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Gastrointestinal Symptoms—A Rare Complication of Untreated CPA Tumor

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Abstract

Cerebellopontine angle (CPA) lesions account for up to 10% of all intracranial tumors. Most CPA tumors are benign, but can cause nerve damage or compress the surrounding structures if left untreated. The typical presentation is with adult-onset sensorineural hearing loss or non-pulsatile tinnitus. In some patients, this goes unnoticed, and presentation is delayed until the lesion is much larger and presents with symptoms related to mass effect. We present the case study of 63 years old gentleman, who had suspected left CPA lesion on CT head done few years ago for dizziness and left-sided facial numbness. MRI could not be done at that time due to his MRI incompatible pacemaker leading to delay in his management eventually causing loss of patient to the follow up. He later developed progressive difficulty in walking which was initially attributed to as secondary to vasovagal syncope and postural hypotension. He eventually presented to us with intractable nausea and vomiting, worsening headache and ataxia. He had an urgent CT head which showed significant growth in the lesion with compression of the surrounding structures and obstructive hydrocephalus. He was given steroids which improved his nausea and vomiting, followed by undergoing surgery in regional center leading to significant improvement in his gait within few days of surgery. He unfortunately continued to have a degree of ataxia and facial numbness. This case illustrates a rare presentation of CPA tumor with symptoms of nausea and vomiting as a result of mass effect of the growing tumor. In addition, this review also shows the importance of regularly following up the

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patients with suspected CPA lesions on initial scans which will help with identifying the increase in size of lesion promptly and potentially preventing advanced complications of CPA tumors. We suggest regular monitoring of these patients to timely manage the lesion and avoid the potential life-threatening complications.

Keywords

Cerebellopontine Angle (CPA), Vestibulocohlear Nerve (CNV111) Vestibular Schwannoma (VS), Sensorineural Hearing Loss, Facial Nerve (CN VII) Vagus Nerve, Microsurgery, Gamma Knife Surgery (GKS, Hydrocephalus)

1. Introduction

A vestibular schwannoma [1], (also known as acoustic neuroma) is one of the CPA tumors which is a benign, usually slow-growing tumor that arises from the vestibulocohlear nerve (CNV111) [2]. The tumor develops from an overproduction of Schwann cells. As the vestibular schwannoma grows, it affects the hearing and balance, usually causing unilateral hearing loss, tinnitus and dizziness/loss of balance. As the tumor grows further, it can affect the ipsilateral trigeminal nerve (CN V) and facial nerve (CN VII) [3], causing facial numbness and facial weakness/paralysis respectively. If the tumor becomes large, it will eventually press against nearby brain structures such as the brainstem, the cerebellum and ventricular system, causing life-threatening complications. Schwannomas are the primary lesions of cranial nerves involving trigeminal and facial nerves, accounting for 75 to 85 percent of all CPA tumors. Intractable nausea and vomiting in advanced cases can be secondary to obstructive hydrocephalus or involvement of vagus nerve (CN X) which is extremely rare.

2. Case Presentation

A 63-year-old gentleman was admitted to our hospital in August 2022 with intractable nausea, vomiting and worsening headache for last three weeks. He had past medical history of essential hypertension, postural hypotension, vasovagal syncope and atrioventricular heart block with a pace-maker in-situ. During the last few years, he had been suffering from left sided facial numbness, dizziness and unsteadiness which hindered his ability to walk without using stick. He was reviewed by ear, nose and throat (ENT) during that time and had CT head which showed suspected lesion in the left cerebellopontine angle, but the requested MRI was cancelled due to presence of pace maker which was incompatible with MRI. The outcome of MDT discussion at that time which included neurosurgeon, ENT specialist, cardiologist and radiologist was to observe the lesion.

On presentation, his blood pressure was 121/65, pulse rate 109 bpm, respiratory rate 19/m and temperature was 36.7C. His neurological examination revealed left side lower motor neuron CN VII palsy and ataxic gait. The remainder

of the physical examination was normal apart from having mild dehydration. He underwent CT head which revealed heterogeneous hypo-dense lesion with enhancing component in left CPA, measuring 3.7/4/3.6 cm arising from the left auditory canal, compressing the adjacent cerebellar hemisphere and affecting the fourth ventricle with obstructive hydrocephalous [4]. MRI could not be done due to the presence of incompatible pace maker. After urgent discussion with neurosurgical team, the patient was commenced on steroid which showed significant improvement in his gastrointestinal symptoms, following initiation of therapy. He was therefore transferred to neurosurgical unit where he was treated surgically.

The patient was able to walk independently without stick only two days following the operation, corresponding to significant improvement in his gait but the left CN VII damage did not show any improvement. He was referred for physiotherapy and was planned to be followed up by ENT team. The result of biopsy confirmed the histopathological diagnosis of schwannoma (**Figure 1** and **Figure 2**).

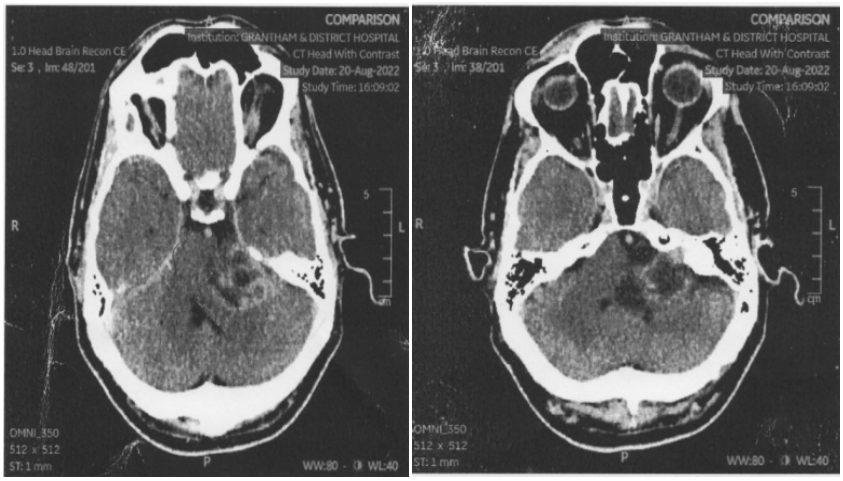

Figure 1. CT head of the patient showing left CPA mass prior surgery.

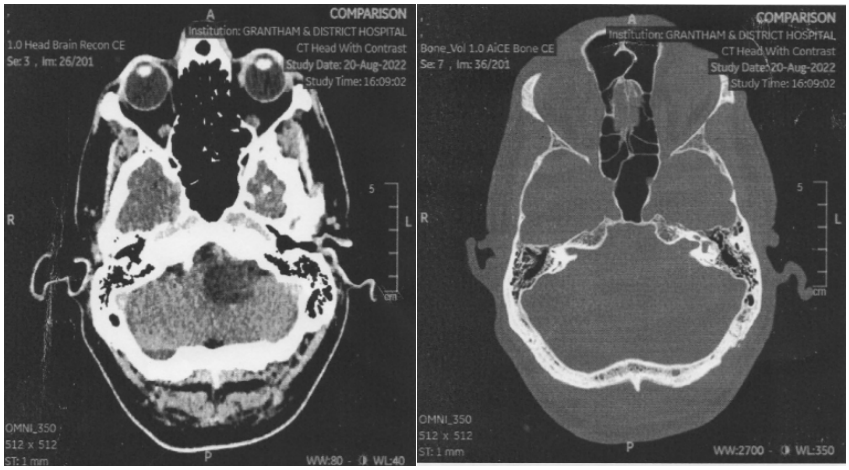

Figure 2. CT head of the patient after surgical removal of the tumor.

3. Discussion

Cerebellopontine angle (CPA) is a triangular space in the posterior cranial fossa that is bounded by the tentorium superiorly, brainstem posteromedially and petrous part of temporal bone posterolaterally. It is an important landmark anatomically and clinically as it is occupied by the CPA cistern, which houses the cranial nerves V, VI, VII, and VIII along with the anterior inferior cerebellar artery.

Our patient had vestibular schwannoma which are benign tumors and usually arise from the intracanalicular segment of the vestibular portion of the vestibulocochlear nerve (CN VIII). They were classically described as originating near the transition zone between glial and Schwann cells but contemporary data suggests they can originate at any point along the nerve. In over 90% of cases, these tumors arise from the inferior division of the vestibular nerve as was the case in our patient). Less than 5% cases arise from the cochlear component of the vestibulocochlear nerve (CN VIII). Non-schwannoma tumors are; meningiomas which account up to 10% to 15%, whereas epidermoids make up 7% to 8% of all CPA tumors. Other less common tumors that roughly contribute 1% are; arachnoids cysts, lipomas and metastatic lesions [5].

The most common presenting symptoms of lesions involving the CPA include hearing loss, tinnitus, dizziness, vertigo, headaches, and gait dysfunction. Hearing loss is mostly unilateral sensorineural and is due to the involvement of the cochlear nerve. Other cranial nerve deficits, brainstem compression symptoms, and hydrocephalus can also be seen with larger tumors compressing these structures. These symptoms are most commonly present between the fourth and sixth decade of life. However, those associated with neurofibromatosis (NF) type 2 [6]; the most common presentation is bilateral acoustic neuromas in younger patients with a positive family history, a condition results from a mutation at the chromosome 22q12.

MRI is the gold standard for diagnosis but unfortunately we could not do it because patient had MRI incompatible pacemaker leading to delay in management of "suspected" CPA tumor on initial CT head. Cerebral angiography [7] as a diagnostic tool is used when involvement of a large vessel is suspected or preoperative embolization is required. It is employed to assess patency in vessels encased by tumor. This was not done in our patient due to absence of any indication.

Current consensus on optimal treatment of vestibular schwannoma remains poorly established; the options include observation, stereotactic radiosurgery [8], microsurgical resection, medical therapy, or a combination of these. The Treatment should be individualized and incorporate the patient clinical features and tumor-specific characteristics known to affect outcome. The treatment should maximize tumor control and minimize functional deficit. Unfortunately our patient had advanced disease due to delay in diagnoses which lead to persistent left sided facial nerve palsy and residual ataxia. This again shows the importance of

regular monitoring and early intervention for any new symptoms.

In large vestibular schwannoma (VS), microsurgery [9] is the main treatment option, and complete resection is considered the primary goal. However, previous studies have documented suboptimal facial nerve outcomes in patients who undergo complete resection of large VSs. Subtotal resection is likely to reduce the risk of facial nerve injury but increases the risk of lesion re-growth. In this situation, Gamma Knife surgery (GKS [10] can be performed to achieve long-term growth control of residual VS after incomplete resection.

On reviewing the literature, we found this retrospective study in which the authors report on the results in patients treated using planned subtotal resection followed by GKS with special attention to volumetric growth, control rate, and symptoms. Case studies of fifty patients were reviewed retrospectively between 2002 and 2009 with median follow up of 33.8 months. There was good outcome with clinical control achieved in 92% and radiological control in 90%. One year after radiosurgery, facial nerve function was good in 94% of the patients. One of the two patients who underwent surgery to preserve hearing maintained serviceable hearing after resection followed by GKS. Considering the good tumor growth control and facial nerve function preservation as well as the possibility of preserving serviceable hearing and the low number of complications, subtotal resection followed by GKS can be the treatment option of choice for large VSs.

4. Conclusion

This patient case illustrates large intracranial space occupying lesion complicated by hydrocephalus in a 63-year-old man with background chronic vasovagal syncope, with a three-year history of symptoms and signs suggestive of CPA tumor, with delay in management partly due to pace maker un-compatible with MRI scanning eventually resulting in loss of the patient to follow up. Physicians must be more alert and aware about this rare but potentially life-threatening complication of slowly growing tumor with unusual presentation such as nausea and vomiting, which is usually difficult to manage. It is also suggested that such patients be reviewed annually or bi-annually to spot any new clinical picture that necessitates urgent specialist intervention or radiological examination. This condition needs multi-disciplinary approach from the time of initial diagnosis involving relevant specialists like neurosurgeon, neurologist, ENT specialist, and radiologist which will help in formulating clear management plan resulting in avoidance of complications and potentially reducing the long-term morbidity and mortality, a situation which require further researches and studies.

5. Disclosures

Human subject: consent was obtained and waived by all participants in this study. Conflict of interest: in the compliance with the ICMJE uniform disclosure form, all the authors have declared the following; payment/service information, all the authors have declared that no financial support was received from any

organization for the submitted work. Financial relationship, all the authors have declared that no financial relationship at present or the past with any organization that might have any interest in the submitted work. Other relationships, all the authors have declared that there is no other relationship or activities that could appear to have influenced the submitted work.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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