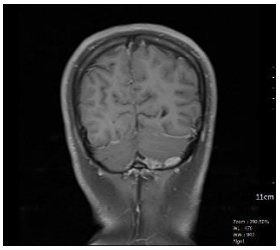

Figure 2. Coronal contrast-enhanced T 1 weighted image, two contrast-enhancing mass lesions seen in the left cerebellar region, consistent with leptomeningeal metastases

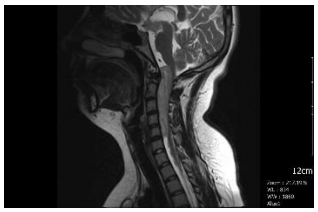

Figure 3. Sagittal FSE T 2 weighted image, a huge mass at the foramen magnum level, extends inferiorly and is severely compressing the cervical spinal cord. This is due to leptomeningeal dissemination of the primary suprasellar tumor which can also be seen on this image

Discussion

Although the JPA (Juvenile Pilocytic Astrocytoma) is considered to be a WHO (World Health Organization) grade 1 neoplasm, in contrast , the PMA has been classified by the WHO to be a grade 2 tumor (1,4). JPAs occur most frequently in the posterior fossa and the hypothalamic-chiasmatic region of children and young adults. These tumors exhibit an indolent clinical course and despite anaplastic evolution or CSF dissemination was reported in some cases, this has been extremely rare (2). The post-operative ten-year survival rate, even with partial tumor removal ranging from 80 % to 100 %. The survival rate with total tumor resection is 100 %. PMAs can occur anywhere along the neuroaxis, but have a strong geographic predilection for the hypothalamic-chiasmatic region and tend to affect a younger age group compared to JPAs (4,5). Local recurrences and CSF dissemination are more likely to occur in PMAs than JPAs. In a study performed by Komotar *et al*., (6), 16 out of 21 PMA patients (76 %) had local recurrences and three of these patients also developed CSF dissemination. Therefore, patients with JMA have higher local recurrences, but shorter survival rates as compared to JPA patients. PMA most commonly presents with symptoms of mass effect and raised intracranial pressure. The hypothalamic-chiasmatic JPA has a characteristic radiological appearance. They may have a solid and cystic component. These tumors usually do not induce peritumoral edema. On contrast-enhanced images, they mostly do not show enhancement except for solid mural nodules. Obstructive hydrocephalus can develop in later periods and 10 % of JPAs may show calcification. These typical radiological features together with the patient's age and characteristic tumor location, enable JPA diagnosis with high confidence. Since PMA is a relatively new recognized tumor, there are very few characteristic imaging findings belonging to this tumor described in the literature. Arslanoglu *et al*., (7) in their

study, described the most prominent radiological features of JMAs in order to distinguish them from JPAs. These tumors are predominantly solid in nature and show homogenous contrast enhancement, hydrocephalus, the extension of T2 signal intensity abnormality into the deep white matter and gray matter and CSF dissemination. JPA can be associated with neurofibromatosis (NF) syndrome and may involve the optic pathways (8). Linscott LL *et al*., (9) in their study, reported 4 PMA cases in the setting of NF type 1 and evaluated them as some other form of NF 1 associated tumors. Linscott LL *et al*., (9) also reported that nearly half of the PMAs in their series occurred in atypical locations, a finding that suggests that this tumor can occur elsewhere in the central neuroaxis. They also found that atypical tumor locations are more commonly encountered in older patients. In contrast to PMA cases, hemorrhage is uncommon in JPA and when present, intratumoral hemorrhage may be an important finding suggesting PMA (9). PMA is a monomorphous neoplasm composed of piloid tumor cells lying within a rich myxoid background (2). The tumor cells usually exhibit a striking angiocentric pattern. Eosinophilic granular bodies, Rosenthal fibers and biphasic pattern are extremely rare in JMA but is commonly encountered in JPA. The main treatment of JMA is gross total tumor resection which provides a favorable outcome in most cases, but the prognosis of these cases is greatly influenced by tumor location. Cerebellar tumors are often cured with surgery without needing any additional therapy, but hypothalamic-chiasmatic gliomas conversely are not amenable to total tumor resection (6,10). PMAs usually require adjuvant chemotherapy or radiotherapy in order to control the disease progression, but there are no specific adjuvant therapy protocols established for PMA (2). PMA has a tendency to appear during infancy and early childhood and its association with NF type 1 indicate that it may be related to a congenital or genetic origin with unique gene mutations (11,12).

PMA is usually considered as an aggressive variant of JPA, but in fact, it is an entirely different clinical and histopathological entity. Although it may have some distinguishing MRI findings, there are actually no pathognomonic imaging findings to discriminate between the two. Here, histopathological evidence is required for correct diagnosis. Clinical suspicion and early imaging detection may lead to better prognosis and survival rates in the management of these tumors.

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