



ORIGINAL RESEARCH PAPER

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CERVICAL SCHWANNOMA BY TRANSCERVICAL APPROACH: OUR EXPERIENCE

KEY WORDS: Cervical Schwannoma, Parapharyngeal Space, Transcervical Approach, Peripheral Nerves Pps - Parapharyngeal Space

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ABSTRACT

Schwannomas are benign neoplasms of the peripheral nerves originating from the Schwann cells. They are rare and usually solitary, with clearly delimited capsules. They occur in the head and neck region, in approximately 25 % of the cases, and may be associated with Von Recklinghausen's disease. About 4% of head and neck schwannomas present as a sino nasal schwannoma whereas Brachial plexus schwannoma constitute only about 5% of schwannomas and 2–5% of neurogenic tumors constitute Cervical vagal schwannomas which are quite rare. They are always a diagnostic dilemma as they are asymptomatic for long time and histopathology is the gold standard for diagnosis.

METHODS- We present a case series of 3 patients of Large schwannomas, arising from the cervical vagus presenting as parapharyngeal Neck Mass. Tumour was completely removed by Extracapsular Excision using transcervical approach.

CONCLUSION- Schwannomas can remain asymptomatic for long time can present as huge Parapharyngeal or Neck Mass specially in adults. As a rule, treatment is surgical and dictated by the location of the tumor and nerve of origin. Due to its rarity, complex anatomical location and morbidity risk post excision, they can pose a formidable challenge to surgeons. This study aims to describe the presentation, workup, surgical technique, and outcome of large head and Neck Schwannomas by Transcervical Approach.

INTRODUCTION

Cervical Schwannomas are a type of parapharyngeal tumor that most often presents as an asymptomatic unilateral neck mass. They are benign in nature, solitary and rare tumors found in the head and neck region. They can arise from the nerve sheath of peripheral, spinal or cranial nerves. These tumors possess a diagnostic challenge due to their rarity, developmental origin, and differential diagnosis. Although they are benign in most of the cases, their surgical removal is needed due to the neurovascular deficits arising because of the anatomical location of these tumors.

The aim of this study is to describe our experience with Large Schwannomas resected via a transcervical route without mandibulotomy. Traditionally mandibular swing approach is used to access these huge lesions, even if it caused delayed postoperative instability and deformities thus requiring a reconstructive procedure. In order to prevent these complications, we have performed the transcervical approach which is an alternative and less invasive technique. We present three cases of large cervical schwannomas out of which one is extending to skull base and was excised by Transcervical Approach.

MATERIAL AND METHODS

Three cases of benign masses in the head and neck that were confirmed by postoperative histopathological examination at the ENT & Head and Neck Surgery Department of Dr. B.R.A.M. Hospital Raipur between 2016 and 2019 are described in our study. The inclusion criteria for this study were the patients who had their postoperative histological diagnosis confirmed to be schwannoma. Pre-operative diagnostic tools included Fine Needle Aspiration Cytology, Contrast CT or MRI scan. The treatment of choice was Surgical Excision of the tumour via Transcervical Approach along with preservation of the nerve of origin. Postoperatively, the data on surgical complications and nerve deficits caused by tumour resection were evaluated.

RESULTS

Complete surgical excision of the tumour without incorporating mandibulotomy or combining with transparotid

approach was feasible in all the above mentioned cases without any partial or complete neurological deficit.

CASE 1

A 17 Year old female patient came to outpatient department of ENT Dr. B.R.A.M. Hospital Raipur with a large swelling on the left side of the neck since Childhood. It was slow in onset and gradually progressive in nature. There was complain of breathing difficulty, change of voice, pain in swallowing. On general examination, patient was moderately built. Vitals were stable. There was no pallor, icterus, clubbing or edema. There was no palpable neck node or enlarged gland. Local examination uvula was displaced due to mass. Ear nose, oral cavity examination were within normal limit. Systemic examination was normal. FNAC was done which showed schwannoma or neurofibroma. The mass contained globular soft tissue mass of size 15x13x6 cm. Outer surface was smooth and encapsulated. On cut section tumor was nodular and variegated in appearance. CECT imaging revealed left sided well defined smooth margined cystic lesion with heterogeneous solid component within it noted at left carotid space, extending superiorly from base of skull, from the middle cranial fossa and extending inferiorly at infra hyoid neck region along cervical part of carotid space just 1cm above the left sternocleidomastoid joint. Lesion was causing medial displacement of pharyngeal mucosal space, anterolateral displacement of parapharyngeal mucosal and carotid vessels and lateral displacement of parotid space. It was also causing right lateral displacement of pharynx. Left internal jugular vein and common carotid artery was drapping around the lateral margins of the lesion with thrombus of the internal jugular vein. Lesion shows heterogeneous post contrast enhancement of solid and peripheral enhancement of cystic component measuring size 13(cc)x8(AP)x((TR) cm. The patient underwent surgery through a trans cervical approach under general anaesthesia and mass sent for histopathological examination which showed schwannoma. The tumor was massive in size weighing around 800gm. Patient was referred to Neuromedicine Department for Anticoagulant Therapy after the Surgery. Fortunately, There were no associated complications or neurological deficits after the surgery. (Figure i,ii,iii)



Figure i Preoperative Clinical picture of the tumor.



Figure ii Pre Operative CECT image of the tumor



Figure iii Intraoperative Picture Showing Enucleated Tumor with intact capsule

CASE 2

A 30 Years old female patient came to outpatient department of ENT Dr. B.R.A.M.Hospital Raipur with a swelling a large swelling on the left side of the neck for last 4 years. It was slow in onset and gradually progressive in nature. There was is no complain of breathing difficulty, change of voice, pain or difficulty in swallowing or pain in ear. On general examination, patient was moderately built. Vitals were stable. There was no pallor, icterus, clubbing or edema. There was no palpable neck node or enlarged gland. Local examination larynx by indirect laryngoscopy was normal. Ear nose, oral cavity examination were within normal limit. Systemic examination was normal. FNAC was done which showed schwannoma or neurofibroma. CECT imaging revealed right sided soft tissue tumor in the carotid space extending superiorly from the base of skull from the middle cranial fossa and extending inferiorly at infra hyoid neck region along cervical part of carotid space just 1 cm above the left sternocleidomastoid joint. The lesion has well defined smooth margined cystic lesion measuring size 10(cc)x 6 (AP)x 7(TR) cm with internal heterogeneously enhancing solid component. The tumor was removed by trans cervical approach under general anaesthesia and sent for histopathological examination which showed schwannoma. (Figure iv,v ,vi,vii)

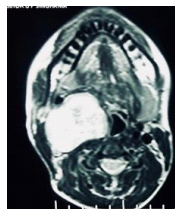
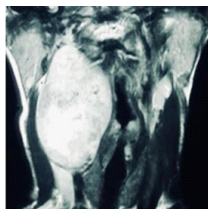


Figure iv and v Showing Pre operative CT image of the Pharyngeal Mass in Coronal and axial Cuts.



Figure vi Showing Intraoperative picture of tumor excision

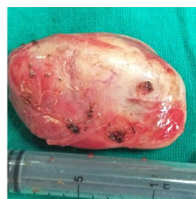


Figure vii Intraoperative Picture Showing Enucleated Tumor.

CASE 3

A 23Year old male patient came to outpatient department of ENT Dr. B.R.A.M. Hospital Raipur with a swelling a large swelling on the left side of the neck for last 3 months. It was slow in onset and gradually progressive in nature. There was no history of breathing difficulty, change of voice, pain in swallowing. On general examination, patient was moderately built. Vitals were stable. There was no pallor, icterus, clubbing or edema. There was no palpable neck node or enlarged gland. Local examination larynx by indirect laryngoscopy was normal. Ear nose, oral cavity examination were within normal limit. Systemic examination was normal. FNAC was done which showed schwannoma or neurofibroma. CECT imaging revealed ride sided ill defined heterogeneously enhancing soft tissue density lesion in right parapharyngeal space encroaching into right carotid space encasing the vagus nerve. By tumor size indication, the patient underwent surgery through a trans cervical approach under general anaesthesia and sent for histopathological examination which showed schwannoma.

DISCUSSION

Schwannoma are the benign tumor of nerve sheath cells. Twenty-five to forty percentage of schwannomas of the body are seen in head and neck region.[1] They were first described by Verocay in 1910. These are slow growing, smooth, and regular benign masses. Fine-needle aspiration cytology (FNAC) has been standard in diagnosing the nature of neck swelling, but in cases of schwannoma, FNAC may not be helpful in giving a diagnosis. The other limitation of FNAC is accessibility of tumors deep-seated in parapharyngeal space in close relation to great vessels. Hence in such scenarios, MRI is the gold standard of imaging which will not only reveal the extent of swelling but also its nature of pathology as well as organ/site of origin. Anatomically, vagus lies in the carotid sheath and hence, tumors arising from the vagus will shift the internal jugular vein laterally and internal carotid artery medially. The cervical sympathetic chain lies postero-medial to carotid sheath, and hence, the tumors arising from cervical sympathetic chain will push the great vessel anteriorly and not splay them. [2,3]. Surgery is the main stay of treatment for schwannomas of the parapharyngeal space (PPS) [4–11]. The choice of operation is mainly determined by the relationship between the tumor and the nerve of origin. The choice of surgical approach is dictated by the size of the tumor, its location, its relationship to the great vessels and the suspicion of malignancy. A number of middle cranial fossa skull base approaches have been described for resection of trigeminal schwannomas involving the root, ganglion and the intra cranial portion of the trigeminal nerve [5,6]. Approaches to the PPS schwannomas include, transcervical, transzygomatic, transmaxillary, the combined transcervical transparotid approach, the combined transcervical transmandibular approach and a combination of the above approaches. The combined transcervical-transmandibular approach is preferred in malignancies in which better exposure facilitates oncologic resection and in cases in which distal control of the carotid artery at the skull base is required [7-9]. It is also the preferred approach for large tumors with superior PPS and extension. This approach is associated with many unique complications, primarily because of incorporation of a mandibulotomy. The complications include infection, temporomandibular joint dysfunction, nonunion, malocclusion, plate extrusion, and tooth loss. More over a tracheostomy is usually performed in conjunction with a transmandibular approach because of the significant upper airway edema caused as a result of the surgical manipulation of the oropharynx.

A combined transcervical-transparotid approach, on the other hand is devoid of the mandibulotomy associated complications. In this approach, the transcervical approach is combined with a transparotid approach by extending the incision superiorly as for a parotidectomy. The facial nerve is

identified and dissected, a parotidectomy is performed and the PPS space is delineated. Although the overall exposure is limited, well encapsulated, radiologically benign appearing. In surgical approaches combined with mandibulotomy, damage to the inferior alveolar nerve, malocclusion and non-union-malunion defects and loss of dentition may occur. Additionally, in some types of osteotomies, lip-splitting may be required. Due to damage to the floor of the mouth during the surgery, tracheostomy and nasogastric tube feeding may be required. Fisch described an infratemporal fossa approach for extremely large PPS tumours invading the temporal bone and middle cranial fossa. [12]

Reviewing the literature on parapharyngeal tumours approached transcervically, two case series were found. Chang et al. reported on 51 cases with the largest tumour size of 6.8 cm, while Presutti et al. described 18 cases with the largest tumour size of 8 cm [13,14] Basaran et al in their study of 44 cases the largest tumour was a pleomorphic adenoma with a horizontal diameter of 11 cm, they emphasised that the vertical diameter should be evaluated rather than the horizontal diameter to determine whether the tumour is suitable for excision through a transcervical route. Basaren et al suggested that if vertical extension of the tumour is suspicious for intracranial extension, a transcervical approach is dispensable. Furthermore, it should be taken into consideration, particularly in neurogenic tumours, that it might be very difficult to dissect the tumour from the surrounding tissues, especially in the vicinity of the cranial base. Additionally, among the largest parapharyngeal tumours reported the giant pleomorphic adenoma reported by Basaren et al was with a diameter of 11 cm which was a good example for large PPS tumours underlining that mandibulotomy is unnecessary just owing to the size of the tumour. [15] In our study the dimensions of the Case 1 was 15cm* 15 cm* 6 cm which is again very huge size reported till now.

The accepted lines of surgical management are complete excision of the schwannoma, but the major drawback of this approach is unreparable neurological damage of the concerned nerve of origin. In cases of the vagus nerve, this will lead to vocal cord paralysis, loss of movement of palate and ipsilateral pharynx leading to hoarseness of voice and aspiration. In cases with involvement of cervical sympathetic chain, postoperatively patient develops Horner's syndrome. In both scenarios, there is a permanent neurological deficit which compromises the quality of life severely. Hence, we attempted Surgical Excision through transcervical approach for the tumors arising from the nerves whose neurological function are intact preoperatively and extending upto skull base. Further studies with larger sample sizes and with long-term follow-ups are advocated to establish the success and drawback of this technique, but till then, this technique should be resorted to as an option in managing schwannomas with no preoperative neurological deficits and extending to skull base.

Although no neurological complications or complications of other types were observed after the operation in the discussed case. The most common postoperative complication post removal is vocal cord paralysis, which occurs in up to 85 percent of all the cases and results in hoarseness.

CONCLUSION

Extra cranial schwannomas in the head and neck region are rare neoplasm. Diagnosis is established by imaging studies such as magnetic resonance imaging or computed tomography, while FNAC is used to rule out other pathology and tissue diagnosis. The accepted treatment for these tumors is surgical resection with preservation of the nerve function. Management of schwannomas has always been a great challenge. Complete surgical excision has been the gold standard for management. Excision of large tumour often requires more radical approach to gain access for removal

but results into a partial or complete neurological functional deficit of the nerve of origin. We have tried using Transcervical Approach for these huge schwannomas and preserved its function postoperatively.

A transcervical approach should be considered as choice for excision of parapharyngeal space tumours, except for recurrent or malignant tumours, considering its advantages of providing direct access to the neoplasm, adequate control of neurovascular structures from the neck and optimal aesthetic outcomes due to preservation of mandibular continuity with minimal morbidity and hospitalisation time. We consider mandibulotomy or other highly invasive procedures unnecessary in such cases as it increases the postoperative morbidity. We advocate use of this approach as choice in huge schwannomas extending to skull base.

CONFLICTS OF INTEREST

None.

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