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Pilocytic astrocytoma: A retrospective study of 32 cases

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ABSTRACT

Pilocytic astrocytoma (PA) is a neoplasia which is considered as a grade I astrocytoma by the World Health Organization (WHO). Its most common location is the cerebellum and it develops during the first two decades of life. Prognosis is mostly excellent if gross-total resection can be achieved, with 10-year survival rates of up to 95%. In rare cases, however, the patient has a bad outcome.

Our aims were to retrospectively describe the clinicopathological features of 32 PAs, and identify factors that may be associated with aggressive behavior.

The study included 21 males and 11 females with a median age of 10.5 years. Tumors demonstrated predilection for infratentorial location (74.9%), especially the cerebellum (59.3%), followed by cerebral ventricles (15.6%), supratentorial location (12.5%) and optic pathway (3.12%). Gross total resection was achieved in 14 tumors only. On histopathology, moderate cellularity (68.7%), microcystic changes (71.9%), Rosenthal fibers (62.5%) and eosinophilic granular bodies (53.2%) were present in the majority of cases. Atypia was present in 62.5% of cases, while endothelial proliferation and necrosis was noted in 3 and 2 cases, respectively. Median follow-up for all patients was 24 months. Four patients died in the postoperative period, one of whom was 62-year-old men and two others had brainstem location or invasion. Recurrence was observed in a 56-year-old patient whom first tumor was locally invasive. The patient died after the second surgery and anaplastic features was found in the recurrent tumor without previous radiotherapy.

PA is a benign tumor, but some clinicopathological factors, such as partial resection, brainstem location and adult age have a worse prognosis.

1. Introduction

Pilocytic astrocytoma (PA) is a rare slowly growing glioma, classified as Grade I by the World Health Organization (WHO) occurring typically in children and young adults [1,2]. Most arise in the cerebellum, particularly in pediatric population; but can also occur in the cerebrum, diencephalon, brainstem, and optic pathway [1,3–5]. Generally, they are regarded as benign tumors in which gross total resection is often curative with a long-term survival greater than 95% [4,6]. Despite the very good prognosis, the outcome is not always favorable, mainly because of surgical morbidity and tumor recurrence. The behavior of this tumor after complete or incomplete resection is unpredictable. As well, pilocytic astrocytomas with anaplastic features are uncommon.

2. Materials and methods

We carried out a retrospective study of 32 cases of pilocytic astrocytoma diagnosed in the Department of Pathology of the Farhat Hached University Hospital, Sousse, and registered in the Cancer Registry of the Center of Tunisia during 27-year period time (January 1984–December 2011). Medical records for each patient were reviewed to determine the sex, age at initial presentation, time to consultation, presenting symptoms and signs and treatment modalities (surgery or surgery and radiation therapy). The extent of resection of resection was estimated by analyzing the surgical reports and the results of postoperative neuroimaging. All results of neuroimaging (computed tomographic and/or magnetic resonance imaging scans) performed during the period of follow-up were obtained. They were judged on the presence or absence of residual or recurring tumor.

Pathology materials from all biopsies and surgical procedures were reviewed to confirm diagnosis using established WHO criteria. Gross characteristics noted included size, consistency, color of surgical specimens, hemorrhagic and/or necrotic foci. Slides

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Table 1
Clinical features of 32 patients with pilocytic astrocytoma.

Patient characteristics	Number of cases (%)	Events, *n* (%)
Age (years)		
0–10	16	2 (12.5)
11–20	9	1 (11.1)
21–30	4	0 (0)
>30	3	2 (66.7)
Median (range)	10.5 (1–62)	
Sex		
Male	21 (65)	5 (23.8)
Female	11 (34)	0 (0)
Time to consultation		
Median (range)	7 months (15–36)	
Type of treatment		
Surgery	21 (95.4)	4 (19)
Surgery + radiation	1 (4.5)	1 (100)
Extent of resection		
Gross total resection	13 (59)	1 (7.7)
Subtotal resection	7 (31.8)	3 (43)
Biopsy	2 (9)	1 (50)
Time to follow-up		
Median (range)	24 months (1–84)	
Outcome		
Died	5	
Alive with tumor	3	
No evidence of disease	11	
Lost to follow-up	13	

were examined for cellular density and types, microcyst formation, Rosenthal fibers, eosinophilic granular bodies, vascularization, vascular hyalinization, chronic inflammation, microcalcification, hemorrhage, endothelial proliferation and necrosis. The evaluation also included an estimation of nuclear atypia and mitotic index. Glial fibrillary acidic protein (GFAP) immunostaining was performed at the time of initial diagnostic workup in a subset of cases.

The clinical status of the patient at the end of the follow-up period was noted. Survival was estimated using Kaplan Meier method. Overall survival (OS) was defined as the time interval from diagnosis to death from any cause or, for patients remaining alive, the time interval from diagnosis to the last follow-up. Disease-free survival (DFS) was defined as the time interval from treatment to the time of disease progression or recurrence, to the last follow-up, or to death occurrence from any cause.

3. Results

3.1. Clinical features

In this study, data pertaining to survival and tumor characteristics were collected from 32 patients (Table 1). The male/female ratio was nearly 2 (21 males/11 females) with a median age at diagnosis of 10.5 years (range 1–62 years). The main presenting symptoms at the time of diagnosis were headache (78%), visual disturbances (43%), vomiting (37%) and gait disturbance (25%). Time to consultation varied from 15 to 36 months (median 7 months). At physical examination, 46% of patients presented with signs of increased intracranial hypertension, 40% with cerebellar syndrome and 28% had papillary edema.

Skull X-ray was carried out in 6 patients. Although not specific, this exam allowed in all cases to detect classic signs of mass effect relative to an intracranial expanding process. More specific preoperative neuroimaging studies were performed in 26 patients and comprised 20 computed tomography (CT) and 12 magnetic resonance imaging (MRI) scans. The locations for the PA tumors are listed in Table 2. In our patient population, infratentorial location (75%) was the most common site, followed by cerebral ventricles (15.6%), supratentorial location (12.5%) and optic pathway (3.1%).

Table 2
Correlation between tumor location and events.

Tumor location	Number of cases (%)	Events, n (%)
Infratentorial	24 (75)	
Cerebellum	19 (59.3)	3 (15.7)
Brainstem	4 (12.5)	2 (50)
Spinal cord	1 (3.12)	
Supratentorial	4 (12.5)	
Hemisphere	3 (9.37)	
Corpus callosum	1 (3.12)	1 (100)
Optic pathway	1 (3.12)	
Cerebral ventricles	5 (15.6)	1 (20)

Cerebellum (59.3%) was the most common infratentorial location. Seventeen PA tumors (65.3%) were cystic with solid mural nodules. The remaining ones were either predominantly cystic (19.2%) or solid (15.4%). Size of tumors varied from 1.5 to 6 cm. Hydrocephalus was detected in 14 cases (53.8%) and 2 patients showed signs of herniation. Information regarding treatment modalities was available in 22 patients. Gross total resection was achieved in 13 tumors (59%); 7 (31.8%) were subtotally resected and 2 patients underwent biopsy only. This later procedure was indicated in the oldest and the youngest patients of our series for neurological status and general condition deterioration. The first one received radiation therapy and died 2 months later. The medical record of the second 1-year-old patient was lost.

3.2. Histopathological features

Histological features are illustrated in Fig. 1. The majority of PAs showed a moderate cellularity (68.7%) and microcystic changes (72%). Rosenthal fibers and eosinophilic granular bodies were present in 62.5% and 53% of tumors, respectively. Vascularization was prominent in 75% of cases and vascular hyalinization observed in 12 cases (37.5%). Endothelial proliferation was present focally in 2 cases and extensively in one case. Three tumors (9.3%) had conspicuous pleomorphic cells and low or moderate degree of atypia was noted in 17 cases (53.1%). Mitotic figures were absent in all tumors. Rare necrotic foci were present in 2 tumors (6.2%). Calcification, inflammation and foci of chronic hemorrhage were rarely seen.

3.3. Follow-up

The mean follow-up period, available in 19 patients, was 29.7 months (median 24 months). Four patients died in the postoperative period; two of them had brainstem tumor location or invasion. The third patient was the oldest one who had not been operated due to his neurological status and general condition deterioration. The last patient died from unrelated septicemia of digestive origin. Tumor progression was observed in one of the remaining patients. This was the second oldest patient of 56 year old, who presented with a right cerebellar tumor and underwent a subtotal resection due to V4 invasion (Fig. 2A). This event supervened 29 months after first surgery. MRI scan showed a recurrent right cerebellar tumor extending to the V4 and accompanied by a second pineal localization (Fig. 2C and D). Pathological exam of surgical material (tumor recurrence) showed more densely cellular compartments with extensive ischemic necrotic foci, endothelial proliferation and variable degree of atypia (Fig. 3). The patient died in the postoperative period.

The Kaplan Meier estimates of DFS and OS, displayed in Fig. 4, show the median DFS was 60.9 months (95% CI, 43.7–78.1 months) and median OS was 61.2 months (95% CI, 44.1–78.2 months).