Case Report

Neuroendocrine tumor of parapharyngeal space

Marlinda Adham, Novra Widayanti
Department of Ear, Nose and Throat, Head and Neck Surgery
Faculty of Medicine University of Indonesia
Cipto Mangunkusumo Hospital
Jakarta

ABSTRACT

Background: Parapharyngeal space tumors account for some 0.5% of tumors of the head and neck, most of them benign. The most common benign neoplasms are salivary gland neoplasm, paragangliomas and followed by neurogenic tumors. The importance of these tumors lies mainly in two aspects, the difficulty of early diagnosis, due to the lack of symptoms in the initial stages and, on the other hand, the extreme risk of complications in performing surgery in the parapharyngeal region. **Purpose:** We present this case to enlighten general practitioners and also otorhinolaryngologist about diagnosis and management of parapharyngeal tumor. **Case:** One clinical case of neuroendocrine tumor in parapharyngeal space on a 37 years old man. **Management:** The patient underwent diagnosis procedure and extirpation of the tumor mass. **Conclusion:** Parapharyngeal tumor is one of head and neck tumors that has good prognosis, especially if diagnosed early and adequately treated.

Keywords: neuroendocrine tumor, parapharyngeal space, benign tumor.

ABSTRAK

Latar belakang: *Tumor parafaring meliputi sekitar 0,5% dari seluruh tumor kepala dan leher, sebagian besar jinak. Tumor jinak yang paling sering adalah tumor kelenjar liur, paraganglioma dan tumor neurogenik. Tumor parafaring ini penting disebabkan sulit untuk melakukan diagnosis dini karena sedikitnya gejala pada tahap awal dan kemungkinan komplikasi yang dapat terjadi pada saat dilakukan tindakan bedah di daerah parafaring.* ***Tujuan:*** *Kasus ini diajukan agar para dokter umum dan spesialis Telinga Hidung Tenggorok dapat mengenali diagnosis dan penatalaksanaan tumor parafaring.* ***Kasus:*** *Dilaporkan satu kasus tumor neuroendokrin parafaring pada laki-laki usia 37 tahun.* ***Penatalaksanaan:*** *Pada pasien ini dilakukan prosedur untuk mendiagnosis tumor dan dilakukan ekstirpasi massa tumor.* ***Kesimpulan:*** *Tumor parafaring merupakan salah satu dari tumor kepala dan leher yang mempunyai prognosis baik terutama bila didiagnosis secara dini dan diterapi secara adekuat.*

Kata kunci*: Tumor neuroendokrin, spatium parafaring, tumor jinak.*

Alamat korespondensi: Marlinda Adham, Department of ENT-HN Faculty of Medicine University of Indonesia- Cipto Mangunkusumo Hospital. Jl. Diponegoro 71, Jakarta. e-mail: marlinda.adham@yahoo.com

INTRODUCTION

Tumors of the parapharyngeal space (PPS) are rare, most of them being diagnosed in adults, but occasionally found in children. PPS tumors often presented as an asymptomatic growth and may be detected in a routine medical checkup or as an accidental finding when scanning for another reason. There are many difficulties in diagnosis of parapharyngeal mass because of late manifestation of the symptoms and the image of the mass on CT Scan or MRI.[1,2]

PPS tumors are infrequent, accounting for some 0.5% of neoplasms of the head

and neck. Most of these tumors (70-80%) are benign and approximately 50% originate from the deep lobe of the parotid or from the minor salivary glands, particularly the pleomorphic adenoma.[3,4]

The symptoms are manifested when the tumor becomes larger than 2.5–3 cm and related to the prestyloid–poststyloid localization.[3,5]

Tumors of the PPS include primary neoplasms, direct extension from adjacent regions, and metastatic disease.[1,2,5]

The diagnosis is done by physical examination, radiological imaging with CT or MRI with contrast and pathological examination which could be performed by fine needle aspiration cytology (FNAC).[3,6]

Choosing the appropriate approach before surgery is very important when dealing PPS. Transcervical approach allows direct access to PPS. Infratemporal fossa skull base surgery approach provides a very wide operative field, which is preferable for the resection of large PPS.[7,8]

We present this case to enlighten ENT surgeons about diagnosis and management of parapharyngeal tumor. Diagnosis performed with nasopharyngeal CT and Magnetic Resonance Angiography (MRA) that showed big mass that had vascularisation from left external carotid artery and the management done by transcervical approach with vessel ligation.

CASE REPORT

A 37 years old male came to ENT Oncology Division in Cipto Mangunkusumo Hospital (CMH) with chief complaint a lump in the left neck since two years. There were difficulty in swallowing, nasal voice and snoring but no pain. Physical examination showed assymetric pharyngeal arch, uvula was pushed to the right side, left lateral pharyngeal wall was bulged to medial and lump in the left neck with the size 5x5x2 cm, hard, fixated and painless. From nasoendoscopic evaluation there was no mass on nasopharynx but fossa Rossenmuller, torus tubarius and Eustachian tube were protruded. Nasopharyngeal computed tomography (CT) scan showed left parapharyngeal mass that heterogenly enhanced after given contrast, pushed vascular structures to lateral, no vascular infiltration, no left neck lymphadenopathy. It was suggested to do neck CT angiography.

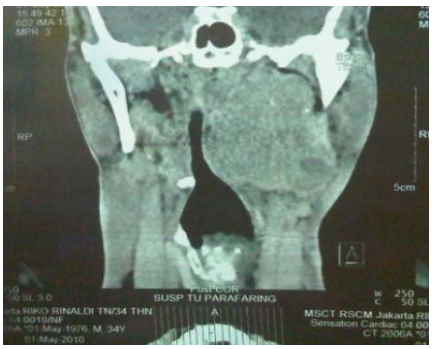

Figure 1. Coronal CT scan of parapharyngeal mass showed left parapharyngeal mass.

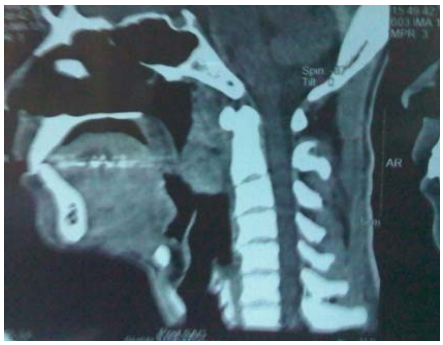

Figure 2. Sagittal CT scan of parapharyngeal mass.

Parapharyngeal biopsy result showed moderate-severe dysplasia with chronic inflammation. FNAB result from the left neck lymphnode showed a metastasis from poorly differentiated carcinoma. MRI evaluation showed a big mass that got vascularisation from left external carotid artery.

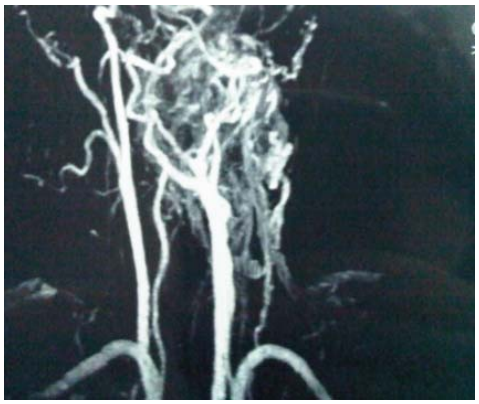

Figure 3. MRA of parapharyngeal mass showed the feeding artery from left external carotid artery.

Histopathology result from parapharyngeal biopsy was carcinoma that difficult to determine the type and origin, could be from neuroendocrine or vascular and suggested for immunohistochemistry examination and the final result was neuroendocrine mass of small cell carcinoma. Thyroid USG result was left lymphadenopathy colli in accord with malignancy with normal thyroid. Extirpation of left parapharyngeal neuroendocrine tumor was done by transcervical approach and preceded by external carotid artery ligation. After the procedure, from flexible laryngoscopy evaluation there was left vocal cord paralysis. Thorax CT scan was taken 15th month after surgery and there was right lung consolidation on 6th segment with diferential diagnosis of lung tuberculosis or metastasis and hepatomegaly. Echo cardiography result was normal. The result of cytology taken by bronchoscopy showed no malignancy sign. The patient was planned to be given chemotherapy.

DISCUSSION

A patient complained of a lump in the left neck, difficulty in swallowing and snoring. These were in accordance with literature that the most common symptoms of parapharyngeal tumors were neck mass, dysphagia, hoarseness, vocal cord paralysis, oropharyngeal mass and hearing loss. The symptoms are manifested when the tumor size 2.5–3 cm and related to the prestyloid–poststyloid localization. Pain in the neck accompanied with lock-jaw and/or paralysis of any of the pairs of cranial nerves suggested malignancy in origin.[3,5,9]

From physical evaluation there were asymetric pharyngeal arch, uvula was pushed to the right side, left lateral pharyngeal wall bulged to medial as there was a big mass on left neck.

Tumors of the PPS may include primary neoplasms, direct extension from adjacent regions or metastatic disease. The presence of a mass in the parotid is often associated with mass in the oral cavity. The presence of pain or neuropathy should direct the clinician to suspect a primary or metastatic cancer but lesions of the posterior PPS may also present with a neuropathy. A tumor originating in the cervical sympathetic chain may produce Horner's syndrome. [2,5]

Malignant tumors can invade the PPS from the nasopharynx, oropharynx, mandible, maxilla, oral cavity, or parotid gland and could extend intracranially through the jugular foramen, or other foramina of the skull base; they may also erode bone or invade or expand to involve other proximal spaces such as the retropharyngeal space or infratemporal fossa. Neoplastic pathology of the parapharyngeal space can mainly be divided into salivary, neurogenic and vascular tumors.[1,2,10]

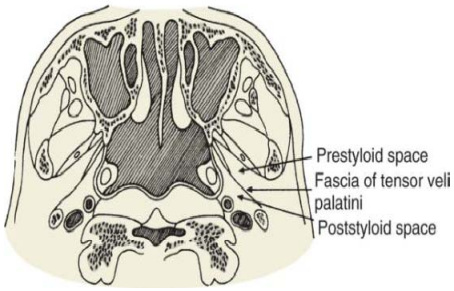

Figure 4. The fascia of the tensor veli palatini muscle divides the parapharyngeal space into a prestyloid and a poststyloid compartment.[2]

From the anatomical point of view the prestyloid PPS contains the deep lobe of the parotid gland, adipose tissues, small blood vessels, lymphatics, and minor nerves. The poststyloid PPS contains the carotid sheath and is also traversed by cranial nerves IX, X, and XII. The cervical sympathetic chain lies posterior to the carotid artery. This tumor should be extirpated but we should consider the possible complication of surgery like bleeding and paralysis of the fascial nerve.[1,2]

The diagnosis is done by physical examination, radiological imaging like CT or MRI with contrast and pathological examination which could be performed by fine needle aspiration cytology (FNAC).

Although MRI is better than CT, contrast CT is the modality of choice. MRI provides useful information on tumor localization and extent, and distinguishes tumors of the deep lobe, neurogenic lesions, intravagal paraganglioma or carotid body tumors and their relations with the internal carotid artery and adjacent structures. Angiography is recommended if paragangliomas or the involvement of the carotid artery is suspected.[2,6]

On CT scan, parapharyngeal mass that pushed vascular structures to lateral was noted. The mass was seen in post styloid region and might be a primary tumor.

FNAC can be an easy, rapid, and effective method of diagnosing these lesions. Diagnostic difficulty persists due to their similar mode of presentations and at times morphological overlap. Hence, some cases can only be confirmed by histopathological examination and some by immunohistochemistry.[3,6]

FNA via a transoral approach for visible parapharyngeal space lesions is an option with an accuracy of 78–86%, but a false-negative rate as high as 19% secondary to inadequate stabilization of the lesion. The value of FNA includes its low invasiveness, the use of small needles in an area where several vascular structures are present.[7]

Open biopsy is not advised, due to the risk of bleeding, opening of the capsule and, accordingly, relapse and seeding to neighbouring tissues. The result from parapharyngeal biopsy was neuroendocrine or vascular, supported by immuno-histochemistry.[9]

Papadogeorgakis et al[11] consider the following five points to be the main parameters when deciding on the best PPS surgical approaches. First, the proximity and the projection of the tumor to the oropharyngeal wall or the neck. Second, the size of the tumor (the bigger the size, the wider the access required). Third, the suspicion of malignancy, because if a malignant PPS tumor must be removed, it is required wide access in order to obtain clear margins. Fourth, the vascularity of the tumor, because in PPS tumors of high vascularity, the intraoperative risks of massive blood loss make wide access important for effective surgical handling. Fifth, the relation of the tumor to the neck neurovascular bundle is of great importance.

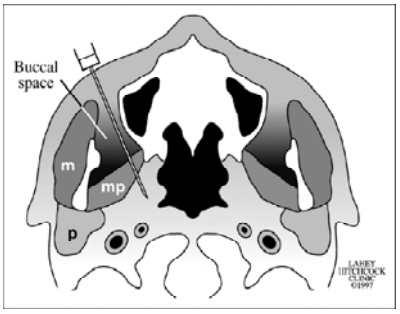

Figure 5. Schematic diagram shows the buccal space and biopsy needle. .m= masticator muscle; mp = medial pterygoid muscle; p= parotid gland.[12]

To maximize the safety of this procedure, wide access to the PPS is recommended. The surgical approach must be as minimal as possible, but wide enough to ensure the complete removal of the tumor and the safety of the vessels and nerves of the neck. During the operation the surgeon must be ready to modify the plan, if unexpected difficulties in the complete and safe removal of the tumor arise.[13]

In our case neuroendocrine tumor extirpation was performed with carotid artery ligation prior the surgery to prevent massive bleeding. Neurogenic tumor was only about 13% from parapharyngeal space tumor. From the literatures, there were three grades of neuroendocrine tumor: low grade, intermediate grade and high grade tumor. The neuroendocrine tumors can be found in lung, thymus and pancreas. Result from bronchoscopy-cytology there was no malignancy.

Transcervical approaches was indicated for removal of tumors originating from minor salivary gland, schwannommas and para-gangliomas in the poststyloid space, and presumably benign tumor. Transcervical approach allow directly acces to PPS. Removal of the tumor is often preceded by vessel ligation. On follow up the patient doesn't have any complaint, no hoarseness nor difficulty in swallowing.[14]

Successful treatment of disseminated neuroendocine tumors requires a multi-modal approach; radical tumor surgery may be curative but is rarely possible. Therapy with radionuclides may be used for tumors exhibiting uptake to a diagnostic scan, either after surgery to eradicate microscopic residual disease or later if conventional treatment or biotherapy fails. Maintenance of the quality of life should be a priority, particularly because patients with disseminated disease may experience prolonged survival.

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