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Pediatric cerebellar pilomyxoid-spectrum astrocytomas:

Report of 2 cases

Jonathan A. Forbes, M.D.¹, Bret C. Mobley, M.D.², Thomas M. O'Lynnner, M.D.¹, Calvin M. Cooper, B.S.³, Mahan Ghiassi, M.D.¹, Rimal Hanif, B.S.³, and Matthew M. Pearson, M.D.¹

¹Department of Neurosurgery, Vanderbilt University Medical Center, Nashville, Tennessee

²Department of Pathology, Vanderbilt University Medical Center, Nashville, Tennessee

³Vanderbilt School of Medicine, Vanderbilt University Medical Center, Nashville, Tennessee

Abstract

Object—Pediatric cerebellar astrocytomas with pilomyxoid features include classic pilomyxoid astrocytomas (PMAs) and intermediate pilomyxoid tumors (IPTs). Since the original description of PMA in 1999, most reports in the literature have described PMAs arising from the hypothalamic/chiasmatic region. To the authors' knowledge, PMAs arising from the posterior fossa have not been discussed in the neurosurgical literature. Intermediate pilomyxoid tumors, or tumors with pathological features of both pilocytic astrocytoma (PA) and PMA, have only recently been described. In this article, the authors present 2 cases that fall within the spectrum of pediatric cerebellar PMA—including a classic PMA and an intermediate pilomyxoid tumor. The authors compare the radiological presentation, surgical results, and postoperative course to findings in a cohort of 15 patients with cerebellar PAs.

Methods—Between 2003 and 2010, 2 patients with pilomyxoid-spectrum astrocytomas underwent treatment at Vanderbilt Children's Hospital. One was a 22-month-old girl who presented with progressive gait disturbance and falls. The other was a 4-year-old girl who presented with ataxia and generalized weakness. In a retrospective review of pediatric cerebellar neoplasms resected by the senior author during this period, these tumors comprised 4% of cerebellar neoplasms and approximately 10% of cerebellar glial neoplasms.

Results—Both patients were treated with midline suboccipital craniotomy for resection. In both cases, tumor invasion anteriorly into the brainstem prevented gross-total resection. the patient in Case 1 was placed on chemotherapy following pathological diagnosis and later developed definitive evidence of leptomeningeal dissemination (LD) 3 years after the operation. The patient

Address correspondence to: Jonathan A. Forbes, M.D., 915 9th Avenue North, Nashville, Tennessee 37208.
jonathan.forbes@vanderbilt.edu.

These 2 patients were presented as part of an analysis of MR imaging findings in 50 patients with cerebellar neoplasms at the Annual Meeting of the American Association of Neurological Surgeons/Congress of Neurological Surgeons Section on Pediatric Neurological Surgery, which was held on December 14, 2009.

Disclosure

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in Case 2 was placed on chemotherapy after exhibiting progressive evidence of local recurrence (findings were negative for LD) 12 months following resection.

Conclusions—Pediatric patients with cerebellar pilomyxoid-spectrum astrocytomas appear to suffer higher rates of local recurrence and LD than pediatric patients with cerebellar PAs.

Keywords

cerebellar; neoplasm; pilomyxoid; T2 hyperintensity; children; astrocytoma; oncology

In 1999, Tihan and colleagues provided the sentinel description of PMAs.¹⁰ Because of their similar histological and radiological features,⁷ PMAs were classified as PAs prior to acceptance of this publication. In contrast to PAs, PMAs demonstrate a more aggressive clinical course¹¹ and appear to be associated with a higher incidence of leptomeningeal spread.⁵ Pilomyxoid astrocytomas are considered to be WHO Grade II neoplasms, and most often arise from the hypothalamic/chiasmatic region. However, PMAs sometimes originate from the posterior fossa.⁵ Whereas hypothalamic PMAs have been well described in the literature, there is a relative paucity of information about PMAs that arise from the cerebellum. Although radiological⁷ and pathological findings⁴ for cerebellar PMAs have been described, to our knowledge, a full clinical characterization of pediatric cerebellar PMAs has not been reported. Thus, it is not entirely certain whether PMAs in the posterior fossa exhibit increased biological aggressiveness relative to PAs, as has been observed in the hypothalamic/chiasmatic region.¹¹ In a recent multicenter review of 84 cases of pediatric astrocytomas with pilomyxoid features, Johnson et al.⁴ reported the histopathological findings in 42 patients with classic PMAs, in addition to another subset of 42 patients with histopathological features typical of both PMAs and PAs. This latter subset was named “intermediate pilomyxoid tumors.” Unfortunately, a lack of clinical information limited the ability of these authors to analyze outcomes in patients with these intermediate tumors. At present, it is therefore also unknown whether intermediate tumors with histopathological features of both types of lesions should be treated more like PAs or PMAs.

In this report, we describe the radiological presentation, histological features, and clinical course in 2 pediatric patients who presented to Vanderbilt Children’s Hospital with pilomyxoid-spectrum astrocytomas. One child was found to have a tumor with histopathological characteristics consistent with a classic PMA, whereas the other harbored an IPT. The literature on this topic is reviewed and management of these neoplasms is discussed.

Case Reports

Case 1

History and Examination—This 22-month-old girl was brought to the emergency department with a 6-month history of progressive gait disturbance and frequent falls. Neurological examination in the emergency department revealed an awake and alert toddler with no cranial nerve deficits and preserved strength. A wide-based ataxic gait was observed. An MR imaging study of the brain revealed a 4-cm heterogeneously enhancing lesion occupying the fourth ventricle, accompanied by obstructive hydrocephalus, with dilation of the third and lateral ventricles (Fig. 1A–F). Preoperative MR imaging of the spine was performed and failed to demonstrate any evidence of meta-static foci.

Operation—The patient underwent a midline suboccipital craniotomy for resection of the posterior fossa mass. Following telovelar dissection, a gelatinous gray tumor was encountered and was gently elevated from the floor of the fourth ventricle. Dissection planes

were developed dorsally and ventrally to the left; however, the superior right and middle aspects of the ventral tumor were noted to be grossly infiltrative into the cerebellar peduncle and dorsolateral brainstem. Meticulous dissection was continued along the ependymal plane in this region, and the tumor was delivered. Some residual tumor was left at the site of tectal invasion. Histological findings on frozen section were thought to be consistent with PA.

Pathological Findings—The H & E–stained sections demonstrated a neoplasm composed of cells with monomorphous round-to-ovoid nuclei. The tumor background was variable, demonstrating myxoid histological features characteristic of PMA in areas of low cellularity, but exhibiting a process-rich fibrillary background characteristic of classic PA in areas of higher cellularity (pilocytic/pilomyxoid transition zone depicted in Fig. 1G). Tumor cells demonstrated angiocentric arrangements in the myxoid regions (Fig. 1H and I). Vascular proliferation was present. Up to 2 mitotic figures were identified per 10 hpf. No calcifications or oligodendroglioma-like cells were observed, and no eosinophilic granular bodies or Rosenthal fibers were seen. The tumor did not demonstrate an infiltrative edge with the adjacent brain parenchyma. Immunohistochemical investigation for glial fibrillary acidic protein revealed strong, diffuse immunoreactivity in neoplastic cells, highlighting piloid processes. The MIB-1 proliferation index was regionally variable, ranging from 1% to 3%. Overall, the combined features of both PA and PMA were retrospectively indicative of an IPT. A diagnosis of “pilocytic astrocytoma with pilomyxoid features” was rendered.

Postoperative Course—Postoperatively, findings on the patient’s neurological examination were stable in comparison with her preoperative ones. Postresection MR imaging of the brain demonstrated interval resection of the fourth ventricular mass, with residual enhancement noted within the superior fourth ventricle, superior cerebellar peduncles, and tectum (Fig. 2A and B). After extensive discussions with members of the neuropathology and neurosurgery departments, the patient’s medical oncology team recommended treatment of the neoplasm as a PMA, with adjuvant chemotherapy. Because it was believed that additional tumor visualized on her postoperative MR imaging studies could be resected, a second operation was undertaken. The patient was returned to the operating room approximately 1 week later, where tumor was visualized and confirmed by Stealth navigation. Portions of residual tumor were resected. However, indistinct margins between the tumor and tectum anteriorly and cerebellum laterally again prevented GTR. Postoperative imaging demonstrated significant interval resection of residual tumor (Fig. 2C and D). Results of the postoperative neurological examination were notable for mild dysmetria and slightly worsened ataxia.

On histopathological examination of the specimen obtained in the second resection, areas with a myxoid background and angiocentric arrangement of tumor cells were identified only focally; the specimen’s histological characteristics were now dominated by a biphasic, compact, spongy architecture classic for PA, with regions of oligodendroglioma-like cytological features seen. The aforementioned histopathological features of this second surgical specimen were thought to be closer to those of a PA than the first specimen. However, given the histopathological findings in the first surgical specimen, pediatric neurooncology specialists formally recommended a chemotherapeutic regimen of carboplatin and vincristine.

The patient developed hydrocephalus in the weeks following her second operation, necessitating ventriculoperitoneal shunt insertion. Repeat MR imaging of the brain obtained approximately 1 month after the most recent craniotomy for resection revealed an enhancing focus that initially was thought likely to be consistent with leptomeningeal disease near the ventral surface of the pontomedullary junction. Additional foci of enhancement were noted at the midthoracic spinal cord and at the conus medullaris extending into the pelvis.

However, these foci had resolved on interval imaging at 5-month follow-up. The patient was noted to be clinically stable, with full strength and improved coordination, at this time. Fifteen months postsurgery, carboplatin and vincristine were discontinued due to intolerable side effects, in favor of temodar and vinblastine. Serial MR imaging studies obtained 36 months after initial resection revealed evidence of disease progression, with new masslike thickening and enhancement of the infundibulum, in addition to a new cystic lesion involving the left choroid plexus and a small focus of abnormal enhancement in the right cerebellopontine angle (Fig. 3). At 45 months postsurgery, MR imaging revealed progression of intracranial disease, in addition to new evidence of leptomeningeal metastases involving the sacrum. The patient's chemotherapy regimen was changed to bevacizumab and irinotecan at this time. The most recent MR imaging studies of the neuraxis obtained at 57 months following surgery revealed stable disease on this regimen.

Case 2

History and Examination—This 4-year-old girl presented with what her parents said was a 1-week history of upper-back and arm pain, in addition to ataxia and generalized weakness in all 4 extremities. Neurological examination revealed no cranial nerve deficits, 4/5 muscle strength in both upper and lower extremities, and an unsteady gait. An MR imaging study of the brain revealed a 4-cm posterior fossa mass with extension into the dorsal left midbrain, with associated obstructive hydrocephalus (Fig. 4A–F). The presence of necrosis and an infiltrating border was thought to be possibly suggestive of a high-grade neoplasm.

Operation—The patient underwent a midline suboccipital craniotomy for resection of the posterior fossa neoplasm. A left paravermian approach was chosen. The pia mater in the left paravermian region was coagulated and opened with microscissors. Bipolar electrocautery and suction were used to expose the tumor, which was noted to have a distinct capsule intimately associated with the cerebellum. The superior aspect of the tumor was noted to be intimately associated with the inferior aspect of the tentorium. Invasion anterior into the brainstem required meticulous dissection. Tumor extension through the incisura was also noted, with intimate association of the tumor with 2 large draining veins of the cerebellum. Because of the of the neoplasm, GTR could not be accomplished.

Pathological Findings—The H & E–stained sections showed a cellular neoplasm with extensive myxoid pericellular material and a radial arrangement of tumor cells around blood vessels (Fig. 4G–I). Neoplastic cells were bipolar, with thick, glial fibrillary acidic protein–immunoreactive processes. There was focal necrosis and an extensive vascular network, including glomeruloid vascular proliferation. The tumor did not demonstrate an infiltrative edge with the adjacent brain parenchyma. Epithelial membrane antigen immunohistochemical studies were performed, and no ependymoma-like epithelial membrane antigen immunoreactivity was seen. The MIB-1 proliferation index was generally low (< 1%). A diagnosis of “pilomyxoid astrocytoma, WHO grade II” was rendered.

Postoperative Course—Following surgery, the patient was noted to be neurologically stable in comparison with results of the preoperative examination. An MR imaging study obtained the day after surgery demonstrated a very small amount of residual enhancement superiorly at the surgical margin of the resection cavity (Fig. 5A and B). The patient developed leakage from the wound approximately 1 week after the operation, necessitating eventual ventriculoperitoneal shunt insertion. An MR imaging study obtained 1 month after surgery demonstrated an increase in residual neoplasm in comparison with the previous studies (Fig. 5C and D). Per her family's request, the patient's neurooncological care was transferred to another facility at this time, where the choice was made to defer chemotherapy.

An MR imaging study of the neuraxis obtained 4 months after the initial surgery again revealed progressive local recurrence, but was negative for evidence of leptomeningeal spread. Subsequent MR imaging studies performed 9 months after surgery revealed stable tumor size without evidence of disease progression. Repeat imaging of the neuraxis 12 months postoperatively again revealed interval progression of local disease. Neurological evaluation at this time revealed a slight improvement in right hemiparesis (4+/5), despite mild progression of ataxia. In the context of interval progression on MR imaging, the patient's family requested that neurooncological care be transferred back to Vanderbilt for initiation of chemotherapy (carboplatin, vincristine, and temozolomide). At the time of this writing, the patient has received a total of 5 cycles of chemotherapy, with plans to repeat neuraxis imaging at the end of her chemotherapy regimen.

Discussion

Under the current WHO grading scheme, PAs are designated Grade I and PMAs are designated Grade II neoplasms.⁸ Because of similar radiological and histological features, PMAs were considered PAs prior to their sentinel description in 1999.¹⁰ In contrast to the fibrillary background and biphasic cellular pattern observed with PAs, PMAs display a predominantly myxoid background with a perivascular arrangement of tumor cells, reminiscent of ependymomatous pseudorosettes. The myxoid background and angiocentric tumor cell arrangement are typically discernible on a low-magnification examination of H & E-stained material. Examination at high power then reveals a monomorphous population of small bipolar cells and an absence of Rosenthal fibers and eosinophilic granular bodies. Following the introduction of PMA as a diagnostic entity, pathologists have come to recognize that occasional tumors show features typical of PMA in combination with features that are more consistent with PA. Called "intermediate pilomyxoid tumors," these lesions show some combination of myxoid substance, with cellular monomorphism and angiocentric growth, and pilocytic features such as compact fibrillary tissue and Rosenthal fibers.⁴ Histopathological differentiation within the pilomyxoid spectrum is based on morphological analysis of H & E-stained material; the utility of other methods of differentiation—including immunohistochemical staining and molecular analysis—remains speculative at this time.^{1,4}

Histopathological distinction between PA and PMA is useful because PMAs have been associated with increased rates of local recurrence and LD, as well as decreased overall survival relative to PAs.⁵ Although it is not uncommon for conventional PAs to spread locally to involve the leptomeninges, the incidence of LD in cerebellar PAs is extremely rare,² and many neurosurgeons do not routinely obtain MR imaging of the neuraxis to rule out LD before or after resection.³ Thus, pathological designation in these patients may influence the frequency and scope of surveillance imaging. The risk-benefit analysis of postresection adjuvant therapy is influenced by histological features as well. Complete surgical excision of a PA is believed to obviate the need for adjuvant therapy¹²; however, adjuvant chemotherapy has been recommended as initial therapy in the treatment of PMAs.¹¹ Although experience with diagnosis and treatment of IPTs is limited at this time, reports describing the behavior of these pilomyxoid-spectrum tumors help to improve our understanding of IPT behavior and influence future treatment decisions.

Most of the knowledge regarding the clinical behavior of PMAs comes from case series predominantly comprising neoplasms located in the chiasmatic/hypothalamic region. These descriptions indicate a significantly higher rate of local recurrence associated with PMAs than what has been documented with PAs. In a report by Komotar et al.,⁵ 76% of patients with PMAs exhibited local recurrence, versus 50% of those with PAs. This group found increased rates of LD as well—3 (14%) of 21 patients with PMA demonstrated evidence of

eventual LD, in comparison with 0 of 42 patients with PA. In a recent report by Tsughu et al.,¹¹ 3 of 5 patients with hypothalamic PMAs had evidence of LD at the time of diagnosis. In contrast, previous published rates of LD in patients with PAs followed over long intervals range from 0% to 1.8%.^{2,9} Komotar et al.⁵ also found PMAs to have significantly decreased mean times of progression-free and overall survival. These trends have been described in other studies.¹⁰ Whereas the literature is replete with clinical series of pediatric patients with hypothalamic/chiasmatic PMAs, descriptions of cerebellar PMAs are rare. In 1 compilation describing the radiological features of 21 patients with PMAs from 7 different institutions, 2 patients (10%) had tumors that arose from the cerebellum.⁷ In another compilation of 84 patients with pathological findings of pediatric astrocytomas with pilomyxoid features, 2 patients harbored cerebellar tumors, in addition to 6 other patients who harbored tumors in an “unspecified” location in the posterior fossa.⁴ To our knowledge, this report is the first in the neurosurgical literature fully to characterize the clinical findings of pediatric cerebellar PMAs.

When salient histopathological features allow a definitive diagnosis of PA or PMA, oncological care of the patient becomes more straightforward. However, tumors with histopathological features of both PA and PMA, which are called “intermediate pilomyxoid tumors,” are encountered in a significant proportion of patients analyzed.⁴ How then does the oncology team approach the treatment of patients with IPTs? The answer will depend on the clinical behavior of these intermediate neoplasms. Although the study by Johnson et al.⁴ did much to address the histopathological spectrum of pilomyxoid tumors, there was limited access to clinical information—precluding an analysis in which tumor histological characteristics could be linked with patient outcome. As a result, the clinical implications of a diagnosis of IPT are currently unknown. Additionally, because the study represented a multicenter conglomeration of referral cases, the true incidence of these tumors could not be estimated from information in the report.

Observations from this series of pilomyxoid-spectrum tumors are consistent with previous studies describing a more aggressive clinical behavior in comparison with PAs. During the 8 years for which data were retrospectively reviewed for this report, 1 patient with a cerebellar PMA, 1 with an IPT, and 15 with cerebellar PAs were identified. The GTR was notably less in the 2 patients with tumors that had pilomyxoid features in comparison with the patients with PAs (0% vs 73%). Additionally, the 2 patients who harbored pilomyxoid-spectrum tumors were found to have higher rates of local recurrence and subarachnoid dissemination than the group of 15 patients with cerebellar PAs (Table 1). Specifically, the patient in Case 1 with the IPT developed questionable evidence of LD 4 weeks after subtotal resection and clear evidence of intracranial and intraspinal LD on repeat imaging approximately 3 years later. The patient in Case 2 harbored a PMA that exhibited evidence of progressive local recurrence on interval imaging and was eventually treated with chemotherapy approximately 12 months after surgery. An MR imaging study of the neuraxis in the latter patient was negative for the presence of LD. Of the 15 patients who harbored PAs, none—with follow-up ranging from 7 months to 4 years—demonstrated evidence of leptomeningeal spread (it should be noted that only 6 of these patients have undergone formal imaging of the neuraxis to rule out LD). It is notable that the rate of LD of patients with cerebellar pilomyxoid-spectrum tumors in this series (50%) was higher than what has been described in previous series of patients harboring hypothalamic/chiasmatic PMAs (14%).⁵ Although data regarding the true incidence of LD in cerebellar PMAs are lacking, routine imaging of the neuraxis in this patient population should be considered.

Given the proclivity of PMAs to recur locally and to disseminate through subarachnoid pathways, in addition to documented associated decreases in overall survival, a lower threshold for adjuvant therapy and an increased frequency of serial imaging of the neuraxis

in patients with PMAs has been proposed by many clinicians.^{6,11} Due to the very recent description of IPTs, current recommendations regarding adjuvant therapy are lacking. Because the tissue specimen analyzed in the patient in Case 1 of this series exhibited pilomyxoid features, the medical oncology team followed a treatment paradigm appropriate for PMA, with administration of chemotherapy immediately prior to the patient's postoperative MR imaging 4 weeks after resection. In this patient, the presumed foci of leptomeningeal metastases visible at 1-month follow-up had regressed by 5 months on a regimen of carboplatin and vincristine. However, due to intolerable side effects, this regimen was discontinued in favor of temodar and vinblastine. The patient was found to have new, definitive evidence of leptomeningeal metastases at 36 months, with confirmed progression at 45 months. The decision was then made to transition to bevacizumab and irinotecan. On this regimen, the metastatic foci were noted to be stable at 57 months. Although the utility of treatment with chemotherapeutic agents in patients with IPTs is presently unknown, a review of this case suggests a possible benefit.

Conclusions

After a review of the charts of 50 pediatric patients who underwent resection of cerebellar neoplasms by the senior author (M.M.P.) since 2003 at Vanderbilt Children's Hospital, 2 patients with pilomyxoid-spectrum astrocytomas were identified: 1 had an IPT and 1 had a tumor with classic PMA histological features. Tumors with pilomyxoid features comprised 4% of cerebellar neoplasms in this series and approximately 10% of cerebellar glial neoplasms. Pediatric patients with cerebellar pilomyxoid-spectrum astrocytomas in this series suffered higher rates of local recurrence and LD than pediatric patients with cerebellar PAs.

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Abbreviations used in this paper

GTR	gross-total resection
IPT	intermediate pilomyxoid tumor
LD	leptomeningeal dissemination
PA	pilocytic astrocytoma
PMA	pilomyxoid astrocytoma

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Fig. 1.

Case 1. **A–C:** Axial cuts from T1-weighted post-Gd MR imaging studies of the brain demonstrating a heterogeneously enhancing 4-cm lesion occupying the fourth ventricle and extending into the tectum anteriorly. **D:** The same neoplasm, as visualized on a midsagittal T1-weighted post-Gd MR imaging study of the brain. **E:** Axial cut from a T2-weighted MR imaging study of the brain demonstrating that the lesion is extremely hyperintense—almost as hyperintense as CSF and vitreous humor. **F:** On diffusion-weighted imaging, the neoplasm is hypointense in relation to normal, surrounding cerebellar tissue. **G–I:** Photomicrographs of surgical specimens prepared with H & E staining. **G:** Low-magnification view demonstrating the transition from a myxoid tumor zone (*left side of field*) to a region with a more process-rich background (*right side of field*) characteristic of classic PA. **H and I:** High-magnification views of myxoid areas showing tumor cells arranged in an angiocentric pattern. Nuclear monomorphism is noted. Eosinophilic granular bodies and Rosenthal fibers are notably absent.

Fig. 2.

A and B Axial and sagittal cuts from T1-weighted post-Gd MR imaging studies of the brain demonstrating resection of the fourth ventricular mass, with residual enhancement noted within the superior fourth ventricle, superior cerebellar peduncles, and tectum. **C and D:** Axial and sagittal cuts from T1-weighted post-Gd MR imaging studies obtained after the second operation, demonstrating significant interval resection of residual tumor.

Fig. 3.

A and B Axial and coronal cuts from T1-weighted post-Gd MR imaging studies of the brain demonstrating new, abnormal thickening and enhancement of the infundibulum (*thick arrows*). **C and D:** Axial and coronal cuts demonstrating a small focus of abnormal enhancement in the right cerebellopontine angle (*thin arrows*) and enhancing lesion in the anterior aspect of the foramen magnum (*arrowhead*). These findings were considered to be a matter of concern for intracranial LD.

Fig. 4.

Case 2. **A–C:** Axial cuts from T1-weighted post-Gd MR imaging studies of the brain demonstrating a 4-cm posterior fossa mass with extension into dorsal left midbrain. **D:** The same neoplasm, as visualized on a midsagittal MR imaging study of the brain. **E:** Axial cut from a T2-weighted MR imaging study demonstrating a moderately hyperintense lesion. **F:** On diffusion-weighted imaging, the lesion is hypointense in relation to surrounding cerebellar tissue. **G–I:** Photomicrographs of surgical specimens prepared with H & E staining. **G:** A low-magnification section revealing an angiocentric pattern of neoplastic cells with prominent myxoid background. **H:** Tumor cell processes extend from a blood vessel cut in a longitudinal section. **I:** Perivascular tumor cells exhibit bland, monomorphous nuclei.

Fig. 5.

A and B Postoperative axial and sagittal T1-weighted post-Gd MR images obtained approximately 24 hours after resection showing a small amount of residual enhancement superiorly at the surgical margin of the resection cavity. **C and D:** Axial and sagittal cuts from T1-weighted post-Gd MR imaging studies obtained 1 month postoperatively demonstrating interval increase in residual neoplasm.

Clinical characteristics, operative results, and postoperative course in 2 patients with cerebellar pilomyxoid-spectrum tumors and 15 with JPAs*

TABLE 1

Histo Finding	No. of Patients	Mean Age (yrs)	% GTR	Mean PFS	% LR	% LD	% OS
PMA	2	3.5 (2–5)	0	1.5 yrs	50	50	100
JPA	15	7.0 (2–18)	73	2.7 yrs	20	0	100

* Histo = histopathological; JPA = juvenile PA; LR = local recurrence; OS = overall survival; PFS = progression-free survival.