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RESEARCH ARTICLE

HYPOGLOSSAL SCHWANNOMA: A CASE REPORT

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ABSTRACT

Hypoglossal schwannoma is a rarely encountered skull base lesion that has generally both intracranial and extracranial parts. This article reports a case of hypoglossal schwannoma that is purely extracranial and presents itself with right-sided hypoglossal palsy and a right-sided neck mass. Magnetic resonance imaging is very important, delineated a mass at the skull base that consisted of both cystic and solid components, consistent with a nerve sheath tumor but not done in this case because of lack of means. Cervical approach was chosen, and the mass was completely removed. Postoperatively, the patient developed hypoglossal nerve palsy because the hypoglossal nerve was sacrificed. Hypoglossal schwannomas are briefly discussed with a literature review.

INTRODUCTION

Schwannomas, which originate from Schwann cells of peripheral nerves, are rare and constitute 5% of all benign soft-tissue tumors (Biswas, 2007 and Ho, 2005). Most of the cranial nerve schwannomas arise from the sensory nerves; hence, schwannomas originating from motor cranial nerves such as the hypoglossal nerve are even rarer.3 About 45% of schwannomas are encountered in the head and neck region (Biswas, 2007). They are named according to the nerve of origin (Kuo *et al*., 2008). The signs and symptoms also depend on the nerve of origin and the size of the tumor as well. Schwannoma is considered to be the most common solitary neurogenic tumor in the neck, with an age range between 20 and 50 years (Kuo, 2008). A case of hypoglossal schwannoma presenting with cervical mass and tongue palsy after surgery is given in this report.

Case Report

A 60 year-old female presented to the ENT department with a two-year history of a progressively enlarging right-sided neck mass. Symptoms on presentation were limited to neck discomfort and mild dysphagia. The patient had an otherwise unremarkable past medical history. On physical examination, a large, mobile, soft level II neck mass was identified. A comprehensive cranial nerve examination was normal. Tongue examination and mobility was normal (Figure 1).

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An CT- Scann of the neck demonstrated a 3.3x 2.3 cm heterogeneous mass at the level of right parapharyngeal space, near the carotid bifurcation (Figure 2). MRI has been indicated but the patient doesn't have the means to pay it. So only CT scan has done. The patient was operated on under general anesthesia, and the cervical approach by Sebileau's incision was chosen because no intracranial extension was noted. Intraoperatively, a grey, gelatinous appearing mass was identified at the carotid bifurcation, and as originating from the proximal segment of hypoglossal nerve, arriving until the right lobe of the thyroid gland which is conserved. The mass had well-defined borders and was fragile on palpation. It was removed en bloc, sacrificing the hypoglossal nerve (Figure 3). The histopathologic diagnosis of the resected mass was schwannoma. The patient had developed right-sided hypoglossal nerve paralysis soon after the surgery, which was more evident on protrusion of the tongue (Figure 4).

DISCUSSION

Schwannomas are benign, slow-growing tumors of the myelinproducing Schwann cells. They account for 5% of all benign soft tissue tumors; however, 25% to 45% of extracranial schwannomas are seen in the head and neck (Biswas, 2007). Actually, schwannomas are uncommon and rarely originate from motor nerves such as hypoglossal nerve (Kuo, 2008). They are usually diagnosed in middle-aged patients (with a median age of 46 years) and are predominantly seen in women (Biswas, 2007; Ho, 2005; Olsen, 1994; McCurdy *et al*., 1976). Odake (Odake, 1989), reported that

14% of hypoglossal schwannomas were associated with neurofibromatosis.

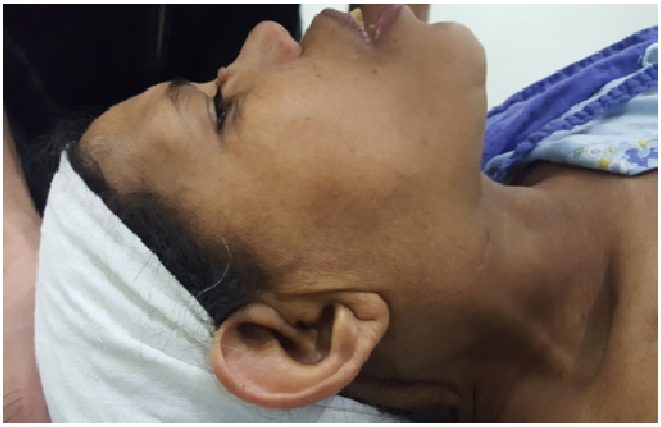

Figure 1. Large, mobile, soft level II neck mass

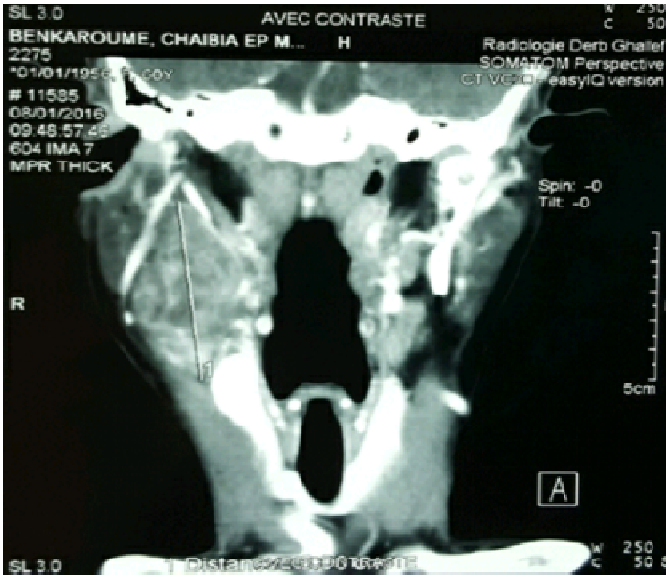

Figure 2. Ct Scan showing the right-sided paraparyngeal mass

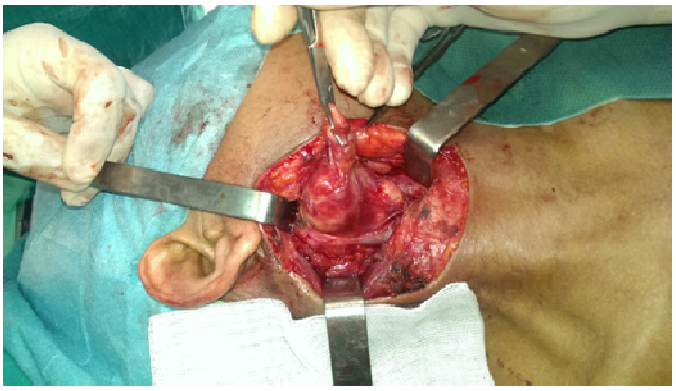

Figure 3. Excision of the mass by Sibileau's incision

After appropriate investigations, the diagnosis of neurofibromatosis was excluded in our patient. Hypoglossal schwannomas usually arise from the intracranial portion of the nerve and then extend extracranially. The most common signs and symptoms in hypoglossal schwannomas are those of hypoglossal palsy, with ipsilateral hemiatrophy, deviation and fasciculation of the tongue, headache due to meningeal irritation, and signs of cerebellar or brainstem compression (Rachinger, 2003). In 94% of cases, hypoglossal palsy is the presenting symptom, which is generally unnoticed and leads to delayed diagnosis (Sato, 1996; Rachinger *et al*., 2003; Spinnato *et al*., 1998). Both paragangliomas and schwannomas are frequently included in the differential diagnosis of parapharyngeal space neoplasms (Olsen, 1994 and McCurdy, 1976). Magnetic resonance imaging with contrast enhancement is considered the most helpful preoperative investigation, which reveals the size, shape, extension of the tumor and refined the differential diagnosis (Mariniello, 2000). But our patient can't pay for the RMI, so only the CT scan of the neck was done.

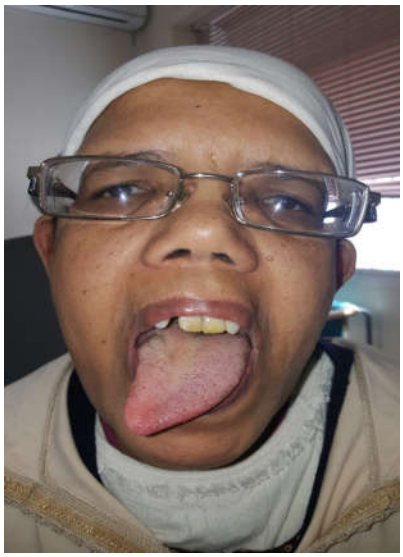

Figure 4. Right-sided hypoglossal palsy evident on Protrusion of the tongue

Because hypoglossal schwannoma is a benign tumor, a complete surgical removal should be indicated if it becomes symptomatic (Aihara, 2005). Complete surgical removal is the main goal. The intracranial as well as extracranial portions near the skull base with anatomic proximity to the vital nerves and vessels may complicate the surgery. The choice of the best surgical approach is determined by the type of tumor extension (Ho *et al*., 2005). Occlusion of the jugular bulb by the tumor can also be fatal (Aihara *et al*., 2005).

Histologically, schwannomas are encapsulated tumors containing cystic as well as solid parts. They are composed of elongated, spindle-shaped cells arranged in either an Antoni A or Antoni B pattern. The Antoni A type refers to a densely packed pattern of cellular arrangement, whereas Antoni B refers to a more loosely arranged pattern (Biswas, 2007). Schwannoma stretches the nerve fibers of its origin during growing phase. Hypoglossal nerve deficit may result from this stretching effect of the mass on the originating nerve within the canal. Our case seems totally extracranial outside the hypoglossal canal, which may prevent or at least postpone the appearance of hypoglossal palsy. Pure extracranial location of the hypoglossal schwannoma, as reported in our case, seems a very rare event. Indeed, these tumors rarely originate extracranially in which they present as parapharyngeal masses (Kuo, 2008; Spinnato, 1998 and Mariniello, 2000).

Conclusion

Hypoglossal Schwannoma account for 5% of all benign soft tissue tumors; however, 25% to 45% of extracranial schwannomas are seen in the head and neck. In front of a palsy of the tongue with a latero cervical mass, we must think of this diagnosis, and an MRI is imposed to assess the extent and size of the mass. A good response to surgical treatment is often obtained. Paralysis of the hemilangue is persistent according to the conservation of the hypoglossal nerve or his sacrifice. This condition is rare and little discussed in the literature.

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