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Siti Nazira Abdullah[1,2], Juani Hayyan Abdul Karaf[2],
Vijayaprakash Rao A/L Ramanna[2], Najah Momin[3], Irfan Mohamad[1]

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# Tłuszczak okolicy przestrzeni przygardłowej naśladujący guz przytarczyc u 4-letniego chłopca

## Parapharyngeal lipoma mimicking parotid tumour in a 4-year-old boy

[1] Department of Otorhinolaryngology – Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, 16150 Kota Bharu, Kelantan, Malaysia
[2] Department of Otorhinolaryngology and Head and Neck Surgery, Malacca General Hospital, Jalan Mufti Haji Khalil, 75400, Malacca, Malaysia
[3] Department of Pathology, Malacca General Hospital, Jalan Mufti Haji Khalil, 75400, Malacca, Malaysia
Adres do korespondencji: Assoc. Prof. Dr Irfan Mohamad, Department of Otorhinolaryngology – Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, 16150 Kota Bharu, Kelantan, Malaysia, tel.: 609-7676420, fax: 609-7676424, e-mail: irfankb@usm.my

Streszczenie

Guz okolicy przestrzeni przygardłowej jest rzadko spotykaną jednostką chorobową w pediatrii. Może to być guz pierwotny wywodzący się ze struktur przestrzeni przygardłowej, proces rozrostowy sąsiednich struktur lub guz przerzutowy. U dorosłych często występują nowotwory ślinianek i przyzwojaki, natomiast u dzieci przeważają guzy neurogenne. Delikatne struktury anatomiczne przestrzeni przygardłowej utrudniają przeprowadzenie badań diagnostycznych, szczególnie w pediatrii. Chociaż biopsję tkanki można wykonać w warunkach sedacji lub znieczulenia miejscowego, histologiczne rozróżnienie między tłuszczakiem a tłuszczakomięsakiem sprawia trudności. W pracy przedstawiono przypadek 4-letniego chłopca, u którego stwierdzono obecność dużej masy zajmującej przestrzeń przygardłową, oraz omówiono zastosowane leczenie.

**Słowa kluczowe:** tłuszczak okolicy przestrzeni przygardłowej, guz okolicy przestrzeni przygardłowej, dostęp przezszyjny, biopsja tłuszczaka

Abstract

Parapharyngeal space tumour is rare in paediatrics. It can either be a primary tumour arising from parapharyngeal space structures, an extension from the surrounding structures or a metastatic tumour. In adults, salivary gland tumours and paragangliomas are common, while neurogenic tumours predominate in paediatrics. The delicate anatomy in the parapharyngeal space makes the diagnostic procedures more complex, especially in paediatrics. Although tissue biopsy can be obtained under sedation or local anaesthesia, it is histologically difficult to differentiate lipoma from liposarcoma. We present a paediatric case with a large parapharyngeal space mass in a 4-year-old boy and the management used.

**Keywords:** parapharyngeal lipoma, parapharyngeal space tumour, transcervical approach, lipoma biopsy

## INTRODUCTION

Parapharyngeal space (PPS) lesion can be complex, depending on the structures it arises or extends from. In paediatric populations, infections and inflammation are the leading cause of lesions, followed by neoplasm. The primary PPS neoplasm in paediatric population is a rare entity, accounting for about 0–17% of cases[1]. Most of the cases are due to a neoplasm of neurogenic origin, followed by other neoplasms, such as lipoma, cystic hygroma, branchial cyst and vasoformative lesions[1]. About 56–67% of paediatric primary PPS tumours were malignant[1,2]. Lipoma is a common benign neoplasm; however, its occurrence primarily in the PPS itself is rare, reported in only about 1–2%[3].

## CASE REPORT

A 4-year-old boy presented with painless right parotid swelling of one-year duration. It was gradually increasing in size. There was no history of trauma or infection in the right parotid area. There was no facial asymmetry noticed, no feeding or speech difficulty.

Examination showed right parotid swelling, extending to the right submandibular region and measuring about 3 × 4 cm, without any skin changes (Fig. 1). The tumour was soft and non-tender. The facial nerve was intact. The throat showed medialisation of the right anterior pillar with the soft palate slightly pushed inferomedially. The Stenson duct was not inflamed and there was no pus discharge seen. The oral hygiene was good. Other examinations showed normal findings.

Contrast-enhanced computed tomography (CECT) scan of the neck showed a well-defined lobulated fat density mass measuring 6.6 cm (height) × 4.2 cm (width) × 4.8 cm (anterior-posterior) (HU: –75 to –90), occupying the superficial and deep lobe of the right parotid gland (Fig. 2). This mass pushed the lateral wall of the nasopharynx medially. There was no obliteration of the fat plane. No cystic or solid nodule. No internal calcification or dilated veins. No enlarged cervical nodes. The angle of the mandible was not eroded. Fine needle aspiration cytology (FNAC) was non-diagnostic. Magnetic resonance imaging (MRI) revealed a large and lobulated soft tissue mass in the right PPS, extending from the infratemporal fossa into the submandibular space. It measured about 4.2 cm (anterior-posterior) × 5.8 cm (width) × 4.9 cm (craniocaudal). It was hyperintense on both T1 and T2, and completely suppressed on fat suppression sequence. No obvious increased vascularity of the mass or the surrounding structures was found. The right PPS was obliterated. The nasopharyngeal wall was minimally compressed. The right pharyngeal wall was displaced medially, causing narrowing of the oropharynx to about 4 mm in width. The right submandibular gland was displaced inferiorly. The right parotid gland was compressed posterolaterally. The right carotid space was minimally displaced posteriorly. There were multiple small (<7 mm) deep cervical lymphadenopathies bilaterally. The clinical picture indicated a large right PPS lipoma with regional structural mass effects.

The patient underwent transcervical excision of lipoma under general anaesthesia. He was placed in a supine position with the head slightly extended and turned away from the surgeon. The parotid swelling was marked on the skin, and after local anaesthesia was given, horizontal transcervical skin incision was made two finger-breadths below the mandible, at the level of the hyoid bone. The subplatysmal flap was raised up to the lower border of mandible. The submandibular salivary gland and digastric muscle were identified. The submandibular gland

*Fig. 1. Right parotid swelling as seen from right lateral and anterior views*



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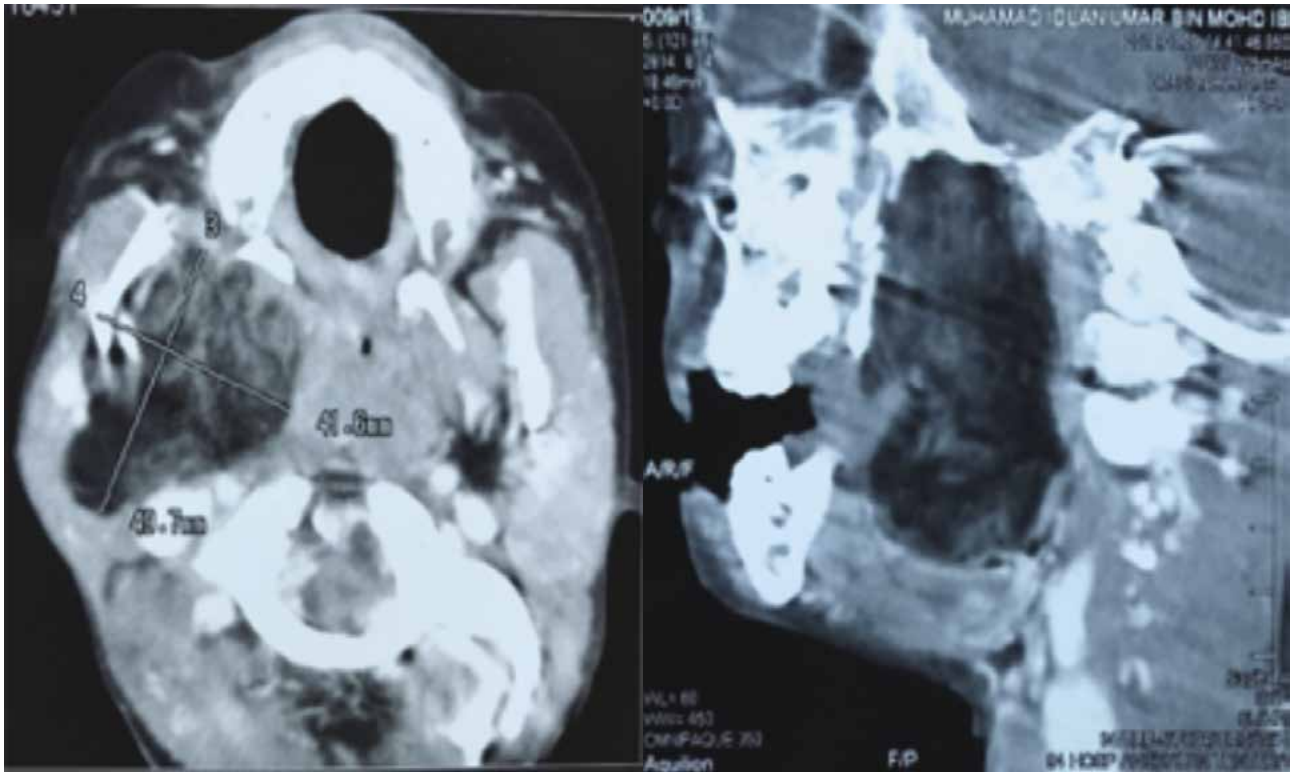

*Fig. 2. Axial and sagittal view of CECT neck, showing well circumscribed heterogeneous lobulated mass in the right parapharyngeal space with mass effect*

was normal, but it appeared pushed outwards by the mass effect of the huge lipoma beneath it. The anterior facial vein was ligated and divided. An incision was made subcapsularly on the submandibular gland, then it was retracted upwards to protect the marginal mandibular nerve. At the posteroinferior part of the gland, the facial artery was encountered, and it was ligated and divided. The hypoglossal nerve and the lingual nerve were identified and preserved.

To visualise the PPS content, the submandibular gland was retracted anteriorly, the posterior belly of the digastric muscle was retracted posteriorly and the mandible was retracted superiorly. The yellowish mass of the lipoma was already visible, and it was excised in a piece-meal fashion. Blunt finger dissection was done to separate the lipomatous mass from the surrounding structures, including the carotid artery. The bulk of the lipoma occupied the lower half of the PPS, while the upper half reached the base of skull. A small dissection area was just adequate to visualize the lipoma up to the base of skull, and haemostasis was achieved by ligation and bipolar electrocautery. After the tumour was cleared, the subplatysmal flap was replaced, vacuum drain was inserted, and the skin was closed in 2 layers. Two weeks after the surgery, the patient was seen well in the clinic, able to take normal diet as usual, and there was no new swelling seen at the operative site. Examination of the oral cavity showed medialisation of the right lateral pharyngeal wall.

The histopathological examination of the specimen revealed homogenous fatty tissue macroscopically, and mature adipocyte tissue separated by fibrous vascular septa microscopically. No evidence of malignancy was seen (Fig. 3). The right level 2 lymph node showed reactive lymphoid hyperplasia.

An outpatient follow-up up to 18 months after the surgery showed no sign of recurrence in the patient. The wound was well healed. There was no parotid swelling, and the examination of the oral cavity and oropharynx was unremarkable. The developmental milestones and growth are up to age.

## DISCUSSION

Preoperative diagnosis is important in planning investigations and surgical approach. FNAC can aid the diagnosis. When FNAC is ultrasound-guided or CT-guided, the accuracy can increase up to 87.5%[4]. It has a positive predictive value of 90% for benign tumours and 75% for malignant PPS lesions[5]. There is no study directly comparing guided with unguided FNAC results for a PPS tumour; however, Mondal and Raychoudhuri found that peroral FNAC diagnosis has 88.2% accuracy, with no false positive reports[6]. Therefore, incisional biopsy can be avoided in this complex anatomical area, while FNAC alone is adequate for suggesting the direction of management to the surgeon. According to current knowledge, per-oral biopsy is no more preferred due to difficult control of haemostasis, fibrosis at incision area which carries the risk of oropharyngeal fistula in the next surgery, as well as the risk of local recurrence associated with pharyngeal mucosa contamination with tumour spillage [7].



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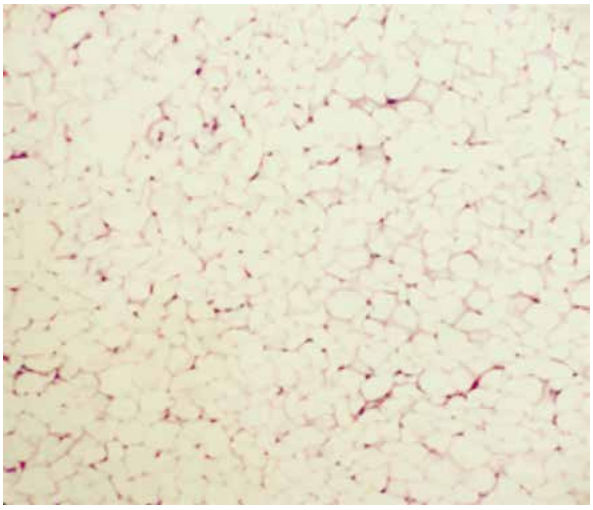

*Fig. 3. Histological image of the lipoma under H&E ×40, showing homogenous mature adipocytes cells, with intervening fibrovascular septae seen. The adipocytes were in distinctive lobular arrangement pattern with clear cytoplasm. No evidence of cellular atypia or metaplasia*

FNAC procedure in a paediatric patient can be troublesome, requires sedation for proper immobilization and it may not yield a significant aspirate. Due to the high anatomical variability and complexity, a PPS tumour poses diagnostic challenge. Unguided FNAC has been carried out transcervically and transorally; however, the results showed its limited role in establishing a correct diagnosis[8]. While another 10 years of a follow up study regarding the management of a PPS tumour showed that the positive predictive value of FNAC was 75% for benign tumours (3/4 were pleomorphic adenomas) and 100% for malignant tumours when using CT or ultrasound guidance[4].

Aspirates of simple lipomas yield small clusters of large univacuolated fat cells with regular nuclei. In contrast, liposarcoma aspirates contain lipoblasts and atypical nuclei and may demonstrate myxoid stroma and/or branching capillaries[9]. FNAC can differentiate lipoma from a liposarcomatous lesion; however, it is unable to accurately differentiate the subtype of liposarcoma itself[9].

CT and MRI are imaging modalities that can help differentiate lipoma and a non-lipomatous lesion due to the specific findings. Heterogeneity in adipocytic tumours suggests liposarcoma, while homogeneity indicates lipoma[9]. Both imaging techniques however will have the same capabilities to show hyper- or hypovascular tumour. Intraoperative frozen section is a very good method to establish a tissue diagnosis[2,8]. When a malignant lymphoma is encountered, the surgery will be abandoned, and the child can be sent for chemotherapy. However, when a surgical disease is encountered, a complete surgical resection is needed.

The diagnostic strategy in approaching a PPS tumour is firstly to request an imaging study (MRI is superior to CT in providing more information) and, after excluding vascular tumour, the surgeon can proceed with a few surgical approaches to PPS, leaving intra-operative biopsy as an option. When the contrast uptake is high during imaging, in line with suspicion of vascular tumour, further test might be considered such as MRA or arteriography, where one should think of possible embolisation as a primary therapy or a surgical adjunct[8]. In our case, the FNAC was done under sedation, unguided by imaging; however, the result was non-diagnostic aspirate. The CT scan, which should have been done prior to FNAC, showed a benign-looking PPS mass with high Hounsfield unit (HU) index, in correlation with fat density. The MRI was done subsequently, and showed a huge primary parapharyngeal lipoma, where the patients was planned for a surgical resection.

Surgical approach to solid PPS tumour can be either transoral or external. In paediatric population, where the oral cavity opening is smaller compared to that of an adult, the transoral approach is not really favoured due to poor visualisation and difficult control of haemostasis[3,10]. In late 80's, a study was done where the author advocated the use of transoral approach in adult patients presenting with a small benign PPS tumour, whereby the outcome showed safe and effective result, provided the lesion occupied more of lower PPS and was not palpable in the head and neck region[11]. However, their patient experienced recurrence rate of 25% after 5 years, which is a high rate possibly due to poor exposure by transoral approach[10]. A recent study suggested transoral approach, combined with a secondary trancervical approach, whereby their patients underwent transoral tumour excision preceded by a supplementary transcervical approach, whose role was to provide assurance for the dissection and safe-guarding the cranial nerves and neurovascular bundle, without any tumour spillage[12]. The follow up of 8 years showed no sign of tumour recurrence[12].

The external approach to PPS is divided into transcervical only, transparotid or transcervico-parotid approach, dictated by the bulk tumour location, tumour extension, tumour size, tumour relations to the surrounding structures as well as the type of the tumour that is either benign or malignant. In our patient, we decided for transcervical approach. The advantage is that the risk of injury to the facial nerve is lower and the cosmetic effect is better. However, the drawback of this approach is the slight difficulty faced during removal of the tumour at the base of skull due to the limited surgical access.

Out of all, the transcervico-parotid approach is the best as it provides wide surgical access, better visualization of the tumour margin as well as better control of major vessels[1]. Obviously, there will be a risk of injury to the facial nerve, but with precaution taken, this can be avoided. Other than that, a combined transcervical-parotid approach can provide greater surgical access and manipulation space in the case of a PPS tumour that occupies the skull base area. When parapharyngeal lipoma or other benign lesion, the specimen can be removed in pieces, in order to reduce the tumour bulk for adequacy of visualisation near the skull base[13]. Furthermore, in respect to the principles of



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oncological radicalism, the tumour should be excised in one block, in order to access the margin clearance and anticipating any need for radio/chemotherapy. Other literature reported the experience in using transcervical approach alone in removing the PPS tumour in toto[9]. The open incision usually will proceed with blunt finger dissection to remove the tumour bulk in PPS, keeping in mind all the major vessels and delicate nerves inside. In the other hands, there is less than 10% cases require additional mandibulotomy with tracheostomy in order to achieve adequate surgical field[14]. In the advance of technology, the surgeons nowadays need to be aware of the availability of other modalities in reaching the tumour in relation to its anatomic border, without much morbidities caused by surgical approach and technical access. As example, the use of combined endoscopic skull base surgery[13], laryngeal laser surgery[13] and transoral robotic surgery are proven to reduce morbidities[15]. The latter is for tumour located mainly in the prestyloid area. In cases of benign tumour that is ensheathed or lies close to major blood vessels, cranial nerves or base of skull, complete removal of the tumour can be attempted with major precaution. On the other hand, malignant tumours which grow infiltratively near these vital structures pose a great challenge for the surgeon in terms of achieving margin-free clearance.

With the current advancement in oncologic treatment, minimal residual tumours are possibly left behind to be treated with adjuvant chemotherapy or radiotherapy, thus avoiding operative morbidities and mortality[1]. In our case, the tumour homogeneously occupied the right PPS from the skull base to the infratemporal fossa and down to submandibular space, displacing other surrounding structures away. The transcervical approach exposed most of the tumour bulk, dissected bluntly in one piece. Few remaining pieces were removed near the skull base as well as those encircling the jugular vein and common carotid and its bifurcation. The surgeon or assistant should clear the surgical field from any blood as it may cause difficulty differentiating lipoma from the surrounding normal fatty tissue. Haemostasis was achieved and wound close primarily after vacuum drain placed.

The complication that can arise from these surgical approaches is the risk of injury to the nerve located inside the PPS or the facial nerve itself due to transparotid incision. Therefore, proper documentation of complete baseline cranial nerve examination is needed prior to the surgery. Apart from that, it must be ensured that there is blood reserved for the patient in case of major vessel injury.

## CONCLUSION

When diagnosing parapharyngeal mass in a paediatric patient, the preferred algorithm is first to raise a clinical suspicion of the nature of the tumour. Then, it is followed by imaging modalities to exclude vascular tumour, features of malignancy and to see the tumour dimension in this complex anatomy. FNAC prior to surgery or intraoperative frozen section are the choice available to establish tissue diagnosis. If the lesion is benign, the aim is to remove the tumour without compromising the surrounding structures, and the patient is always reviewed again to see any signs and symptoms of recurrence. However, in case of malignancy, the surgeon needs to weight between the risk the benefit and the quality of life in approaching the tumour, and the removal is either curative or palliative. There are multiple surgical approaches available for PPS, and all are tailored to the tumour size and location, in order to provide good surgical access with least complication possible.

### Conflict of interest

*The authors has no potential conflicts of interest.*

### Piśmiennictwo


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