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Case Report

# RADICAL RESECTION OF CEREBELLAR JUVENILE PILOCYTIC ASTROCYTOMA - A 22-YEAR SURVIVAL: A CASE REPORT

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## Summary

Juvenile pilocytic astrocytoma (JPA) is a low-grade glioma, a most common astrocytoma in young patients. It is a tumor with relatively well-defined margins. Pilocytic astrocytomas (PA) comprise approximately 5-6% of all gliomas. Gross total resection ensures a radical cure of patients and long-time survival. In the literature, the data on the survival rate of more than 20 years is scarce. A 5.5-year old boy with a history of 3-month complaints of headache, dizziness, and vomiting was diagnosed after CT to harbour a hypodense cerebellar tumor mass, situated in the midline-right hemisphere. The compression of the fourth ventricle resulted in rostral hydrocephalus with transependymal resorption. Within a week, a VP shunt was applied, followed by a radical Nafziger-Town operation. Gross total resection of the tumor was achieved. Profound clinical improvement was observed immediately after the operation. Postoperative CT scans, including the ones 22 years after the operation, remained practically normal. The patient is now 28-year old and is a perspective economist now. He leads a healthy working life. In general, the prognosis is excellent. If the tumor is completely removed by surgery, the chances of being "cured" are very high. Pilocytic astrocytoma has a five-year survival rate in over 96 percent in children and young adults, which is one of the highest survival rates of any brain tumor. However, there is even a small percent possibility for malignant transformation (1-4%).

**Keywords:** pilocytic astrocytoma (PA), juvenile pilocytic astrocytoma (JPA), low-grade glioma

## Introduction

Juvenile pilocytic astrocytoma (JPA) is a low-grade, slow-growing astrocytoma that is most common in young patients. It is classified as WHO grade I tumor in the current 2016 WHO classification of CNS tumours and has a relatively good prognosis. On CT images, a hypodense tumor mass often with cystic lesion and calcifications is detected.

Typically, JPA develops from the cerebellum. Other sites of origin include optic nerve pathways, the brainstem, hypothalamus, or other supratentorial localizations. Raised intracranial pressure is often detected. Light microscopy shows Rosenthal fibers. These astrocytomas emerge and develop in situ and do not spread, being the most non-malignant of all

astrocytomas.

Pilocytic astrocytomas (PA) comprise approximately 5-6% of all gliomas. PAs are typically seen in children and young adults, and their overall incidence is 0.37 per 100,000 persons/year. The median age of patients with a juvenile pilocytic astrocytoma (JPA) ranges between 5 and 14 years. JPA is unusual in infants, and very rare in adults. Approximately one-third of pilocytic astrocytomas involving the optic nerves have associated Neurofibromatosis type 1 (NF1).

The histological entity pilocytic (juvenile) astrocytoma is believed to be a tumor, rather benign in its biological and clinical behavior. Gross total resection ensures a radical cure of the patient and long-time survival.

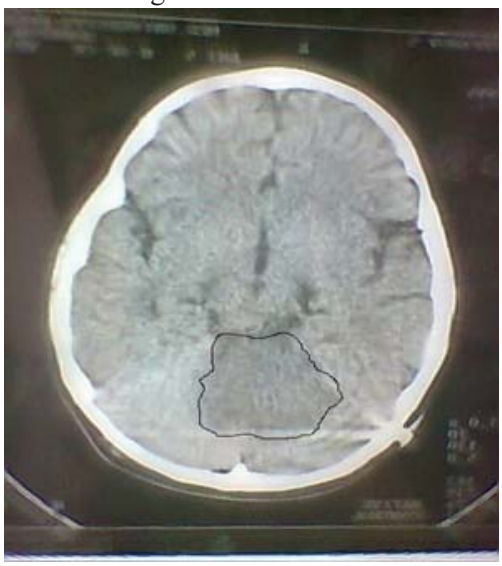

**Figure 1.** Preoperative CT scan (marked is pilocytic astrocytoma)

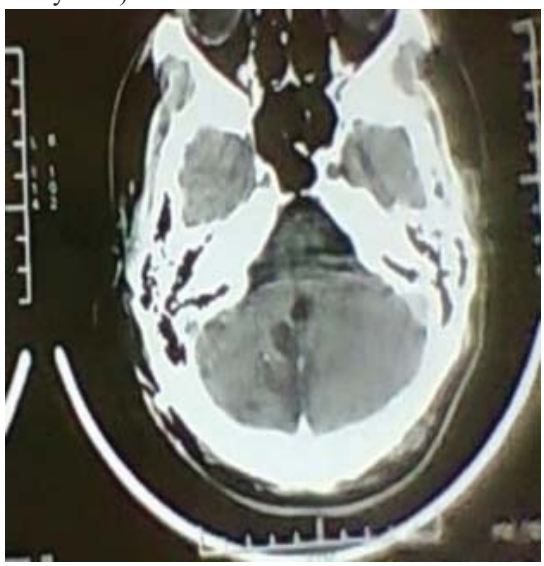

**Figure 2.** Postoperative CT scan (no cerebellar tumor-residual disease is visualized)

## Materials and methods

A 5.5-year old boy presented with a history of headache, dizziness, and vomiting for three months. A CT scan revealed a hypodense cerebellar tumor mass, situated in the midline-right hemisphere. Cystic volumes were not detected. The compression of the fourth ventricle resulted in rostral hydrocephalus with transependymal resorption. Several days after the diagnosis, a VP shunt was applied, followed by radical Nafziger-Town operation using standard craniometric points and a CT scan (topogram). A gross total resection of the tumor was achieved (Figures 1, 2, 3) (1,2).

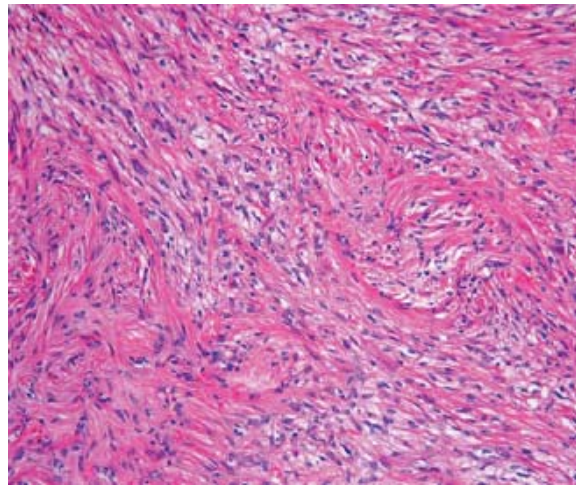

**Figure 3.** Histology: Pilocytic Astrocytoma 20X HE. Bipolar neoplastic cells with elongated hair-like processes that are arranged in parallel bundles and resemble mats of hair. Rosenthal fibers (tapered corkscrew shaped, brightly eosinophilic, hyaline masses), often associated with eosinophilic protein droplets (resembling foamy macrophages); may have microscopically infiltrative margin; mural nodule may be highly vascular; often calcifications. Rarely malignant degeneration with hypercellularity, mitotic figures and necrosis. Characterized by a biphasic pattern with varying proportions of piloid areas alternating with spongy areas. Piloid areas are formed of compacted strongly GFAP+ bipolar cells (with hair-like bipolar processes) associated with Rosenthal fibers. Spongy areas are loosely textured and formed of weakly GFAP+ multipolar cells (protoplasmic astrocytes) associated with microcysts and eosinophilic granular bodies (brightly eosinophilic PAS+ globular aggregates). Vascular changes are common including glomeruloid vascular proliferation and vascular hyalinization. Degenerative changes include hyalinized blood vessels, infarct-like necrosis, degenerative nuclear atypia, calcification, hemosiderin deposits and lymphocytic infiltrate. Leptomeningeal extension is not a sign of malignancy. Some cerebellar tumors show a diffuse growth pattern.

## Results

Profound clinical improvement was observed immediately after the operation. Postoperative CT scans, including the one, performed 22 years after the operation, have been found as practically normal. The shunt was found to function adequately. The 28-year old man is a perspective economist at present. He leads a healthy working life. (Figures 4, 5)

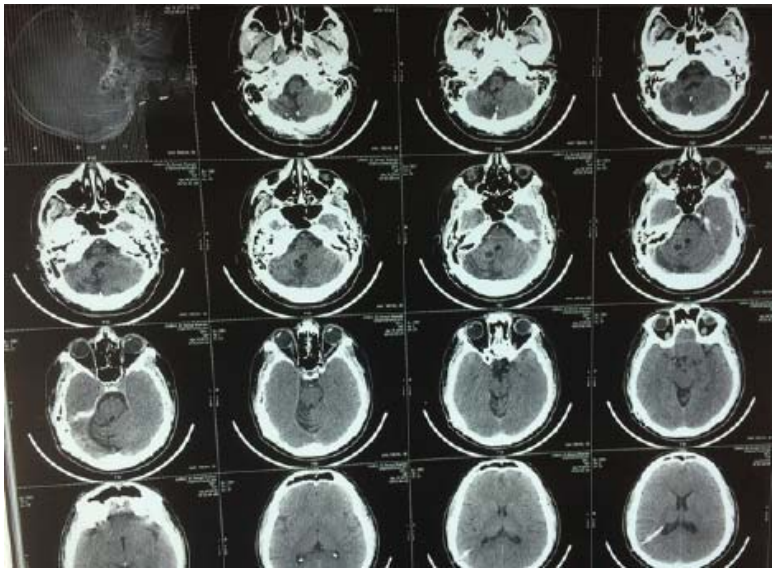

**Figure 4.** Twenty-two years after operation without recurrence; with the clearly discernible VP shunt

**Figure 5.** The young 28 year-old man with his operator, 22 years after the operation

## Discussion

A 5-year survival rate after radical operation for pilocytic astrocytoma is reported to be between 75% and 96% [1, 3]. Therefore, operative radicalism should be the goal of a neurosurgeon in order to expect a long-term survival rate. Nevertheless, we have observed a malignant transformation of pilocytic astrocytoma to glioblastoma multiforme (GBM) localized in the left frontal lobe, histologically verified, three years after radical resection of the benign tumor. Normal neurological and CT status are observed during this period. The reasons for this are still unknown (1). One to four percent of PAs undergo malignant transformation. Histological anaplastic features (pleomorphic nuclei or multinucleated, high mitotic activity, microvascular proliferation, among others), have lately been considered as prognostically important. Such histological findings are more common in older patients [4, 5]. Furthermore, specific signaling pathway alterations within the tumor, give reasons to expect an aggressive behavior even in the absence of anaplasia: mitogen-activating protein kinase (MAPK) pathway alterations: KIAA1549-BRAF (67%), other BRAF or C-RAF fusion (5-10%), BRAF V600E (5-10%), NF1 (5-10%), FGFR1(5%), NRTK1 (<3%), KRAS (<2%), PRPN11(<2%). These newly found facts about PAs help chemotherapists evaluate new modern drugs such as MAPK inhibitors in Phase II or III study trials with promising initial results, such as Selumetinib, Vemurafenib, Dabrafenib, Trametinib combined with or without Carboplatin and Vincristine [4].

Other studies suggest that a better prognosis is seen in children, due to the more favorable tumor location with the possibility for gross total resection [6, 7]. Moreover, a residual tumor should be suppressed by radiation therapy. Such therapy is not recommended for children under the age of 5, because it can affect brain development. However, some authors believe radiation therapy is recommendable in patients with recurrent or progressive pilocytic astrocytoma, localized in critical (eloquent) areas, where additional surgery is not possible.

Some studies have documented that survival decreases with increasing age in patients, especially in adult patients [8].

## Conclusion

In general, the prognosis is excellent. If the tumor is completely removed by surgery, the chances of being cured are very high. Pilocytic astrocytoma has a five-year survival rate in over 96 percent in children and young adults, which is one of the highest survival rates of any brain tumor. However, there is even a small percent possibility for malignant transformation [1-4%)].

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