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Case Report
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Schwannoma causing greater occipital nerve neuralgia: Case report

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ABSTRACT

Schwannomas are benign tumors that originate from the Schwann cells of the nerve sheath. They can arise from any myelinated nerve. The pre-operative diagnosis of schwannoma is difficult and should be suggested by clinical features and supported by investigations based on techniques such as ultrasonography, computed tomography (CT), magnetic resonance imaging, and fine-needle aspiration cytology. Schawannomas can present with very subtle symptoms or morbid sequel. A 19-year-old male patient presented with 6 months history of swelling in the upper left side part of the neck. Local examination showed a 5 cm × 3 cm single globular mass in the left suboccipital region. The contrast enhanced CT scan showed a 44 mm × 46 mm × 39 mm well defined heterogeneous mildly enhancing mass on the left side of the upper three cervical vertebras abutting them. The mass was excised under general anesthesia by transcervical approach. After extensive search of English literature we came across only three case reports where schwannoma of greater occipital nerve presented with neuralgia.

Key words: *Greater occipital nerve (Arnold nerve), schwannoma, suboccipital*

Introduction

Schwannomas or neurilemmomas are capsulated, benign and slow growing tumors that originate from the Schwann cells in the collagenous matrix of the neural sheath of motor and sensory myelinated nerve fibers. Schwannoma characteristically run along the course of a nerve or attached to peripheral, cranial, spinal or sympathetic nerves. These lesions are frequently seen in head and neck region (25-45%) [1]. The pre-operative diagnosis of schwannoma is difficult and should be suggested by clinical features and supported by investigations based on techniques such as ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), and fine-needle aspiration cytology (FNAC) [2,3]. Most extracranial schwannomas generally occur in parapharyngeal space [4] but are rare in the suboccipital region [5-7]. The location of tumor, nerve of origin and way of presentation is a unique feature described in this article.

Case Report

A 19-year-old male patient presented with 6 months history of swelling in the upper part of neck left side. It was insidious in onset and gradually progressive. There was a history of intermittent sharp pain in suboccipital

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region and difficulty in movement of neck. Local examination showed a 5 cm × 3 cm single globular mass in left suboccipital region. The rest of local and systemic examination was normal. The personal and family history was not contributory. The contrast enhanced CT scan showed a 44 mm × 46 mm × 39 mm well defined heterogeneous mildly enhancing mass on the left side of the upper three cervical vertebras abutting them. The mass was compressing and displacing adjacent posterior neck muscles with insulating between first and second cervical vertebrae (Figure 1). FNAC showed a cluster of spindle cells with intercellular fibrillary material suggestive of a benign spindle cell tumor. Mass was excised under general anesthesia by transcervical approach. The mass was removed in toto by blunt dissection (Figure 2). In post-operative period the paraethesia was present in the distribution of Arnold nerve. The final histopathology report was schwannoma and the specimen showed presence of Antoni A and B cells without nuclear or cytological atypia (Figure 3). The patient is under follow-up for the past 12 months and disease free.

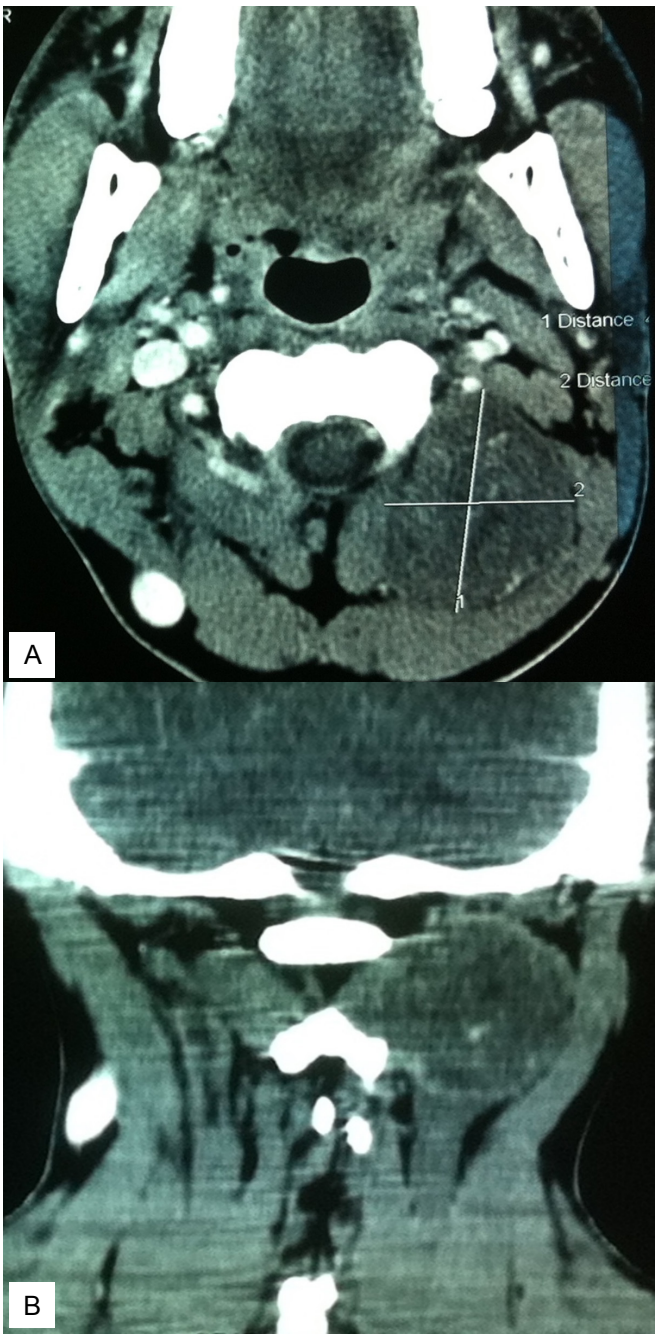

Figure 1. (A and B) Contrast enhanced computed tomography scan (axial, coronal cuts) showed a 44 mm × 46 mm × 39 mm well defined heterogeneous mildly enhancing mass on the left side of the upper three cervical vertebras abutting them.

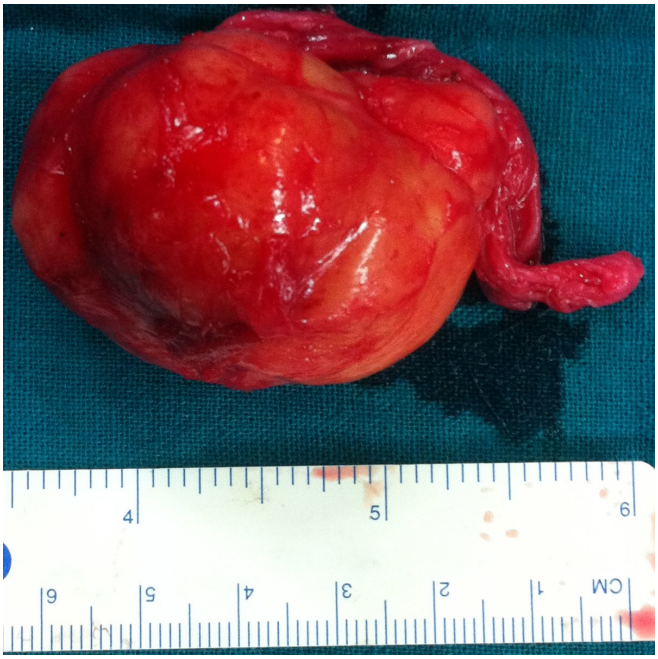

Figure 2. Specimen in toto.

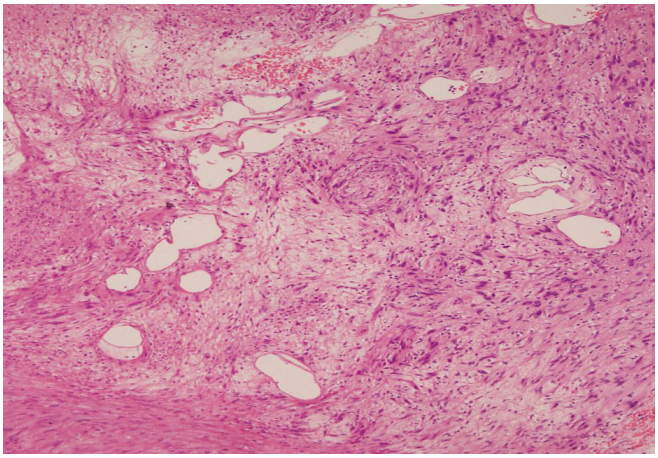

Figure 3. Alternating hypocelullar and hypercelullar areas in tumor with hyalanised vessel (H and E, ×10).

Discussion

Schwannomas are benign tumors that originate from the Schwann cells of the nerve sheath. Schwann

cells are neural crest-derived glial cells that are responsible for providing myelin insulation to peripheral nervous system axons [8]. The exact cause of origin is not known but it can be a genetic disorder or there is a family history of having neurofibromatosis. In the case here described, the tumor was presented lateral to upper three cervical vertebrae with insulating between first and second cervical vertebrae, which is the site of origin of Arnold nerve (greater occipital nerve). The schwannomas are the most common benign lesion of the nerve sheath and are typically reported in females between third and six decades [9]. Extra cranial schwannoma commonly present as a solitary mass (70%) but by expansion they may induce symptoms due to increased pressure affecting adjacent tissues. As such schwannomas are benign but malignant forms have also been reported in the literature. The malignant form is related with neurofibromatosis type II and it requires detail examination of family members.

The pre-operative investigation includes FNAC and radiographic imaging such as US, and CT or MRI. With the lack of symptoms and physical examination findings, imaging plays the central role in diagnosing the nerve of origin. On non-contrast CT, schwannomas are hypodense as compared with muscle. Contrast administration results in some degree of enhancement, which may be homogeneously solid or heterogeneous and patchy similar finding in our case [10]. On MRI, schwannomas are well-circumscribed homogenous masses that exhibit high-signal intensity on T2-weighted images and a relatively homogeneous low-signal intensity on T1-weighted images. The characteristic feature of schwannoma on histopathology is the presence of Antoni A (hypercellular condense texture with eosinophilic band between nucleus rows) and Antoni B cells (hypocellular, loose connective tissue). Schwannomas are frequently difficult to characterize on FNAC. Liu et al., reported that the accuracy of FNAC was only 20% [11]. As for the management of schwannomas, multiple treatment options exist including observation, complete tumor excision, and intracapsular enucleation. Their slow growth, low recurrence rate, and noninvasive nature, however, often allow for an observational approach. The neoplasm's are relatively radio resistant, so complete surgical resection remains the treatment of choice [12]. Intracapsular enucleation is most feasible for tumors situated eccentrically without being splayed by nerve fibers. It is usually attempted for schwannomas arising from major nerves. Because of the substantial chance of nerve palsy after operation, obtaining an accurate pre-operative diagnosis - and preferably - with the identification of the nerve of origin is crucial to the management of the disease. Only three cases have been reported in the literature, where schwannoma of greater occipital nerve presented with neuralgia [5-7].

Conclusion

The surgical resection of schwannoma often results in post-operative neurologic deficits. With careful clinical examination and proposed investigations, it is possible to predict with considerable precision the nerve giving rise to a schwannoma. This information allows the surgeon to counsel the patient on specific anticipated post-operative neurologic deficits. Only two additional cases have been reported in the literature where schwannoma of greater occipital nerve presented with neuralgia.

Author Contribution

Dr. Hitesh Verma has made substantial contributions to the conception and design of the study, acquisition of data, analysis and interpretation of data, and was involved in drafting the manuscript and revising it critically for important intellectual content. He agrees to be accountable for all aspects of the work in ensuring that any question related to the accuracy or integrity of any part of the work will be appropriately investigated and resolved. Dr. Arjun Dass, Dr. Surinder K. Singhal, Dr. Nitin Gupta have given final approval of the version to be published. Dr. Amirinder Kaur has made substantial contributions to the acquisition of data.

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Conflict of interest statement

The authors have no conflicts of interest to declare.

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