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Original Article

Pilocytic Astrocytoma with Massive Microcalcification in a Child: A Tumor with Excellent Prognosis

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Abstract

Background: Pilocytic astrocytoma (PA) is the most common primary astroglial neoplasm in children and adolescents. Massive calcification is a rare feature of PA and may complicate radiologic and pathologic diagnosis. Herein, we report a case of PA with extensive microcalcification in western Iran.

Case Presentation: A 12-year-old boy presented with a two-year history of headache and vomiting that had worsened during the previous 15 days. Physical examination revealed a conscious and hemodynamically stable patient with no focal neurologic deficits. Histopathologic examination demonstrated a fascicular and microcystic tumor composed of cells with oval to spindle-shaped nuclei and bipolar cytoplasmic processes within a fibrillary matrix. Hyalinized arborizing ectatic vessels, Rosenthal fibers, and extensive areas of microcalcification were also identified.

Conclusion: Calcification is an uncommon feature of PAs and may create diagnostic challenges for radiologists and pathologists. Calcified PAs may occur at any age, with an approximately equal male-to-female ratio. Headache, vomiting, seizures, and visual disturbances are among the most common presenting symptoms in patients with calcified PA.

Keywords: Child, Calcification; Pilocytic astrocytoma

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Introduction

Pilocytic astrocytoma (PA) is the most frequently occurring primary astroglial neoplasm among children and teenagers (1-7). It accounts for approximately 10% of cerebral tumors and 85% of cerebellar tumors in pediatric patients (1). PAs are classified as WHO grade I tumors and are considered the most benign subtype of astrocytoma because of their favorable prognosis (1, 2).

The neuroimaging characteristics of PAs, including findings on conventional and advanced magnetic resonance imaging (MRI), vary according to tumor size, location, and extent of infiltration into adjacent structures (3). Massive calcification is a rare finding in PA and may lead to diagnostic difficulties in both radiologic and histopathologic evaluations (4). Immunohistochemically, PAs express astrocytic lineage markers and typically demonstrate strong positivity for glial fibrillary acidic protein (GFAP) (1).

The present study reports a case of PA with extensive microcalcification in a child from western Iran.

Case Report

A 12-year-old boy from a village in western Iran was admitted to the neurosurgery ward on February 7, 2017, with a two-year history of headache and vomiting. His symptoms had worsened during the preceding 15 days. The headaches were generalized and more severe at night. There was no history of convulsion, gait disturbance, or visual loss; however, the patient had experienced insomnia and malaise during the previous two years. His past medical and drug histories were unremarkable.

On physical examination, the patient was conscious and hemodynamically stable. Neurologic examination, including cranial nerve and spinal examination, was normal.

Brain MRI performed on January 17, 2017, revealed an approximately 4 × 5 × 6 cm posterior fossa mass lesion located posterior to the fourth ventricle, with cystic changes and possible massive calcification or hemorrhage, accompanied by mild hydrocephalus. The initial radiologic impression was medulloblastoma.

The imaging studies were subsequently reviewed by two additional radiologists. Their differential diagnoses included medulloblastoma, PA, and ependymoma because of the atypical radiologic appearance of the lesion.

The patient underwent surgical resection on February 19, 2017, and the specimen was submitted for histopathologic examination. Grossly, the specimen consisted of fragmented gray soft tissue measuring 5 × 4 × 3 cm in aggregate.

Microscopic examination demonstrated a tumor with fascicular and microcystic architecture composed of cells with oval to spindle-shaped nuclei and bipolar cytoplasmic processes embedded within a fibrillary matrix. Hyalinized arborizing ectatic vessels, Rosenthal fibers, and extensive areas of microcalcification were also observed (Figure 1).

Immunohistochemical analysis showed diffuse positivity for GFAP and negativity for epithelial membrane antigen (EMA) (Figure 2). Based on these findings, the tumor was diagnosed as PA, WHO grade I.

Postoperative gadolinium-enhanced MRI performed on February 21, 2017, demonstrated mild ventricular dilatation, a shunt within the left lateral ventricle, and an irregular hyperintense area in the vermis region with minimal irregular gadolinium enhancement. Follow-up MRI was recommended to evaluate for residual neoplastic tissue.

At follow-up on April 19, 2017, the patient was in good clinical condition.

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Table I: The cases with pilocytic astrocytoma and calcification

Reference	Sex	Age, year	Presenting symptoms	Duration of symptoms, months	Tumor location
(2)	F	17	Headache off and on/ Nonbilious, and nonprojectile vomiting	1 month/2 week	Lateral ventricles in the proximity of the foramina of Monro
(4)	M	3	Vomiting and imbalance	~3	Right lateral ventricle
(5)	M	13	Headache and visual disturbance	6	The suprasellar region was associated with enlargement of the lateral and third ventricles
(6)	F	7	Seizures associated with headache	6	Intraventricular
(6)	M	58	Headache and diminution of vision in both eyes	12	Left lateral ventricle extending into the third ventricle
(8)	F	11	Headache, retro-orbital pain, vomiting, and bilateral diminution of vision	1	Pineal region
(8)	F	22	Gradually progressive severe headache /altered sensorium and recurrent episodes of seizures	6 months/1 week	Pineal region and extended into the third ventricle and lateral ventricles
(9)	M	7	Headache, visual disturbance, and psychomotor seizures	7	Right temporal
(10)	M	36	NA	NA	Right parietal lobe
(11)	F	28	Difficult swallowing solids and progressive left hemiparesis	~144	Forth ventricle
(12)	F	39	progressive pain and numbness, predominantly in the S-1 dermatome	12	Tip of the conus medullaris
(13)	M	6	Vomiting, increasing head size, altered sensorium, weakness of lower limbs, loss of appetite and loss of weight	NA	Suprasellar
(14)	F	15	Complex partial seizure	NA	Left lateral ventricle
(15)	F	38	Acute confusional state and right-sided spastic hemiparesis	NA	Left temporal
(16)	M	21	Acute onset of generalized Tonic-clonic seizure	NA	Right frontal subcortical region
The present case	M	12	Headache and vomiting	24	left lateral ventricle

Abbreviations: M, male; F, female; NA, not available

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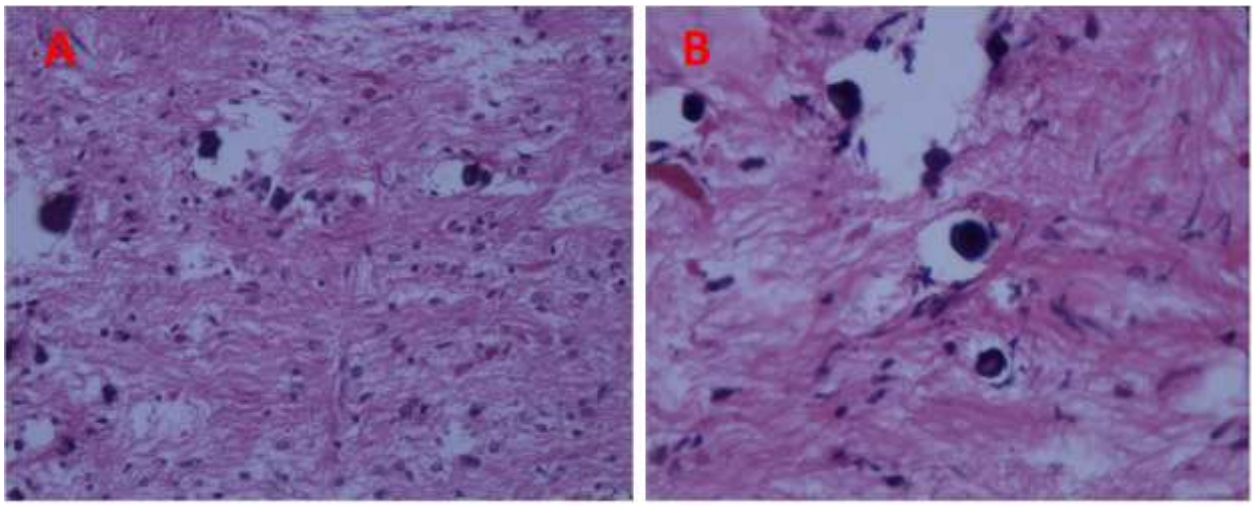

Figure 1: Pilocytic astrocytoma with microcalcification: (A) Hematoxylin-Eosin staining with magnification of ×200, (B) Hematoxylin-Eosin staining with magnification of ×400

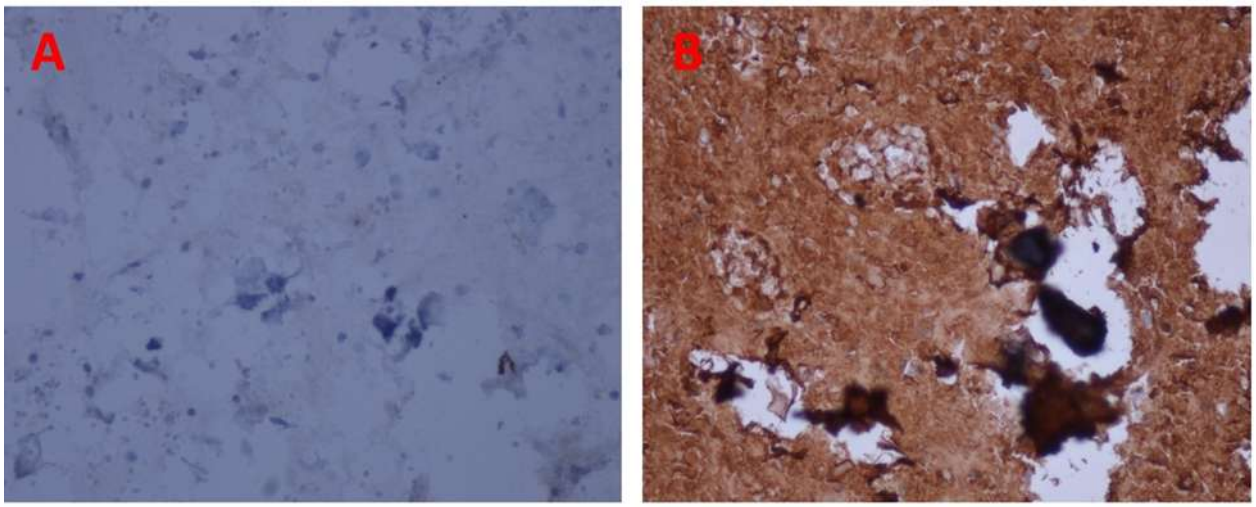

Figure 2: Immunohistochemistry of (A) EMA with magnification of ×200, (B) GFAP with magnification of ×200

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Discussion

The present study describes a pediatric case of PA with extensive microcalcification involving the left lateral ventricle. Histologically, PA should be differentiated from several entities, including pleomorphic xanthoastrocytoma, diffuse astrocytoma, glioblastoma multiforme, oligodendroglioma, and piloid reactive gliosis.

Calcification occurs only occasionally in PAs and is usually limited to thin or fleck-like deposits (1,4,5). Extensive tumor calcification is rare and is thought to reflect the chronic nature and slow growth of the lesion (5). MRI findings in calcified PAs have been described as showing low signal intensity on both T1- and T2-weighted sequences, or as appearing iso- to hypointense on T1-weighted images and hyperintense on T2-weighted images, often accompanied by prominent peripheral contrast enhancement (5,7,8).

Reported cases of PA with calcification indicate a mean age at diagnosis of 20.8 years (range: 3–58 years), with approximately equal sex distribution. Common presenting symptoms include vomiting, seizures, headache, and visual disturbances. Symptom duration has ranged from approximately 3 to 144 months. Tumor locations reported in the literature are summarized in Table I (2, 4, 5,6, 8-16).

PAs occur most frequently in children and young adults between 0 and 19 years of age and account for approximately 10% of cerebral tumors and 85% of cerebellar tumors (1). These tumors commonly involve the cerebellum, optic pathway, midbrain, and medulla (17). Sattar et al. reported that PAs may arise throughout the neuraxis, with approximately 67% occurring in the cerebellum and optic pathway. PAs generally have an excellent prognosis, and calcification is considered a benign feature associated with slowly growing tumors (2).

Neoplasms that develop in the lateral ventricles represent under 1% of all brain tumors, although they occur with greater frequency in children than in adults. Clinical manifestations of PA vary according to tumor location and may include headache, nausea, vomiting, seizures, ataxia, and visual disturbances (18,19). In cerebellar PAs, headache, vomiting, gait disturbance, blurred vision, diplopia, and neck pain are among the most common presenting symptoms (5).

Conclusion

Calcification is a rare finding in PAs and may create diagnostic challenges for both radiologists and pathologists. Calcified PAs may occur at any age, although most reported cases occur in patients younger than 30 years. The male-to-female ratio is approximately 1:1. Headache, vomiting, seizures, and visual disturbances are among the most common presenting symptoms in patients with calcified PA.

Availability of Data

None

Ethical Considerations

None

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Authors' Contributions

M.R and M.S: Designing and writing of manuscript.

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Conflict of Interest

The authors declare no conflict of interest.

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