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Review article

Primary cerebellopontine angle melanoma: Case Report and Systematic Review

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ABSTRACT

Introduction: Primary cerebellopontine angle melanoma (PCPAM) is one of the most rare primitive brain tumors. Its management and prognosis are drawn from very few published cases.

Aim: The aim of this study was to provide an approach to help practitioners diagnosing similar cases.

Material and methods: A systematic review was conducted. A PCPAM case is also presented.

Results: The search yielded 13 cases of PCPAM (53% were males) with a median age of 32 years (range 26–56 years). The most frequent symptoms were hearing loss (77%) and ataxia (62%). All underwent neurosurgical removal of the tumor due to worsening of symptoms. Post-surgery follow-up showed that only half of the cases had free-of-disease survival beyond a year. The other half had a poor outcome few months after surgery. We present a 77-years-old female patient with PCPAM with clinical characteristics different from those described in the literature.

Discussion: Results indicate that diagnosis criteria are still lacking specificity. Therefore, clinical features have led clinicians to reconsider the diagnosis more than once. New techniques, such as PET-scan and genetic analysis can greatly assist in the search for the primary tumor. Nowadays, complete resection and radiotherapy are still the gold standard treatment. Prognosis differs between the cases, but age and tumor biology are the main indicators of survival.

Conclusions: We strongly suggest strengthening the surveillance of patients with PCPAM as the management and prognosis differ significantly from those with metastatic melanoma.

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1. Introduction

Primary melanocytic lesions arising from central nervous system (CNS) include localized lesions presented as leptomeninges masses that can be benign tumors (e.g. melanocytoma); melanocytic tumors of intermediate differentiation (MID); and malignant ones, such as melanoma.[1] Primary cerebellopontine angle (CPA) melanoma (PCPAM) only represents 1% of all human melanoma tumors.[2] The incidence has been estimated to be 0.005 cases per 100 000 but we lack stringent diagnostic criteria for its calculation.

CPA is a rare site for tumors as it represents 6%–10% of all intracranial tumors.[3] The most frequent mass lesions of the CPA region are vestibular schwannomas and meningiomas, while melanocytic lesions appear to be a very uncommon finding.[4]

Bailey first described PCPAM in 1948.[5,6] Since then, few cases were reported in the literature. To date, diagnosis remains on exclusion of a primary malignant lesion outside the CNS. Clinical presentation and auxiliary test results may differ among patients receiving this diagnosis. Therefore, the real origin of this tumor is uncertain. The low number of existing published case reports also reveals a lack of evidence-based guidelines for the management of PCPAM.[7]

2. Aim

The aim of this study was to review PCPAM cases and studies published in the literature, in order to guide practitioners managing similar cases. Additionally, we present a case report of a PCPAM, as an instance of this condition.

3. Material and methods

A systematic review of the literature was conducted in Medline, Embase and central databases with the terms 'melanoma,' 'brain tumor,' 'cerebellopontine angle' and their MeSH-term synonyms. The search was restricted to English publications from 1948 to 2013. Non-published studies were collected after tracing down references of the included studies. The selection criteria included PCPAM: (a) case reports; (b) systematic reviews and/or meta-analysis; and (c) clinical trials in which any interventions, drug or surgical, were tested. If multiple studies reported similar results, a selection was made to minimize redundancy. After scrutinized titles and abstracts, we retrieved full text of those that met the selection criteria. Then, we independently extracted the data to a standard form that focused on demographic data, symptoms and signs reported; imaging, surgical and pathological findings; and finally treatment and prognosis reports of the patients with this tumor. We performed quantitative analysis and qualitative analysis depending of the variable nature. We used the Microsoft Excel 2011 version 14.1.0 and STATA 12.0 for both analyses. Figures were designed with Review Manager (RevMan) version 5.2.

Numerical data are presented in percentages, median and 25% and 75% percentiles due to the skewed distribution of the variables and the small sample. Qualitative data are described in Table 1. A meta-analysis was not performed because of the heterogeneity and shortage of reports. This study was conducted according to the recommendations of the Cochrane Collaboration,[8] and is reported following the Prisma Statement.[9]

4. Results

The search yielded 58 studies, and after abstract screening, 13 matched the selection criteria. Then, 2 studies were excluded due to language issues (articles in French). Finally, 11 studies were included involving 13 cases of PCPAM for the review (Fig. 1). Those 13 cases of PCPAM were included in the qualitative and quantitative analysis (Table 1).[10–13]

The median age of subjects was 32 years old (26–56 years old); 54% were male; and the median timing from onset of symptoms to presentation into the hospital was 5 months (2–48 months), with a range of 1 month to 15 years.

The most frequent symptoms and signs presented in patients with PCPAM (Table 2) were unilateral hearing loss (77%), ataxia or gait unsteadiness (62%), unilateral facial palsy (46%) and headache (38%).

Magnetic resonance imaging (MRI) was the preferred imaging technique used in the diagnosis beyond 1990.[14] The most recurrent findings were T_1- and T2-weighted hypointense mass (64% and 55%, respectively).

All the cases underwent neurosurgery due to worsening of symptoms. The most frequent surgical technique to approach the tumor was suboccipital and translabyrinthine craniotomy (23% each). The most frequent intraoperative findings reported the tumor to be a black (69%), highly vascular (62%), and tough (30%) mass. Total resection of the tumor was reported in 46% of the cases.

The pathology findings followed a similar profile, consisting of large, polygonal and pleomorphic cells, arranged in nest or sheets, with vesicular, large and central nuclei and prominent nucleoli. Almost all the tumors showed a high-mitotic rate (only one did not[15]). In total, 54% of reports used positive staining with both HMB45 and S-100 stains as confirmation of the melanocytic origin of the tumor, consistently after 2001 (Table 1).

Methods for the study of a primary origin of the tumor outside the CNS were diverse; 69% reported a dermal and ophthalmologic evaluation, while 23% used one of these imaging techniques: chest X-ray, abdominal ultrasound, PET-scan, CT-scan, Bone-scan.

In 10 cases (77%) patients received radiotherapy after surgery. The other 3 (23%) ones did not: (1) because the tumor had benign behavior[15]; (2) because complete resection of the tumor,[16] and (3) because the patient died 5 days after surgery of post-operative complications.[17] One patient was treated with temozolomide adjunctive to radiotherapy.

The follow-up report showed that 46% of patients had a long free-of-disease survival period, from 1 to 8 years after surgery. Conversely, 46% of the patients died within 1–10 months and had a median survival period of 4 months after hospital discharge.

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