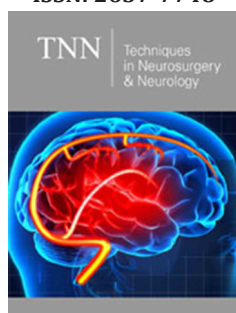


# Management of Tumors of the Cerebellopontine Angle in Developing Countries: Experience of Fann Teaching Hospital in Dakar

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## Abstract

**Introduction:** Cerebellopontine angle (CPA) tumors account for about 5 to 10% of intracranial tumors in the literature. The majority of lesions in this region are benign, so surgery is the main treatment. The literature on the management of these lesions is poor in our context. In this way, we will draw up the epidemiological profile and analyze the diagnostic, therapeutic and evolutionary aspects of this pathology in Dakar.

**Materials and methods:** This is a retrospective study of 51 patients hospitalized in the neurosurgery department of the FANN teaching hospital for the management of the CPA tumors, from January 2007 to December 2018.

**Results:** CPA tumors accounted for 2.8% of intracranial tumors. The average age was 43.4 years and the sex ratio was 0.8. The main signs found were: vestibulo-cochlear syndrome in 66.7% of cases, cerebellar syndrome in 66.7% and HIC syndrome in 56.9% of cases. On imaging, the average size of the tumor was 4.8 cm. The most common tumor was meningioma. Hearing loss and facial paralysis were the most common sequelae. The most common complications are infectious.

**Conclusion:** The tumor pathology of CPA is essentially composed of benign tumors, but of large size in our context making the management of its lesions delicate. Surgery is the main prognostic element found.

**Keywords:** Cerebellopontine angle; Meningioma; Dakar

## Introduction

Tumors of the cerebellopontine angle (CPA) represent about 10% of intracranial tumors in adults and about 1% in the pediatric series. The most common histological types are vestibular schwannoma, meningioma and squamous cyst. These three tumors represent more than 90% of the tumors of the cerebellopontine angle. These are essentially benign lesions [1-4]. Surgical removal is the mainstay of treatment, moving from the era of preservation of the vital prognosis to the era of preservation of the functional prognosis. To date, the literature on CPA tumor surgery is uncommon in our developing countries where access to care is late and the technical platform is not sufficient [5,6]. Thus, our objective is to report our experience on this pathology; we will establish the epidemiological profile and analyze the diagnostic, therapeutic and evolving aspects of this pathology in our context in Senegal.

## Materials and Methods

We conducted a descriptive study, with retrospective data from January 2007 to December 2018, a 12-year period. The study was conducted at the neurosurgical department of the FANN National University Hospital. We have included in the study all patients who have had clinical examination and medical imaging (CT scan or brain MRI) for an accurate topographic diagnosis of cerebellopontine angle tumor. We have excluded incomplete records, tumor lesions of the anterior or middle floor of the skull base extended to the cerebellopontine angle and airway lesions extended to the cerebellopontine angle. We also excluded brainstem and cerebellar lesions spread at the cerebellopontine angle. The parameters evaluated were epidemiological, clinical, paraclinical, therapeutic and evolving. Data analysis was performed on EXCEL 2016 and the statistical analysis software "SPSS statistics 20".

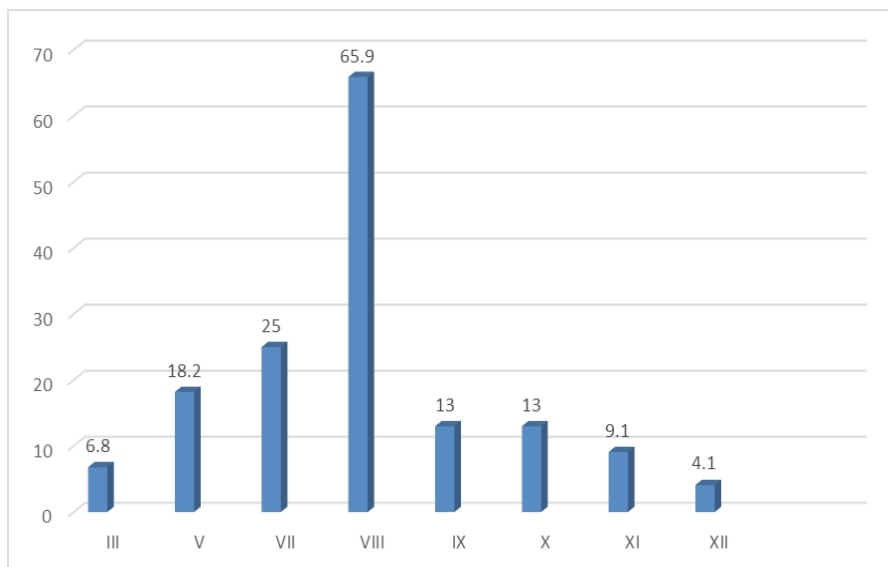
**Results**

**Study population**

During the study period, we reported 51 cases of cerebellopontine angle tumors on 1805 intracranial tumors, representing 2.8% of all intracranial tumors. The mean age of the patients was 43, 4 years, with extremes ranging from 1 year to 77 years. The most represented age group was 41 to 65 years, representing 45.1%. The sex ratio of our series was 0.8.

**Clinical aspects**

The average consultation time was 8.8 months with extremes between 15 days and to 3 years. Headache was the most common cause of consultation, accounting for 82.4% of cases. 45.1% of patients complained of hearing loss. The examination of clinical signs was dominated by cerebellar syndrome (66.7%), vestibulo-cochlear syndrome (66.7%) and intracranial hypertension syndrome (56.9%). Figure 1 illustrates the impairment of the cranial nerves.

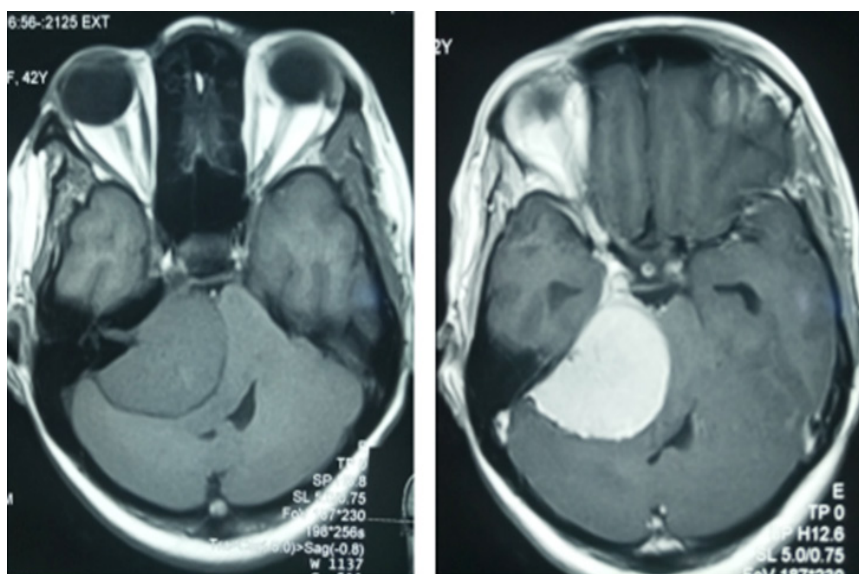


**Figure 1:** Summary percentage rate of cranial nerve damage.

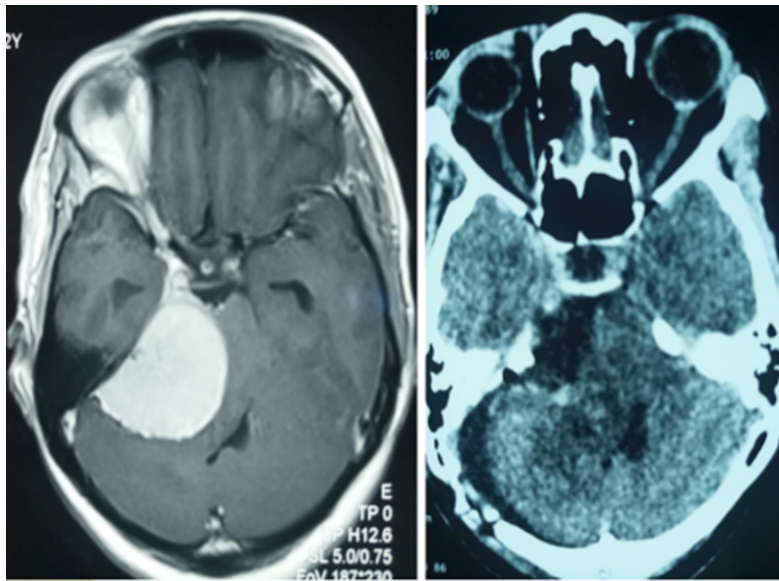
**Paraclinical aspects**

All our patients have had, at least, a morphological exploration. Cerebral CT was performed in 74.5% of patients (Figure 2 & 3) and cerebral MRI was performed in 37.3% of patients and both

investigations were performed in 11.8% of patients. The average tumor size was 5.2cm with extremes from 2cm to 8.9cm. Associated hydrocephalus was found in 72.7% of cases. No neurophysiologic exploration has been carried out.



**Figure 2:** Injected T1 and T1 brain MRI sequences showing a meningioma of the right APC with a significant mass effect on the brain stem and 4th ventricle.



**Figure 3:** Cerebral CT at day 4 post-exeresis of a meningioma of the right CPA.

### Therapeutic aspects

Surgery was performed in 48 patients (94.1%). There were 2 cases of surgical abstention and 1 case of radio clinical surveillance. Gross total resection was performed in 35.3% of the cases, the retro sigmoid approach was the most common, and accounting for 70.6% of cases and 49% of patients received a CSF shunt. The average duration of removal surgery was 4.7 hours with extremes of 3 hours to 8.2 hours. One of our patients received radiotherapy and chemotherapy.

### Evolving aspects

The average length of hospitalization was 27 days with extremes ranging from 4 days to 2.5 months. The average follow-up was 6 months (1 month-4 years). We reported 7 cases of death or 20.5% of cases. There were 4 cases of recurrence, representing 11.8%. The complications (Table 1 & 2) identified in our series were: cranial nerve involvement, infections, CSF leaks, hydrocephalus and a hematoma of the surgical site.

**Table 1:** Distribution according to histological results.

Meningioma	5
squamous cyst	2
medulloblastoma	2
hemangioblastoma	1
schwanoma vestibular	1
high grade glioma	1

**Table 2:** Distribution by post-surgical complications (%).

Cranial nerve injury (VII+++)	26,3%
Infections	26,3%
hydrocephalus	11,8%
CSF leak	8,8%
Hematoma	5,9%

### Discussion

#### Frequency

The literature on CPA tumors as a whole is rare. Most studies focus on specific histological types, especially on vestibular schwannoma and meningioma. These two tumors represent approximately 80% and 10% of the CPA tumors of the different series [7-10] respectively. During a 13-year study period in Nigeria, out of 612 cases of intracranial tumors, [6]. In a South African study, had an annual prevalence of 115 cases of vestibular schwannoma [11]. In this study, there were only 3 cases of black patients, in contrast to the high number of Caucasian patients. This finding is similar to other South African and American studies. It indicates a low prevalence of vestibular schwannoma in black populations [5]. In our study, tumors of CPA represent 2.8% of intracranial tumors. This rate is low compared to the data in the literature. It suggests a low frequency of CPA tumors in our context. Of the 44 patients, collected between 2007 and 2017, 12 cases had an aspect to vestibular schwannoma imaging (27.3%). These results are similar to the South African and Nigerian series. They reflect the low prevalence of schwannoma in sub-Saharan populations. As vestibular schwannoma is the main tumor of CPA, its low prevalence in our population could be an element of explanation for the relative rarity of CPA tumors in our regions.

#### Age and sex

Female predominance is the most common in many series [12,13]. The average age was between the 4th and 5th decade in the different series [9,12,14-16]. We found an average age of 43 years, which is in line with the published data.

#### Clinical aspects

The median consultation time was 44.4 months in the series of 1000 cases of vestibular schwannoma by Samii et al. in Germany [14]. Talfer et al. [16] found an average delay of 26 months in France [16] and 25 months for Voss et al. [13] on a series of 24 meningioma

cases. Our results are in accordance with the literature, the long consultation time is explained by the reason that otological symptomatology rarely leads patients to consult. They come in the stages of neurological signs. In most occidental series, vestibulo-cochlear symptomatology is the most common with hearing loss and vertigo [9,10,12,14-20]. In our series, cerebellar signs, signs of intracranial hypertension and involvement of the long pathways predominated as well as vestibulo-cochlear symptomatology. This is due to the fact that our series is mainly composed of large tumors.

### Paraclinical aspects

As with all current series, all patients have had a CT scan or/and brain MRI to diagnose tumors of the cerebellopontine angle. Both explorations were performed in all patients in the current series. In our series, cerebral CT is the most performed radiological exploration, 77.3% benefited from it. The facility to perform CT scan and its accessible cost compared to MRI makes it the most requested exam in our context. The size of the tumors in our series being large or medium, brain CT had a good sensitivity. However, the gold standard for exploring a suspected pathology of CPA is brain MRI. CPA meningioma was the most common tumor. On the other hand, in the occidental series, there is a clear predominance of vestibular schwannoma in this location. They represent more than 80% of CPA tumors and meningioma represents 5 to 10%. This difference is explained by the fact that our series is mainly composed of sub-Saharan patients. Indeed, in South Africa, of a series of 95 cases of vestibular schwannoma, Seedat et al. [11] found only 3 black patients. In Nigeria, in a series of 612 cases of intracranial tumors, Ohaegbulam et al. [6] had only 3 cases of vestibular schwannoma. The same team had 252 cases of intracranial meningiomas out of 612 cases. Meningioma is reported to represent the most common tumor in their area. Regarding the size of the tumor, we found an average size of 5.2 cm. Only 2 cases out of 51 CPA tumours were less than 3 cm in size. Thus, our series was essentially composed of large or giant tumors. In contrast, in the Occidental series, there is a predominance of stage II and III tumors [14,21-23].

This predominance of large tumors in our series is explained by the delayed diagnosis. Indeed, the lack of a technical platform and specialists in our regions does not allow for early diagnosis. Otological investigations are necessary for the preoperative and postoperative evaluation of patients with APC tumors in the current series [24-29]. The preservation of hearing is one of the challenges of CPA surgery [5]. No patients have had an otological examination in our series. This is a pitfall in the objective evaluation of preoperative and post-operative cochlear involvement.

### Therapeutic aspects

In Algeria, out of a series of 151 unilateral vestibular schwannomas with a major axis in the CPA between 30 and 60mm, all patients underwent retro sigmoid surgery [20]. In South Africa, out of a series of 115 cases, the retro sigmoid pathway was used in 53% of cases, the trans labyrinthine pathway in 8% of cases and the sub-temporal pathway (supra petrous) in 7% of cases [11]. In Germany, Samii and Coll operated on 4000 cases of vestibular schwannoma retro sigmoidally [7]. Results are more dependent on the surgeons'

experience than on the advantages or disadvantages of a particular approach. Excellent results in terms of complete removal rates and functional preservation of facial nerve function have been achieved in hands experienced with each of these approaches.

In the literature, the quality of excision varies according to the histological nature and size of the tumor [22,30]. In the Maghreb, Boublata had 83% of gross total resection on series of vestibular schwannomas [20]. Sekhar and Samii found respectively 99 and 98% gross total resection on schwannoma series [9,29]. The same authors, Sekhar and Samii, found a rate of 56% and 85% of gross total resection, respectively, on CPA meningioma series [10]. Agawar, Ari J. Kane, and Nakamura found 50%, 54% and 85.9% gross total resection, respectively, in contemporary series of CPA meningiomas [15,22,30]. In our series, we only had 35.3% gross total resection. This is due to the fact that most of our series consists of large tumors (greater than 5cm); without intraoperative monitoring, the tumor portions adhering to the cranial nerves and brainstem was not removed. This attitude allows us to reduce the morbidity of the surgery. All our patients have been operated on under an operating microscope, but the lack of drills makes it difficult to properly approach these lesions.

Hydrocephalus was present in 72.7% of our patients at admission. This high frequency is explained by the large number of tumors larger than 4 cm in our series. CSF diversion was performed in 52% of patients. Out of a series of 400 cases of Samii et al. [29] 53 patients had preoperative hydrocephalus, 48 of the 53 patients received removal surgery, 11.3% of patients required additional hydrocephalus treatment [29]. Pirouzmand F et al. [31] of 284 patients with CPA tumors, they found 39 cases of hydrocephalus; only 4 required an emergency diversion [31]. Our high frequency of derivation is explained by the reason that most patients are coming to a table of intracranial hypertension.

Radiosurgery is becoming a non-invasive alternative for most CPA tumors, such as schwannomas and meningiomas smaller than 2cm [25,27,28,32,33]. Not having this technique in Senegal, no patient has had radiosurgery in our series. In our study context, standard radiotherapy is available, it is mainly indicated for malignant tumors. Chemotherapy is primarily indicated as an adjunct to removal surgery for WHO grade III or IV tumors. We had a single case of desmoplastic medulloblastoma that benefited from it.

### Evolutionary aspects

Low or no mortality is found in most current series [12,16]. We had a high rate (20.6%) compared to the results of the literature. This is explained by the high prevalence of large tumors increasing the morbidity and mortality of procedures and by the unavailability of intraoperative monitoring. Patel et al. [12] out of a multicenter series of 14,928 patients in the USA, 10.1% had post-operative facial involvement [12]. In Germany, out of a series of 1000 cases, Samii et al. [29] had 45% facial paralysis [29]. The preservation of the facial nerve is one of the challenges of this surgery, not having monitoring of this nerve intraoperative, our attitude was very conservative.

Indeed, the tumor portion very adherent to the facial nerve were not removed. This attitude allowed us to have 11.8% post-operative involvement. Concerning the damage to the cochlear nerve, it is the most affected nerve preoperatively in the pathology of CPA. In our series, without an objective pre-operative and post-operative evaluation, it is difficult to have an exact assessment of the postoperative outcome. Samii had 3% of meningitis out of a series of 1000 cases [29]. Patel et al. [12] found 0.4% out of 14,928 cases [12]. We had 5.8% of meningitis cases. This is a classic complication of surgery but decreases with the experience of the teams. CSF leaks are a common complication of CPA surgery. But only a few cases require surgical management.

Samii et al. [29] had 9.2% fistulas per 1000 operated patients [29]. Patel et al. had 3.4% fistula in 14,928 patients [12]. Our results are similar to those in the literature. In our study, one patients had a hematoma at the surgical site; and required surgery to evacuate the hematoma. Samii et al. [29] had 22 cases, 15 of which required evacuation [29]. Patel recorded 1.6% hematoma [12]. Hematomas are a serious postoperative complications, they represent, together with diffuse posterior cerebral fossa edema, the main causes of delayed awakening. Thus, a control CT scan is imperative before any delay in waking up.

CPA meningiomas recur more often, compared to vestibular schwannomas. Recurrence is correlated with the quality of meningioma removal (SIMPSONS) and histological grade. Indeed, Agaward et al. [22] on a recent series of 24CPA meningiomas, observed 3 cases of recurrence; all three had had subtotal excision and two had grade II meningioma [22]. In our series, no cases of meningiomas or schwannomas have recurred. However, there is not enough perspective to conclude. In the literature, for WHO grade I meningiomas, the recurrence rate varies between 5 and 20% [22]. On the other hand, the recurrence rates of atypical and anaplastic meningiomas are very high (50 to 95% of cases), regardless of how they are managed, even after radical removal [15,34].

For squamous cysts, the rate of complete excision varies widely in the literature. Yasargil on a series of 22 cases, had total excision in 97% of cases. He observed no recurrence over a follow-up period of about 6 years. Samii et al. [29] out of a series of 40 cases, had 75% complete removal rate. And over the same follow-up period, they had a 7.5% recurrence rate [35,36]. We observed only one case of recurrence of squamous cyst two years after surgery. Medulloblastoma is a very rare tumor in APC. There is currently no consensus on ownership. The evolution is mainly marked by recidivism. Jaiswal et al. [37] in a series of 14 cases, 50% recurred over an average of 6 months of follow-up. In our series, we had two cases, both patients had a recurrence in less than a year, they had a second excision surgery. Like medulloblastoma, the literature on hemangioblastoma in the APC is poor. Out of a series of 23 cases, Cheng et al. [38] had 4 cases of recidivism or 17% [38]. We had a had a case of CPA hemangioblastoma, he recurred after four years of follow-up.

## Limitations of the study

The size of our cohort is not large and the absence of some data makes it difficult to draw convincing conclusions; however, this is a preliminary study. It was also noted that most clinical and paraclinical classifications were not found in the files and ophthalmological and otological examinations were not performed. This pushes us to harmonize the evaluation of patients. We also note that more than half of the patients did not obtain their anatomopathological results. It takes on average 3 months, in our context, to get the results. Not all patients had an otological evaluation and only 22 patients had a control imaging because of the cost of these examinations. Facial nerve monitoring, and neuronavigation is not available in our unit.

## Conclusion

Surgery of tumors of the cerebellopontine angle is always a challenge in our context. The tumor pathology of this region is dominated by benign, but large, tumors. This aspect increases the morbidity of the surgical procedure in general and especially in our context where we do not have intraoperative monitoring of the cranial nerves. This makes the complete removal of these tumors very morbid and affects the quality of treatment provided. The next step would consist in early detection of its tumors, on the one hand, and on the other hand, to have intraoperative monitoring allowing a complete removal of these tumors with reduced morbidity.

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