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Pilocytic Astrocytoma: A Case Report

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Abstract | **Case Report**

Pilocytic astrocytoma (PA) is a WHO grade I glial tumor, most frequently affecting children and adolescents, with a predilection for the cerebellum. Vermian involvement presents unique surgical challenges due to its proximity to critical midline structures. Histologically, PA displays a biphasic architecture with Rosenthal fibers and eosinophilic granular bodies, and generally carries a favorable prognosis when treated appropriately. In cases where gross total resection is not possible, adjuvant chemotherapy with vincristine and carboplatin is often employed, particularly in young patients, to delay or avoid radiotherapy. This case report describes the anatomopathological features, radiological presentation, and therapeutic management of a cerebellar vermian pilocytic astrocytoma in an adolescent, highlighting the importance of a tailored, multidisciplinary approach

Keywords: Astrocytoma, Glial tumor, Vermis, prognosis, chemotherapy.

INTRODUCTION

Pilocytic astrocytoma is a slow-growing, generally benign brain tumour, classified as grade I by the World Health Organization. It is the most common glioma in children and adolescents, making up about a quarter of central nervous system tumours in this age group and around 5% across all ages. First described over a century ago, it is now well recognized for its typical appearance under the microscope, with a characteristic mix of dense and loose areas, Rosenthal fibres, and eosinophilic granular bodies.

Pilocytic astrocytoma most often develops in the cerebellum, especially in the posterior fossa, where it frequently appears as a cyst with a bright, contrast-enhancing nodule on imaging. Although its prognosis is generally favorable, the tumour's location can greatly influence treatment. When it grows in delicate midline areas such as the cerebellar vermis, complete surgical removal can be challenging and carries a higher risk of complications. In such situations, additional treatments like vincristine and carboplatin-based chemotherapy are often used to control the tumour and postpone radiotherapy, particularly in young patients to protect long-term brain function.

We present the case of an 18-year-old female with a pilocytic astrocytoma of the cerebellar vermis, managed at the Oncology-Radiotherapy Department of the Mohammed VI University Hospital in Marrakech.

CASE PRESENTATION

An 18-year-old high school student with no relevant medical, surgical, or toxic history, was referred for further care after the diagnosis and partial resection of a cerebellar tumor. She had previously enjoyed good health, and there was no family history of cancer.

Her symptoms began about two years earlier, initially with occasional vertigo and mild occipital headaches that gradually became more intense. Over time, she developed sensitivity to light and a progressive decline in vision in her right eye. Her condition significantly worsened after a year, when she developed right-sided weakness, followed by a generalized tonic-clonic seizure, prompting urgent evaluation.

A brain MRI was performed and revealed a large heterogeneous mass in the right cerebellar hemisphere, measuring 55.5 × 51 × 50 mm. The lesion had mixed cystic and solid components with lobulated borders, perilesional edema, and compression of the fourth ventricle, resulting in active tri-ventricular hydrocephalus and early tonsillar herniation. The mass also displaced the brainstem and cerebellar peduncles. No other abnormalities were found. The imaging raised

the possibility of either a medulloblastoma or a pilocytic astrocytoma.

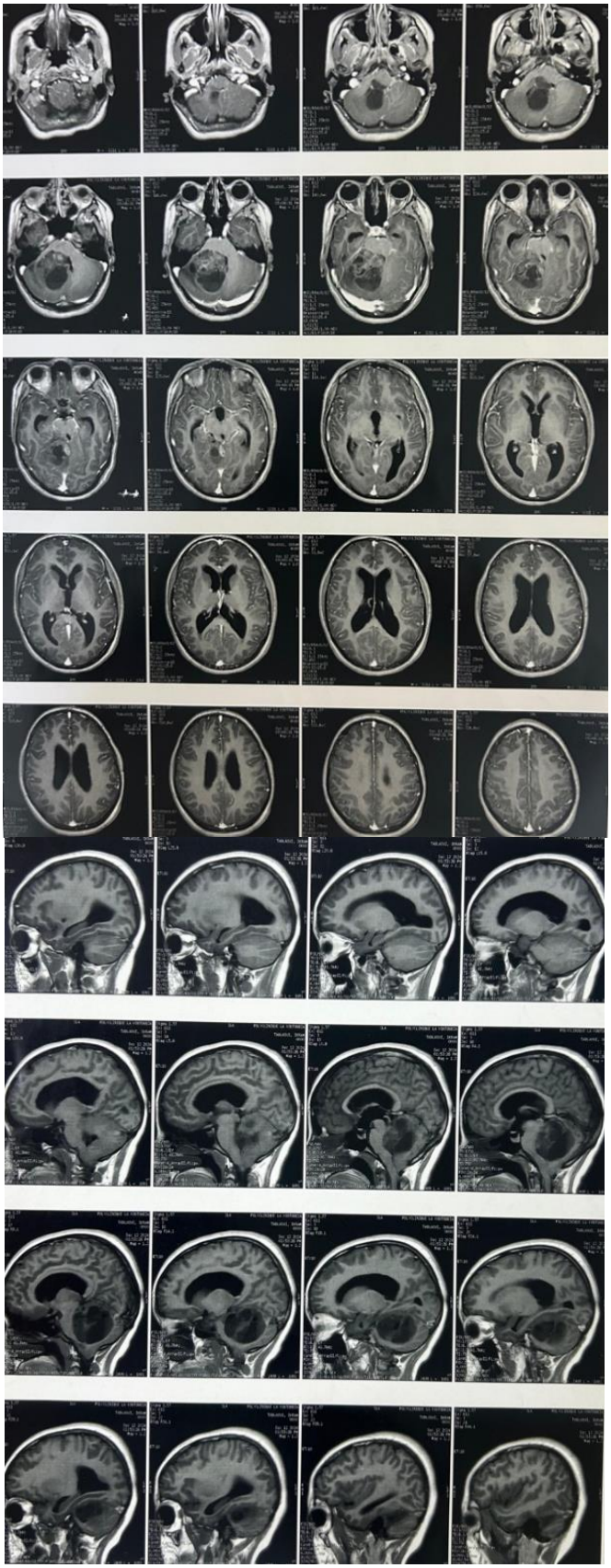

Figure 1 and 2: Compressive right cerebral hemispheric mass

She was admitted to the neurosurgery department at CHU Ibn Tofaïl. The first intervention involved placing a ventriculoperitoneal shunt to manage the hydrocephalus. then, she underwent a second surgery on march the 27th 2024 for biopsy and partial removal of the cerebellar lesion.

Histological examination of the specimen revealed a low-grade glial tumor. Immunohistochemical analysis confirmed the diagnosis of a pilocytic astrocytoma, with tumor cells staining positive for GFAP, OLIG2, and ATRX, and negative for P53 and IDH1-R132H. The Ki-67 proliferation index was low, around 3%, which supported the diagnosis of a low-grade glioma.

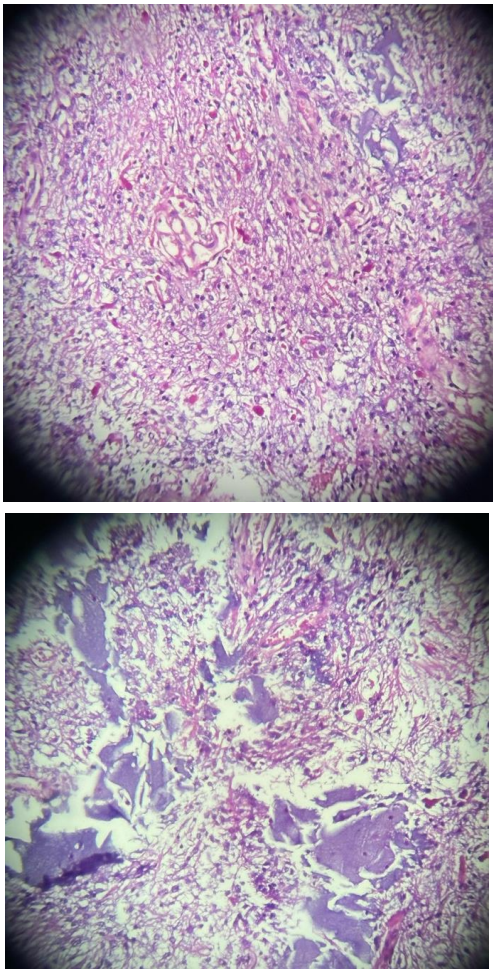

Figure 3 and 4: Histopathological features suggestive of a low-grade glial tumor

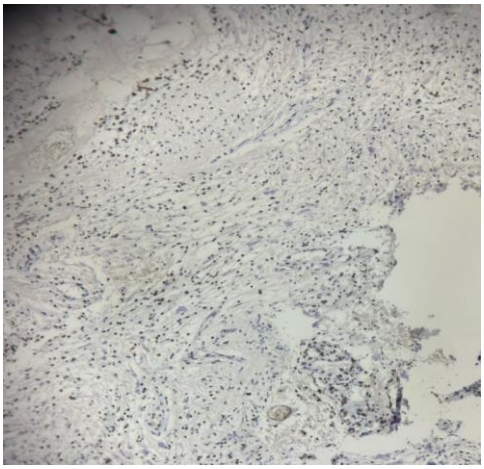

Figure 5: ATRX immunohistochemistry showing nuclear expression in tumor cells

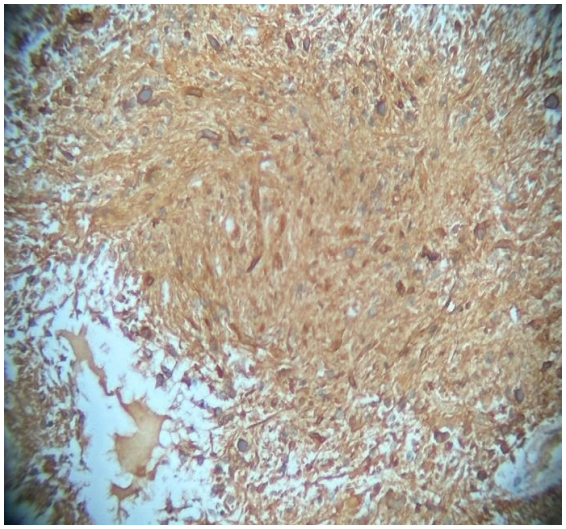

Figure 6: GFAP mmunohistochemistry showing nuclear expression in tumor cells.

A 3 months postoperative MRI showed a marked reduction in tumor size, measuring 40 × 31 × 36 mm. The lesion still had a thin-walled cystic component and a solid nodular part with heterogeneous contrast enhancement. The hydrocephalus had resolved, and there was no longer any evidence of tonsillar herniation. No new vascular anomalies were seen. However, postoperative changes were noted, including meningeal enhancement and infiltration of the posterior cervical muscles at the surgical site.

On clinical examination, she was fully alert (GCS 15/15), with a performance status of 0. She had mild vertigo, occasional tinnitus in the right ear, slight gait deviation to the right, and a subtle coordination deficit on the same side. Muscle strength was preserved in all limbs, reflexes were normal, cranial nerves intact, and there were no sensory or sphincter disturbance

Given the persistence of a residual tumor that could not be completely resected, the patient was started on adjuvant chemotherapy with vincristine and carboplatin, following standard treatment protocols for pediatric and adolescent low-grade gliomas. The aim was to stabilize the disease and delay or avoid radiotherapy, especially in consideration of her young age.

DISCUSSION

Pilocytic astrocytomas are slow-growing, well-defined tumors, most often found in the posterior fossa. Classified as grade I by the World Health Organization, they typically have a benign course, low risk of aggressive progression, and favorable outcomes when completely resected [1].

In children, PAs are among the most common brain tumors, representing 15–25% of pediatric CNS tumors. The cerebellum is the most frequent site, accounting for nearly 60% of cases [2,3]. Radiologically, they often appear as cystic lesions with an enhancing mural nodule and are commonly discovered due to signs of raised intracranial pressure, particularly when obstruction of the fourth ventricle leads to hydrocephalus [4].

Tumor location significantly influences surgical approach and prognosis. Cerebellar hemispheric tumors are generally more accessible and amenable to complete excision. In contrast, vermian tumors, as in our case, are more challenging due to their proximity to the brainstem and deep cerebellar nuclei, which are critical for coordination and balance. This closeness increases the risk of surgical complications and may limit the possibility of total resection [5].

In our patient, the tumor's size and the degree of compression on adjacent structures allowed only partial removal. Such outcomes are common in vermian tumors, especially when associated with brainstem compression or tonsillar herniation [6]. To manage the residual lesion and reduce recurrence risk, adjuvant chemotherapy was initiated postoperatively. This approach aims to achieve disease control while avoiding or delaying radiotherapy, which is particularly important in younger patients to minimize long-term neurological sequelae.

Cerebellar PAs generally have favorable long-term outcomes, but surgical strategy is heavily influenced by their location, size, and proximity to critical structures. When complete resection is not possible, as in our patient, adjuvant chemotherapy becomes the preferred next step. This is now a well-established approach for residual disease, particularly when further surgery poses excessive risk due to the tumor's relationship with vital neurological anatomy.

The vincristine–carboplatin combination is the most widely used regimen in this context. Its efficacy has been confirmed in major cooperative trials, including those by the Children's Oncology Group (COG) and the SIOP-LGG Consortium, showing consistent rates of

disease stabilization or tumor shrinkage. Importantly, these benefits are achieved with side effects that are generally manageable, especially when compared to the potential long-term consequences of radiotherapy [7,8].

This strategy is particularly valuable in young patients, where delaying or avoiding radiotherapy is a priority. Although radiotherapy is effective, its use during brain development can lead to serious late effects, such as neurocognitive decline, hormonal dysfunction, growth retardation, and secondary malignancies [9]. As such, chemotherapy serves as a protective bridge, controlling disease progression while keeping the option of radiotherapy in reserve for when it becomes absolutely necessary

In our patient's case; the decision to initiate chemotherapy aligns well with current clinical guidelines and expert consensus. The goal here is clear: to prevent tumor progression, maintain quality of life, and preserve neurological function, all while postponing the need for radiation therapy.

In recent years, molecular profiling has become increasingly important in the management of low-grade gliomas, including pilocytic astrocytomas. Most of these tumors, particularly in pediatric and adolescent patients, harbor alterations in the MAPK (mitogen-activated protein kinase) pathway, most often involving BRAF gene fusions or mutations [10]. The most common is the KIAA1549–BRAF fusion, a hallmark of classic cerebellar pilocytic astrocytoma, which leads to constitutive BRAF activation and promotes tumor growth in a typically less aggressive manner than BRAF V600E mutations [11].

Identifying these alterations has diagnostic value and potential therapeutic implications, as it opens the door to targeted treatments. In cases where tumors are unresectable or progress after standard therapy, BRAF and MEK inhibitors (dabrafenib, trametinib, or selumetinib) have shown promising results in clinical trials, leading to tumor stabilization or shrinkage with a generally favorable safety profile compared to conventional chemotherapy or radiotherapy [12,13].

Although our patient has not yet undergone molecular profiling, this step could become crucial if her tumor fails to respond adequately to first-line chemotherapy. Detecting a targetable mutation would provide an alternative therapeutic pathway, allowing for a more personalized treatment approach that minimizes long-term toxicity while maintaining effective disease control.

Following the completion of chemotherapy, close and structured surveillance becomes essential in the management of pilocytic astrocytomas, particularly when subtotal resection was performed and residual disease remains. Regular follow-up is typically based on serial MRI scans, with the initial imaging done 6 to 8 weeks after the end of treatment to assess tumor response, and then at progressively longer intervals; usually every 3 to 6 months during the first few years, depending on the clinical course [14].

Following the completion of chemotherapy, close and structured surveillance is essential in managing pilocytic astrocytomas, especially when subtotal resection leaves residual disease. Monitoring typically relies on serial MRI scans, with the first control imaging performed 6 to 8 weeks after treatment, followed by evaluations every 3 to 6 months during the first few years, depending on clinical stability [14].

The objectives of follow-up are detecting early radiological progression and monitoring for delayed treatment-related complications, particularly in adolescents and young adults. Long-term assessments often include neurological examinations, neurocognitive evaluations, and endocrine testing when the hypothalamic–pituitary axis may have been affected by tumor location or prior therapy.

When residual tumor remains stable and the patient is asymptomatic, a "watchful waiting" approach is generally adopted. However, radiological or clinical progression warrants re-evaluation of therapeutic options. Rechallenge with chemotherapy can be considered if there was a good prior response, while molecular profiling becomes critical for identifying targetable mutations in refractory cases.

Radiotherapy, though effective, is generally reserved as a last resort for progressive disease in younger patients due to its potential long-term toxicities. When required, conformal techniques such as proton therapy or IMRT are preferred to limit radiation exposure to surrounding healthy brain tissue [16].

In summary, the post-treatment strategy for cerebellar pilocytic astrocytoma hinges on tumor behavior, age, and symptomatology. Management should remain flexible and tailored, balancing disease control with quality of life and minimizing long-term treatment sequelae.

CONCLUSION

This case highlights the complexity of managing cerebellar pilocytic astrocytoma in adolescents, particularly when gross total resection cannot be achieved. While these tumors are typically slow-growing and associated with favorable long-term outcomes, their location, especially in midline structures like the vermis, can pose significant surgical challenges. In such cases, a multidisciplinary approach combining neurosurgery, oncology, and radiology becomes essential to tailor treatment plans that balance efficacy with the patient's long-term well-being.

The use of vincristine and carboplatin as first-line chemotherapy remains a well-established option for residual disease, especially when radiation therapy is best avoided due to age-related risks. Advances in molecular diagnostics now offer the possibility of targeted therapy in progressive or refractory cases, paving the way for more personalized treatment strategies.

Ultimately, the management of pilocytic astrocytoma extends beyond tumor control. it requires careful, long-term follow-up to monitor for recurrence, treatment side effects, and psychosocial impacts, particularly in adolescent patients. As research continues to refine our understanding of low-grade gliomas, integrating clinical, radiological, and molecular data will be the cornerstone of truly individualized care.

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