

Collision Tumour of Cerebellopontine Angle in a Patient without Neurofibromatosis Criteria: Case Report

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Abstract

The coexistence of meningioma and schwannoma as two distinct components within the same tumor is extremely rare. They are usually associated with Type 2 Neurofibromatosis (NF-2) but there are some isolated reports of cases without confirmed diagnosis or in association with cancer and/or post irradiation cancer patients. The authors present a case of a collision tumor of the CPA with a presumed diagnosis of NF-2 and a literature review.

Keywords: Neurofibromatosis; Collision tumors; Meningioma; Schwannoma

Abbreviations: CPA: Cerebellopontine Angle; NF-2: Type 2 Neurofibromatosis; GTR: Gross Total Resection; CN: Cranial Nerve; MRI: Magnetic Resonance Imaging

Introduction

Since the first description by Cushing and Eisenhart in 1938, just over a dozen case reports of mixed schwannomas and meningiomas (both components seen together in the same tumor) arising in patients with NF2 were reported [1]. The simultaneous occurrence of primary brain tumours of different histology in the same anatomical site its' a rarely observed situation, more rarer particularly on the CPA [2,3]. Such tumours of different origin might be considered as collision, concomitant or contiguous tumours and have been mainly reported in patients with NF2, less often in cases without clinical signs of NF2 [4]. The CPA it's the most reported site of these lesions, but there are a fewer reports of collision tumors of the spine [4,5].

There are some genesis theories that these tumors occur due to a total or partial loss of chromosome 22 and an overexpression of epidermal growth factor that induces schwannomas stimulate the growth of meningiomas in a paracrine fashion [6,7].

The radiographic differentiation of schwannomas and meningiomas of the CPA have been well described: *Meningiomas* tend to be broad-based, eccentric to the internal auditory canal, and often have adjacent dural enhancement (dural tails). Vestibular *schwannomas* usually involve and are centered on the internal auditory canal, are more rounded, and rarely have an adjacent dural enhancement. Another distinguishing feature is the hyperostosis seen with 70% of meningiomas, which rarely occurs with vestibular schwannomas [8,9]. The treatment is usually based on the same algorithm used for Schwannomas and meningiomas (GTR and Radiosurgery in specific cases) [8,10].

Case Report

A 54-year-old male with no past personal or familial medical history, presented a 1-year complaint of unilateral vertigo, headache and a 3-month difficulty for swallowing and face numbness. Patient was referred to Neurosurgery outpatients service after investigation in a private practice and on Physical examination at admission presented: alert and oriented, no gait disorders, a grade IV left hemiparesis and a cranial nerves (CN) syndrome as follows: right V1-V3 hypoesthesia; right VII cranial nerve

palsy (House-Brackman 2); right VIII(hypoacusia), IX(decreased right gag reflex), XII(tongue deviation and atrophy).

Investigations

Haemogram; Urine examination, renal function test, and liver

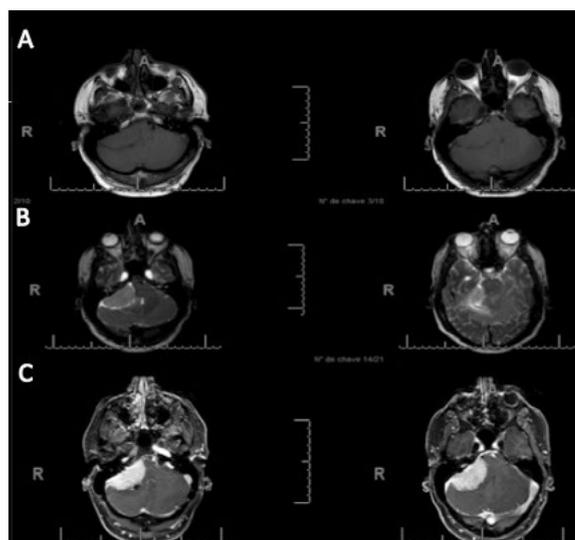


Figure 1: Axial Brain MRI A: T1 weighted image; B: T2 weighted image; C: T1GD weighted image all an extra-axial CPA lesion showing a broad dural base, homogeneous and well circumscribed, with contrast enhancement (C) compressing the brainstem.

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function test were normal. Brain MRI revealed a broad-based lesion on right CPA with brainstem compression and IV ventricle obstruction and secondary Hydrocephalus as shown on Figures 1 and 2.

After case discussion, the authors concluded that the patient had a Meningioma due to the symptoms and image characteristics.

Treatment

The patient underwent surgery, a retrosigmoid craniotomy was performed and Simpson II resection was achieved, during the surgery an incidental discovery (Figure 3), a small schwannoma of the VIII cranial nerve. Conservative conduct was decided due to the small size of the lesion and the possibility for radiosurgery eventually on follow-up. Patient presented on the immediate postoperative worsening of VII nerve palsy (HB 2 to HB 4) and slight improvement of the symptoms of V nerve deficit. A postoperative brain CT showed an adequate resection of the lesion (Figure 4).

The specimen was sent for routine histopathological examination.

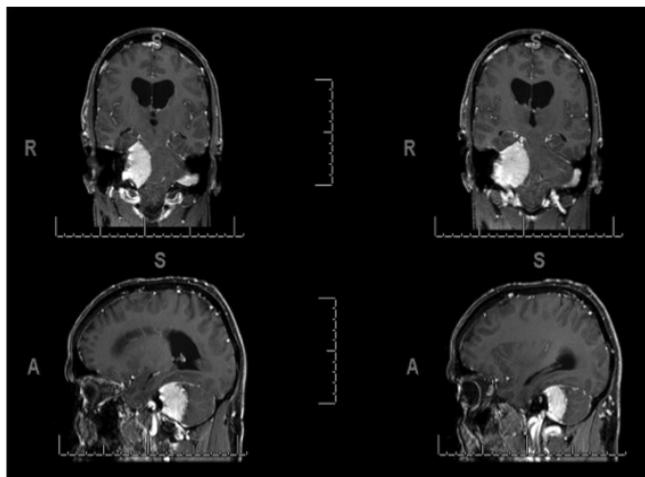


Figure 2: Coronal and Sagittal Brain MRI T1 GD weighted image CPA lesion showing a broad dural base, homogeneous and well circumscribed, with contrast enhancement (C) compressing the brainstem.

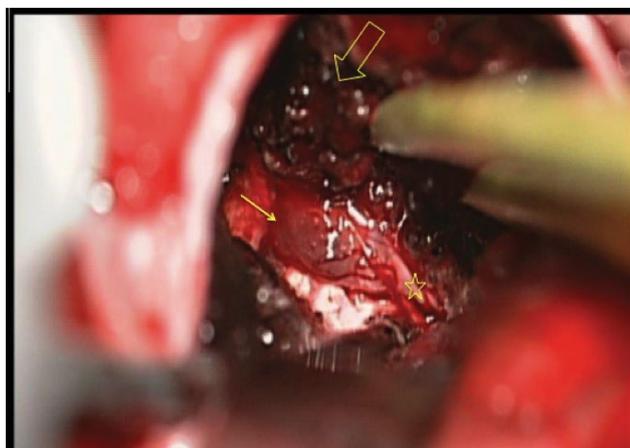


Figure 3: Intraoperative imaging: incidental finding of a VIII cranial nerve Schwannoma (small arrow) and involved VIII cranial nerve (star); partial resected CPA meningioma (big arrow).

Multiple paraffin sections were studied using haematoxylin and eosin. The sections showed a tumor composed by two histological components (Meningioma and Schwannoma).

Outcome and follow-up

On the first month outpatient visit, patient presented improvement of the previous reported immediate facial palsy (from HB 4 to HB 2) and maintained the VIII cranial nerve deficit and awaits for a new audiometry for comparison.

Discussion

When two tumors of discrete pathology occurring simultaneously and in close proximity to each other have been termed 'collision tumors' [10]. Meningioma and vestibular schwannoma (VS) are the most and second most common intracranial benign tumors. In the cerebellopontine angle (CPA), these tumors comprise 6-15% and 80%, respectively, of all tumors [11].

Multiple primary brain tumors are common in patients with phakomatoses, such as NF-2. This disease also known as "multiple inherited schwannomas, meningiomas, and ependymomas syndrome" (MISME syndrome) is an extremely rare (Table 1), inherited autosomal dominant disease characterized by the development of bilateral schwannomas in the region of the vestibular or eighth cranial nerve (in 90% of cases) [4,11].

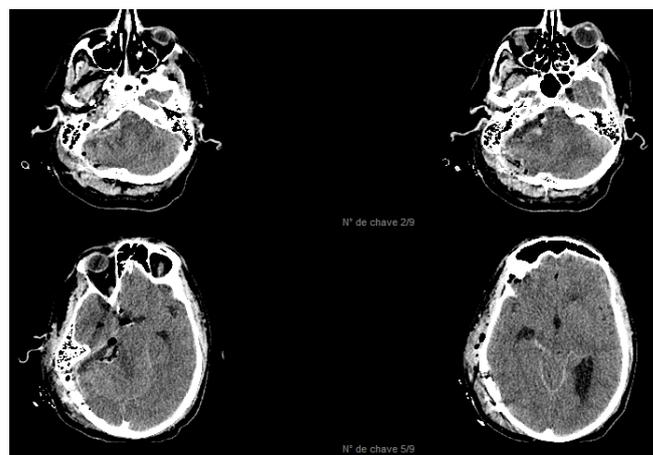


Figure 4: Brain CT: immediate post operative scan revealing adequate resection.

| Author | Year | Tumor side | Age/sex | Radiological differentiation of two distinct tumors on preoperative. Imaging |
|-------------------------|------|------------|---------|--|
| Gardner and Turner [12] | 1939 | Left | 48/F | Not possible (only Roentgenograms available) |
| Thomassin et al. [13] | 1991 | Right | 64/F | Not possible |
| Wilms et al. [14] | 1992 | Right | 47/F | Possible |
| Chandra and Hedge [15] | 2000 | Right | 35/F | Possible |
| Izci et al. [16] | 2007 | Left | 57/F | Possible |
| Grauvogel J et al. [11] | 2010 | Left | 46F | Not possible |
| Present case | 2015 | Right | 53M | Not possible |

Table 1: Summary of previously reported cases of Collision tumors of CPA.

In spite of the overlapping morphological features, cases have been reported, such as the present one, in which the two components of the mixed acoustic tumor show different antigenic profiles, with S-100 protein immunoreactivity being positive in schwannomatosis areas and negative in meningotheliomatous foci [7-16].

Conclusion

In conclusion, meningioma and Vestibular Schwannomas (VS) may very rarely occur coincidentally in the same

CPA. These tumors can usually be safely and completely removed with a single surgical suboccipital approach [11].

Disclosures

The authors declared no potential conflicts of interest.

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