Giant Schwannoma of Neck - A Rare Case Report

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Abstract: Schwannomas, also known as neurilemmomas, are benign peripheral nerve sheath tumors. They can originate from any nerve covered with schwann cell sheath. Schwannomas constitute 25 - 45% of tumors of the head and neck. The half of them are found intracranially, within the cochleovestibular nerve (n. VIII) leading to deafness. The slow growth and the lack of characteristic clinical signs influence clinically silent development of the tumor. Presence of the clinical signs may by connecting with the pressure on the surrounding anatomical structures or the damage of the affected nerve. The treatment of the choice of the extra cranial tumors is surgical resection. Here we report case of a 65 year old male patient who presented with unilateral schwannoma of neck. Diagnosis was attained with the help of investigations like MRI neck, FNAC and trucut biopsy. In the presented case, the tumor development was without any signs. The tumor was successfully surgically enucleated.

Keywords: schwannoma neurilemmoma neck mass fnac trucut enucleation resection

1. Introduction

Schwannomas are tumors of Schwann cell origin, either central, peripheral, or autonomic nervous system. Schwannomas do not arise from the optic and olfactory nerve as they lack schwann cells. Perineural fibroblast tumor, nerve sheath tumors, neurilemmomas are some of the other nomenclatures. Around 25% - 45% of these tumors are present in the head and neck region. Schwannoma generally presents in the 4th to 6th decade of life with no sex predilection. They are usually seen in the intracranial cavity, oral and nasal cavity, over the face and even scalp.

In the head and neck region, the most common site of tumor is in the parapharyngeal space. Clinical presentation varies depending on its anatomic site. In the parapharyngeal space, the vagus nerve is the most common nerve of origin (NOO).

Pre - operative diagnosis may be made by radiological imaging like computed tomography (CT) and magnetic resonance imaging (MRI). Imaging can also give an idea about the preoperative estimation of the parent nerve of origin. It also helps to differentiate between different origins of schwannoma. Fine needle aspiration cytology (FNAC) is generally not performed to prevent injury to vital structures. Complete surgical excision is the treatment of choice.

2. Case Report

A 65 year old male patient presented to department of General surgery with history of swelling over left lateral side of neck since last 15 years. On examination there was a 15*10 cm swelling located for left lateral side of neck possibly from posterior triangle of neck. The mass was firm and mobile with smooth surface and displacing the internal jugular vein and external carotid artery outwards. There was no complaint of hoarseness of voice or voice change, no pain over swelling was elicited, no complaints of FNAC from the mass.
Trucut biopsy showed histological features of spindle cell lesion.

TRUCUT from the mass.

With this background the tumor was completely resected under general anesthesia. The entire tumor was safely separated from the nerve of origin and totally excised while preserving the anatomic continuity and the functionality of the nerve of origin.
Excised schwannoma from neck
Histological examination confirmed the diagnosis of schwannoma, with the nerve of origin most probably arising from the cervical sympathetic chain.
Excisional biopsy of the mass
The postoperative period was uneventful, and the patient was discharged on the seventh postoperative day. Upon follow-up after 12 months, he remains asymptomatic with no signs of recurrence.

3. Discussion

Schwannomas and neurofibromas arise from schwann cells. Schwannomas arises exclusively from schwann cells, while neurofibroma comprises of a mixture of three cells, namely schwann cells, perineural cells, and perineural fibroblasts. Hence neurofibromas are inseparable from NOO (nerve of origin).

Head and neck schwannomas always present as lateral neck swelling, in few rare cases they might present also present with symptoms of pressure. In our case the patient presented with asymptomatic unilateral neck swelling.

Schwannomas of the head and neck are slowly growing benign tumors. There is an incidence rate of 8 - 13% of malignant transformation OF schwanna. They are usually asymptomatic, with the most common presentation being a symptomless solitary mass. Symptoms, like dysphagia, nasal obstruction, dyspnea and hoarseness of voice might occur in the later stages, due to the increased size of the mass compressing the surrounding anatomical structures.

The differential diagnosis of these lesions include inflammatory cervical lymphadenitis, metastatic lymphadenopathy, lymphoma, paraganglioma, salivary gland neoplasms, branchial cleft cyst, carotid body tumors, neurofibroma, and carotid aneurysm.

Imaging studies like CT and MRI play an important role in distinguishing between the entities included in the differential list as it can depict the relationship of the mass with the adjacent structures, and provide some insight into the mass composition. Differentiation between vagal schwannomas and cervical sympathetic chain schwannomas can be made on the basis of displacement of the carotid sheath contents. The cervical sympathetic schwannomas displace both the carotid artery and internal jugular vein laterally since they are not inside the carotid sheath, while vagal schwannomas tend to separate them. In our case the nerve of origin was suspected as a branch of sympathetic cervical chain.

The role of FNAC is debatable because schwannomas are deep-seated and usually adjacent to the large vessels, and injury may occur. Hence, FNAC is not recommended, and is performed only for malignancy to be ruled out. In our case, ultrasound guided FNAC and later trucut biopsy was performed by interventional radiologist.

Schwannoma are resistant to radiotherapy, hence complete surgical removal of the tumor is the mainstay of treatment. Literature mentions two different surgical methods of resecting schwannomas. One involves the dissection of the nerve fascicle from which the schwanna origin, removing the schwanna with its capsule (total resection), while the other, more conservative, method involves the enucleation of the schwanna from inside its capsule with the preservation of the nerve of origin. In our case the latter was performed.

In the study by Liu et al., the rate of nerve palsy after operation were 100%, 67%, and 50% for resection with division of NOO (nerve of origin), intracapsularenucleation, and debulking of the tumor, respectively. Hence intracapsular dissection does not guarantee the preservation of NOO, if the tumor is large.

A recent review by Sandler et al, comparing these two surgical techniques showed that the enucleation method was statistically superior in terms of nerve - sparing and postoperative course. More patients in the enucleation group reported that they were asymptomatic postoperatively. Furthermore, the patients who reported postoperative symptoms of hoarseness or vocal fold paralysis were fewer.
in the enucleation group compared to those who underwent total excision.

The main aim of the treatment is excision of the tumor with minimizing the postoperative neurological deficit. However, the postoperative course also depends on the surgeon’s skills and familiarity with the technique as well as the size and location of the lesion.

In this case it was difficult to make out the nerve of origin on preoperative investigations. After surgical removal of tumour, the specimen was sent for histopathological examination which suggested schwannoma. Our presumptive nerve of origin for this case is a branch from cervical sympathetic ganglion. On follow - up after 12 months, the patient remains asymptomatic with no signs of recurrence.

4. Conclusion

Schwannoma being a benign rare tumor is one of the differential diagnosis of a unilateral slow growing mass in the head and neck region, especially in an adult. Schwannomas present as a diagnostic dilemma as they are asymptomatic in most cases. Diagnosis of schwannoma is established by clinical examination, FNAC, and radiological imaging. FNAC should always be done after CT/MRI scan to rule out paragangliomas. Sympathetic and vagal schwannomas can be diagnosed depending upon the displacement of carotid artery and internal jugular vein. Nerve of origin cannot be made out in all cases except if it arises from major nerves. The gold standard management of schwannoma is surgical and depends on the location of the tumor and nerve of origin. Treatment of extracranial schwannomas involves tumor enucleation or total surgical removal together with the affected nerve segment. Conservative treatment can be given in patients with small, slow - growing, asymptomatic tumors. Due to their rarity, complex anatomical location and morbidity risk post excision, they can pose a formidable challenge to surgeons.

References