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Case Presentation

Over Thirty Years of Experiences in Management the Rare Cases of Glossopharyngeal Neurinoma, Case Series and Literatures Review

Farajirad M. [1*], Mohazzab Torabi S. [1], and Farajirad E. [2]

[1] Department of Neurosurgery, Mashhad University of Medical Sciences, Iran

[2] Resident Physician in Adult Neurology University of Toronto, Canada.

Corresponding Authors: **Farajirad M.**, Department of Neurosurgery, Mashhad University of Medical Sciences, Iran.

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Abstract

Glossopharyngeal neurinomas are rare neurogenic tumors that arise from the glossopharyngeal nerve. They account for less than 1% of all head and neck tumors and can result in significant morbidity if left untreated. The present review aims to provide an over thirty years experiences of the clinical presentation, diagnosis, and management of glossopharyngeal neurinomas.
We present 15 cases of glossopharyngeal neurinoma who were underwent surgical resection in Ghaem faculty of medicine, Mashhad, Iran from 1991 to 2022. Age distribution was between 24 years old to 63 years old. One case wase dumble shape with concurrent intra-extra cranial invasion and other were just intra-cranial. On primary neurological assessment, all patient were totally alert and 14 cases were complaint hearing loss,8 patient facial palsy, three cases hoarseness, three cases gag reflex dysfunction and two cases of pain in the tongue or throat (glossodynia). All patients were undergoing surgery via retrosigmoid approaches with gross total resection which was confirmed with post operative MRI in all cases.
In conclusion, glossopharyngeal neurinomas are rare but important tumors to be aware of in neurosurgery practice. Early diagnosis and treatment are crucial for an optimal outcome, and the combination of surgical resection and postoperative radiotherapy appears to be the most effective management strategy.

Key words: glossopharyngeal neurinomas; cerebellopontine angle; surgery; radiosurgery

Introduction

A glossopharyngeal neurinoma is a rare type of benign tumor that develops on the ninth cranial nerve, also known as the glossopharyngeal nerve [1]. This nerve is responsible for controlling the muscles of the tongue and throat, as well as providing sensation to the back of the tongue and pharynx. Symptoms of a glossopharyngeal neurinoma may include difficulty swallowing, pain in the tongue or throat, and difficulty speaking. Treatment options include surgical removal of the tumor and radiation therapy [2]. The prevalence and incidence of glossopharyngeal neurinomas is considered to be quite rare. According to a study published in the Journal of Neurosurgery, the incidence of glossopharyngeal neurinomas is estimated to be between 0.1 and 0.5 cases per 100,000 people [3]. Although it is considered a rare tumor, it should be considered in the differential diagnosis of patients with symptoms such as otalgia, dysphagia, or dysarthria. It is typically diagnosed in middle-aged adults and is more common in men than women [3]. The majority of cases are sporadic, however, familial cases of glossopharyngeal neurinomas have also been reported.

Glossopharyngeal neurinomas are benign tumors that typically arise from the schwann cells, which are the cells that make up the myelin sheath that surrounds nerve fibers [4]. They are slow-growing tumors that can compress or damage the surrounding structures, such as the VII, VIII, X nerves. They can be classified into two types: intracranial or extracranial. Intracranial tumors are located inside the skull and typically compress the nerve within the skull. Extracranial tumors are located outside the skull and can compress the nerve as it exits the skull. Both types can cause similar symptoms, but the treatment options and prognosis can be different [5]. Pathologically, these tumors are composed of spindle-shaped cells that are arranged in a pattern called Verocay bodies. They are usually encapsulated and well-circumscribed. They are usually composed of bland cells with minimal atypia and mitotic activity. The tumor cells are positive for S100 and glial fibrillary acidic protein (GFAP) [6]. The diagnosis is usually made by imaging studies such as magnetic resonance imaging (MRI) and biopsy. In some cases, a fine-needle aspiration (FNA) can be used to diagnose the tumor, but a biopsy is typically needed for definitive diagnosis. Here, we will present over thirty years of experiences, clinical manifestation, diagnostic methods and treatment of glossopharyngeal neurinomas as a rare brain tumor.

Case Presentation

We present 15 cases of glossopharyngeal neurinoma who were underwent surgical resection in Ghaem faculty of medicine, Mashhad, Iran from 1991 to 2021. Pre-operational diagnosis was acoustic neurinoma in 11

cases, jugular glomus in one patient and three cases of other cranial nerve neurinoma. Age distribution was between 24 years old to 63 years old. one case were dumble shape with concurrent intra-extra cranial invasion and other were just intra-cranial. Right CPA tumor was seen 8 patients and other were on left side. Sign of hydrocephalous in3 patients no invasion to brain stem were seen in all patients. On primary neurological assessment, all patient were totally alert and 14 cases were complaint hearing loss, 8 patient facial palsy, six cases hoarseness, 5 cases gag reflex dysfunction and two cases of pain in the tongue or throat (glossodynia). All patients were undergoing surgery via retrosigmoid approaches. Gross total resection was confirmed with post operative MRI in all cases. Despite two transient hoarseness which had been resolved in three months post-operatively, no other significant complications were observed. Patients' demographic data, clinical presentation, tumor side, surgical approaches and complication were demonstrated in (table 1). In addition, Imaging of one patient with Glossopharyngeal Neurinoma was shown in (Figure 1).

	Sex	Age	Tumor side	Hearing loss	Hoarseness	Gag dysfunction	Glossodynia	Facial palsy	Complication
1	Female	25	Right	+	+	-	-	-	-
2	Female	31	Left	-	-	+	-	+	-
3	Male	41	right	+	-	-	-	-	-
4	Female	46	left	+	-	+	-	-	Short term Hoarseness
5	Male	37	right	+	+	-	+	-	-
6	Female	40	right	+	-	-	-	+	-
7	Female	32	left	+	+	-	+	+	Short term Hoarseness
8	Male	49	right	+	-	-	-	-	-
9	Male	44	left	+	-	+	-	+	-
10	Female	24	right	+	-	-	-	+	CSF cyst
11	Female	32	right	+	+	-	-	-	-
12	Male	48	left	+	+	-	-	+	-
13	Female	63	right	+	+	+	-	-	-
14	Female	24	left	+	-	-	-	+	-
15	Male	58	left	+	-	+	-	+	Short term Hoarseness

Table 1: Demographic and clinical data of patients

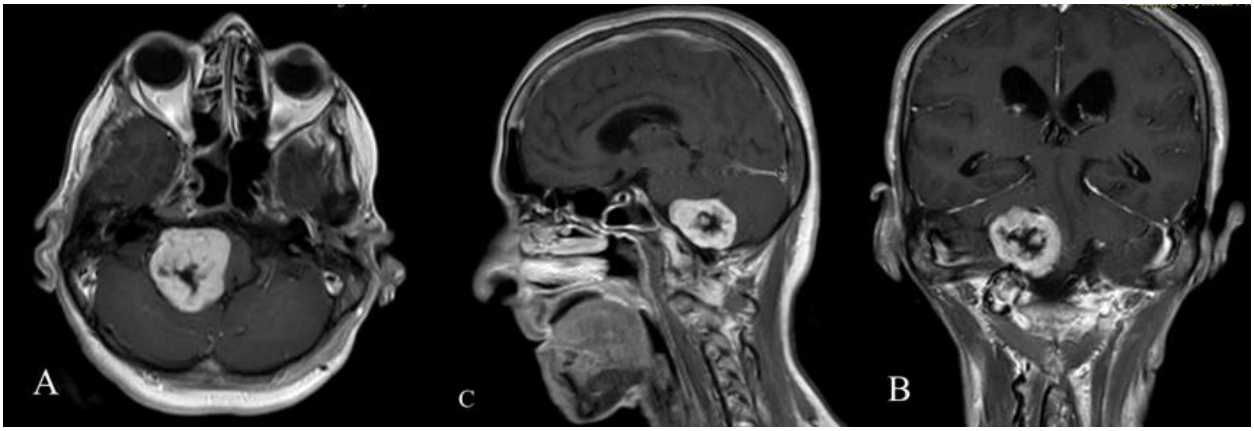

Figure 1: Axial (**A**), sagittal (**B**) and coronal (**C**) T1+ Gad sequences demonstrated hyper signal Glossopharyngeal Neurinoma in right CPA

Discussions

Glossopharyngeal neurinomas, also known as glossopharyngeal schwannomas, are rare neurogenic tumors that arise from the glossopharyngeal nerve. They account for less than 1% of all head and neck tumors and can result in significant morbidity if left untreated [7]. The prevalence of glossopharyngeal neurinomas does not appear to be affected by sex, with studies reporting similar incidence rates in both men and women [8]. In a study of epidemiology of glossopharyngeal neurinomas, the male to female ratio was 1:1 [9]. As we showed, hearing loss is a most common symptoms of this tumor. These findings suggest that there is no significant difference in the incidence of glossopharyngeal neurinomas between men and women [9]. Clinical presentation of glossopharyngeal neurinomas is typically nonspecific and can include symptoms such as hearing loss, facial palsy, difficulty swallowing (dysphagia), hoarseness (dysphonia), and pain in the tongue or throat (glossodynia). In addition, some patients may present with Horner's syndrome, which is characterized by drooping of the eyelid, constriction of the pupil, and decreased sweating on the affected side of the face [10]. The presence of Horner's syndrome is indicative of involvement of the sympathetic fibers of the glossopharyngeal nerve [11, 12]. Some patients may also present with a combination of the aforementioned symptoms.

The preoperative diagnosis of glossopharyngeal neurinomas from acoustic neurinoma is almost impossible and most of the reported cases are identified during the operation. usually, the image findings are similar to those of acoustic neurinomas. These imaging studies can help to identify the location, size, and extent of the tumor, as well as its

relationship to surrounding structures. In addition, these imaging modalities can help to differentiate neurinomas from other tumors that may present with similar symptoms, such as a jugular foramen schwannoma or a hypopharyngeal carcinoma.

The prognosis of glossopharyngeal neurinomas is generally good, with a high rate of complete resection and preservation of glossopharyngeal nerve function [13]. However, the long-term outcome can be affected by the size and location of the tumor, as well as the patient's overall health and medical history [14]. In addition to surgical resection, there have been reports of successful treatment of glossopharyngeal neurinomas with radiosurgery, particularly in patients who are not suitable candidates for open surgery [15]. This treatment modality has been shown to be effective in controlling tumor growth and preserving glossopharyngeal nerve function in some patients. The mainstay of treatment for glossopharyngeal neurinomas is surgical resection. The aim of surgery is to completely remove the neoplasm while preserving the function of the glossopharyngeal nerve. Furthermore, we will discuss about some researches that the surgery was successful and the patient experienced improvement in his symptoms after the operation [16]. The surgical approach will depend on the location and size of the tumor, as well as the patient's overall health and medical history. In rare cases, postoperative radiotherapy may be necessary to prevent recurrence. In the case report published in the Journal of Clinical Neuroscience in 2019, the patient was a 61-year-old man who presented with difficulty swallowing and pain in the tongue and throat [17]. In the case report published in the Journal of Craniofacial Surgery in 2020, the patient was a 55-year-old man who presented with difficulty speaking and swallowing. He was diagnosed with a glossopharyngeal neurinoma and underwent surgery to remove the tumor [18]. In the case report published in the journal of Otolaryngology-Head and Neck Surgery in 2018, the patient was a 55-year-old man who presented with difficulty swallowing and pain in the tongue. He was diagnosed with a glossopharyngeal neurinoma and underwent surgery to remove the tumor. The surgery was successful and the patient experienced improvement in his symptoms after the operation [19]. A case report published in the Journal of Medical Case Reports in 2015 describes the case of a 45-year-old woman who presented with difficulty swallowing and a hoarse voice. She was diagnosed with a glossopharyngeal neurinoma and underwent surgery to remove the tumor. The surgery was successful and the patient experienced improvement in her symptoms after the operation. Additionally, the report states that the patient received postoperative radiotherapy, and the patient's symptoms were resolved [20].

We mentioned three patients who had experienced transient hoarseness which was improved within three months. Unfortunately, it is difficult to provide specific percentages for complications of surgery for glossopharyngeal neurinomas, as the literature on this topic is limited and the studies that have been conducted have relatively small sample sizes [16]. However, some studies have reported on the incidence of specific complications in their patient populations.

Here are a few examples of complication. patients with glossopharyngeal neurinomas, 37.5% of patients experienced some degree of glossopharyngeal nerve injury [21]. Another study reported no cases of major haemorrhage during surgery, but one patient experienced a minor bleed that required transfusion [22]. Another literature patients with glossopharyngeal neurinomas, 6.25% of patients experienced recurrence of their tumor after surgery [23]. It's important to note that these studies are not representative of all cases, and the complication rate may vary depending on the tumor size, location, and surgical method and experience of surgeon.

Additionally, the report states that the patient received postoperative radiotherapy, and the patient's symptoms were resolved. Radiosurgery is a non-invasive method that uses precisely focused radiation beams to target the tumor while minimizing damage to surrounding healthy tissue. Case reports have shown that surgery and radiosurgery can be effective in treating glossopharyngeal neurinomas. For example, a case report by Park et al. described the successful use of radiosurgery in a patient with a large glossopharyngeal neurinoma that was not amenable to open surgery [24]. The patient had a good outcome with preservation of glossopharyngeal nerve function and no recurrence at a 5-year follow-up. Another case report by Zhao et al. described a patient with a glossopharyngeal neurinoma who underwent open surgery and had an excellent outcome with preservation of glossopharyngeal nerve function and no recurrence at a 5-year follow-up [25]. In addition to surgical resection and radiosurgery, there have also been reports of successful treatment of glossopharyngeal neurinomas. In our patients who underwent surgery, no recurrence was seen until the time of writing this article.

Conclusion:

In conclusion, glossopharyngeal neurinomas are rare but important neurogenic tumors that can result in significant morbidity if left untreated. The corner stone of treatment is surgical resection, with preservation of glossopharyngeal nerve function being the primary goal. Radiosurgery and a combination of surgery and adjuvant radiotherapy can also be effective treatment options, particularly in patients who are not suitable candidates for open surgery. Close monitoring and follow-up are essential to detect any recurrences or complications. It is important for neurosurgeons to be familiar with the clinical presentation, diagnostic evaluation, and management options for glossopharyngeal neurinomas in order to provide optimal care for patients with this condition.

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