

# Rare Tumours and Tumour-like Lesions of Pharynx That Cause Dysphagia

## *Farinksin Disfaji Yapan Tümör veya Tümör Benzeri Nadir Lezyonları*

Original Investigation  
Özgün Araştırmalar

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### Abstract

**Objective:** The aim of this article is to discuss rarely encountered tumours and tumorous masses at the different levels of pharynx that were diagnosed in patients admitted to our hospital with the complaint of dysphagia.

**Methods:** Twenty-one cases with masses causing dysphagia at the oropharyngeal, hypo-pharyngeal and laryngeal levels were identified among patients admitted to our hospital with the complaint of difficulty with swallowing between the years 2009 and 2011. Detailed anamneses and physical examinations were performed. In addition, for the purpose of diagnosis, the following were applied: Magnetic Resonance Imaging with and without contrast, Computerized Tomography, Three-Dimensional Computerized Tomography, Multi-

ple row-Detector Computerized Tomography, MR Angiography and 99m Tc Scintigraphy.

**Results:** Twenty-one patients, 12 women (57.1%) and 9 men (42.9%), were included in the study. Their mean age was 49.23±32.71 years old (min. 15-max. 80). There was partial obstruction at the oropharyngeal level in 11 cases; there was partial obstruction at the laryngeal and hypopharyngeal level in the other 10 cases.

**Conclusion:** A detailed history and examination along with endoscopic, radiological and laboratory evaluations are important for early diagnosis and management of rare tumours and tumour-like lesions of the oropharynx and neck causing dysphagia.

**Key Words:** Pharynx, tumour, tumour-like lesions, dysphagia

### Özet

**Amaç:** Bu makalenin amacı disfaji şikayetleri ile başvuran hastalarda tespit ettiğimiz, farinksin değişik seviyelerinde gözlenebilen çok nadir tümör ya da tümör benzeri kitleleri tartışmaktır.

**Yöntemler:** 2009-2011 yılları arasında hastanemize yutma güçlüğü şikayeti ile başvuran hastalar arasında disfajiye sebep olan orofaringeal, hipofaringeal ve laringeal düzeyde kitlesi bulunan 21 vaka teşhis edildi. Detaylı bir anamnez ve fizik muayene ile birlikte tanı amaçlı olarak kontrastlı-kontrastsız Manyetik Rezonans Görüntüleme, Bilgisayarlı Tomografi, 3 boyutlu Bilgisayarlı Tomografi, Multiple row-Detector Bilgisayarlı Tomografi, MR anjiyografi ve 99m Tc sintigrafi görüntülemeleri kullanıldı.

**Bulgular:** On ikisi kadın ve 9'u erkek 21 hasta çalışmaya dahil edildi. Yaş ortalaması 49.23±32.71 idi (min. 15-maks 80). On bir olguda, orafaringeal seviyede, kalan 10 olguda ise laringeal ve hipofaringeal seviyede parsiyel obstrüksiyon saptandı.

**Sonuç:** Endoskopik, radyolojik ve laboratuvar değerlendirmelerle birlikte detaylı hikaye ve muayene, disfajiye sebep olan orafarinks ve boynun az reastlanılan tümör ve tümör benzeri lezyonlarının erken teşhisinde ve tedavisinde çok önemlidir.

**Anahtar Kelimeler:** Farinks, tümör, tümör benzeri lezyon, disfaji



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## Introduction

The pharynx is located within the neck. It is part of both the respiratory and digestive systems since it serves as a conduit for both air and food. Anatomically, the pharynx is divided into three parts: nasopharynx, oropharynx and hypopharynx from cranial to caudal. Tumours arising from adjacent structures of the pharynx may indent the walls of the pharynx causing swelling and dysphagia. There are two types of dysphagia: pharyngeal and oesophageal dysphagia (1). Pharyngeal dysphagia is caused by difficulty in initiating the swallowing process, resulting in failure of bolus transfer from the mouth to the oesophagus. There are several pharyngeal neoplastic and non-neoplastic structural lesions such as hamartoma, inflammatory polyp, lipoma, haemangioma, lymphangioma, schwannoma and cervical hypertrophic osteoarthropathy, which cause dysphagia (2, 3). In this study, we present 21 lesions including rare tumours and tumour-like lesions of the pharynx and adjacent structures of the pharynx causing dysphagia.

## Methods

The medical files of the cases presented to our clinic with dysphagia between the years 2009 and 2011 were retrospectively reviewed. Dysphagia associated with neurological disease, central disorder, trauma or

surgery was excluded from the study. Twenty-one patients (12 females, 9 males) aged between 15 and 80 years old (mean age:  $49.23 \pm 32.71$  years) suffering from dysphagia caused by obstruction or compression at the pharyngeal level were included in the study (Table 1).

Detailed anamneses and physical examinations were performed. In addition, for the purpose of diagnosis, the following were applied: Magnetic Resonance Imaging (MRI) with and without contrast, Computerized Tomography (CT), Three-Dimensional CT (3DCT), Multiple row-Detector Computerized Tomography (MDCT), MR Angiography, and  $99m$  Tc Scintigraphy. All screening of the patients was conducted in the same clinic using the same devices. MDCT scans were performed using a Toshiba Aquilion 64° system at 120 KV and 75 mA with 5 mm intervals and a gantry tilt, a 2-second time. All images were captured using 1.5 T Philips Achieva Magnetic Resonance (MR) scanner using a clinical gradient system (30 mT/m, 150 mT/m/ms).

## Results

Eleven patients had obstructions at the oropharyngeal level. The causes of obstruction were as follows: soft palate-uvula pleomorphic adenoma in one case (Figure 1), fibrous dysplasia of

maxillary bone in one case, right parapharyngeal glomus vagale in one case (Figure 2), ectopic lingual thyroid in one case (Figure 3), antrochoanal polyps in one case, angiofibroma extending to oropharynx in one case, mass of the oropharynx in one case, aberrant internal carotid artery in three cases (Figure 4), and fibrous dysplasia at the level of C2 in one case (Figure 5).

Obstruction was at the level of the larynx/hypopharynx in 10 cases, including a laryngeal haemangioma (Figure 6), a bilateral carotid body tumour (Figure 7), proximal oesophageal and hypopharynx squamous cell cancer (Figure 8), interarytenoid stalked polyps, Killian Jameson diverticulum and left glomus caroticum, with one case of each, and diffuse idiopathic skeletal hyperostosis (DISH) in four cases (Figure 9). The levels of the lesions causing dysphagia are listed in Table 1.

Ten cases underwent surgery, in which post-operative relief of dysphagia was observed. Since radical surgery was refused by the patient who had epidermoid carcinoma located in the proximal oesophagus-hypopharynx, radiation therapy was recommended. Management involved follow-up in seven cases that refused the recommended surgery. Three of these were lost to follow-up.

The clinical results of the cases and the sizes of the lesions are presented in Table 2.

## Discussion

The pharynx is an anatomically and functionally complicated segment of the gastrointestinal tract. Anatomically, the pharynx is

**Table 1.** Levels of lesions that cause dysphagia

Lesions That Cause Dysphagia at the Level of the Oropharynx	Lesions That Cause Dysphagia at the Level of the Larynx and Hypopharynx
Pleomorphic Adenoma	Killian Jameson
Fibrous Dysplasia in Maxilla	Laryngeal Haemangioma
Fibrous Dysplasia in the C2 Vertebrae	Proximal Oesophageal Squamous Cell Carcinoma
Ectopic Lingual Thyroid	Glomus Caroticum
Mass of the Oropharynx	Diffuse Idiopathic Skeletal Hyperostosis (DISH)
Parapharyngeal Glomus Vagale	
Aberrant Internal Carotid Artery	
Antrochoanal Polyps	
Angiofibroma	



Figure 1. Sagittal T1-weighted MRI without (a) contrast and with (b) contrast imaging of a 59-year-old female patient revealed a pleomorphic adenoma located in the soft palate and uvula (white arrows)

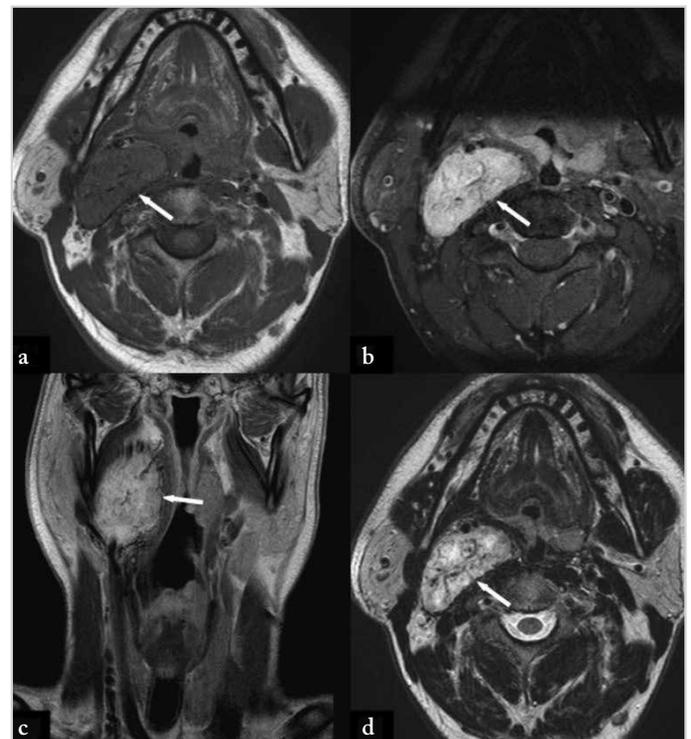


Figure 2. Axial T1- (a) and axial T2-weighted (b) without contrast and coronal (c) T1-weighted with contrast and axial (d) T1-weighted with contrast MR images of a 45-year-old female patient revealed right parapharyngeal glomus vagale (white arrows)



Figure 3. 99m Tc thyroid scan (a) of a 45-year-old female patient revealed functioning thyroid tissue at the base of the tongue (orange arrow) and absence of isotope uptake in the normal thyroid position; coronal (b), sagittal (c) and axial (d) MDCT with contrast images of the same patient demonstrated an oval-shaped mass secondary to ectopic thyroid at the base of the tongue causing sub-occlusion in the oropharynx (orange arrows)



Figure 4. Three-Dimensional MDCT (a, b) and coronal (c) MDCT Maximum Intensity Projection (MIP) images of a 67-year-old man showing medially deviated and tortuous bilateral internal carotid artery at second cervical vertebral level (white arrows)

divided into three parts: the nasopharynx, oropharynx and hypopharynx from cranial to caudal. The pharynx is a complex tube involved in respiration, speech and swallowing (1). Thus, pharynx dysfunction may result in choking, globus and dysphagia. Occasionally, dysphagia may result from extrinsic compression due to space-occupying lesions within the pharynx or neck (2-4).

Tumours of the parapharyngeal space are rare entities, representing 0.5% of all head and neck neoplasms. They are often associated with diagnostic and therapeutic difficulties due to various non-specific symptoms and the complex anatomy of the region. They are comprised of various neoplastic and non-neoplastic structural lesions such as salivary gland tumours, neurogenic

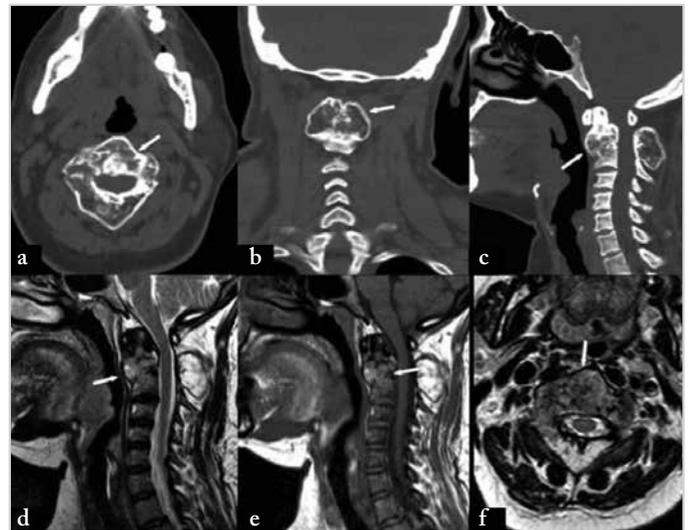


Figure 5. Axial (a), coronal (b) and sagittal (c) MDCT; sagittal T1- (d), T2- (e) and axial T2-weighted (f) MR images of a 50-year-old female patient showing the involvement of the vertebral body and posterior elements of the second cervical vertebra with an expanded and lytic lesion secondary to fibrous dysplasia. Extension of the C2 vertebral mass to hypopharynx caused dysphagia

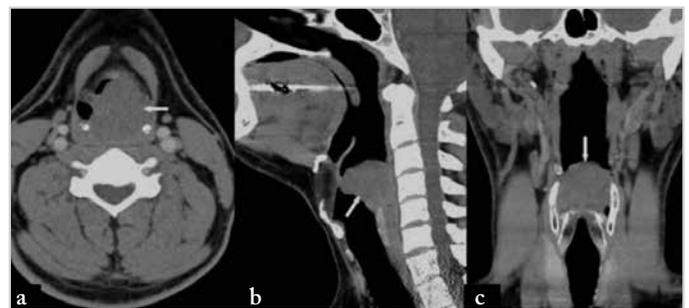


Figure 6. Axial (a), sagittal (b) and coronal (c) MDCT images of a 38-year-old man showing oval-shaped mass at the supraglottic level of the larynx causing sub-occlusion in the hypopharynx and larynx (white arrows)

tumours such as paraganglioma and schwannoma, hamartoma, inflammatory polyps, lipoma, haemangioma, lymphangioma, aneurysm, brachial cleft cyst, meningioma, chordoma, sarcoma and cervical hypertrophic osteoarthropathy (2-4).

Several different imaging modalities and specialized tests such as Video-Fluoroscopic Swallowing Study (VFSS), Fibre-optic Endoscopic Swallowing Study (FESS), Oesophagoscopy, Electromyography (EMG), Manometry-Manofluorography, Scintigraphy, Barium Swallow, Sonography, CT, MRI and Angiography are used for the diagnosis of dysphagia (5-8). Oesophagoscopy, Barium Swallow, Sonography, Scintigraphy, CT scan and MRI of the neck provided information about the adjacent pharyngeal anatomy and gave us valuable information on the character of the neoplastic and non-neoplastic structural oropharyngeal lesions.

The minor salivary glands are scattered throughout the hard and soft palate, lips, buccal mucosa, tongue, base of the mouth,



Figure 7. Coronal (a) and axial (b) MDCT; coronal 3D MDCT (c) and coronal MDCT Angiography (d) images of a 34-year-old woman showing bilateral huge oval-shaped masses at the carotid bifurcations splaying the external and internal carotid arteries (white arrows). The upper poles of the masses extend up to the level of the skull base. There are numerous central and peripheral enlarged feeding arterial branches within the bilateral carotid body tumours

tonsil, pharynx, retromolar trigone and nasal cavity (9, 10). Salivary gland tumours represent about 3% of all neoplasms, and tumours of the minor salivary glands comprise 22% of salivary gland neoplasms. Pleomorphic adenoma is the most common benign tumour of the salivary glands (11). Pleomorphic adenoma located in the uvula is extremely rare and causes dysphagia, speech disturbance and snoring. There are a few cases of pleomorphic adenomas located in the uvula (12). The pleomorphic adenoma diagnosed in our study was larger than all previously reported cases. It was located in the soft palate-uvula and caused dysphagia and sub-occlusion at the level of the oropharynx. It was treated by surgery.

A paraganglioma (glomus tumour) is a rare tumour that can be either unilateral or bilateral, and may originate from any of the neural ganglions in the body. Glomus tumours of the head and neck are associated with four primary locations including the jugular bulb, middle ear cavity, vagal nerve and carotid bifurcation (13). Carotid body tumours are the most common type, where glomus tympanicum tumours are one of the most rare glomus tumours of the head and neck. Carotid body tumours

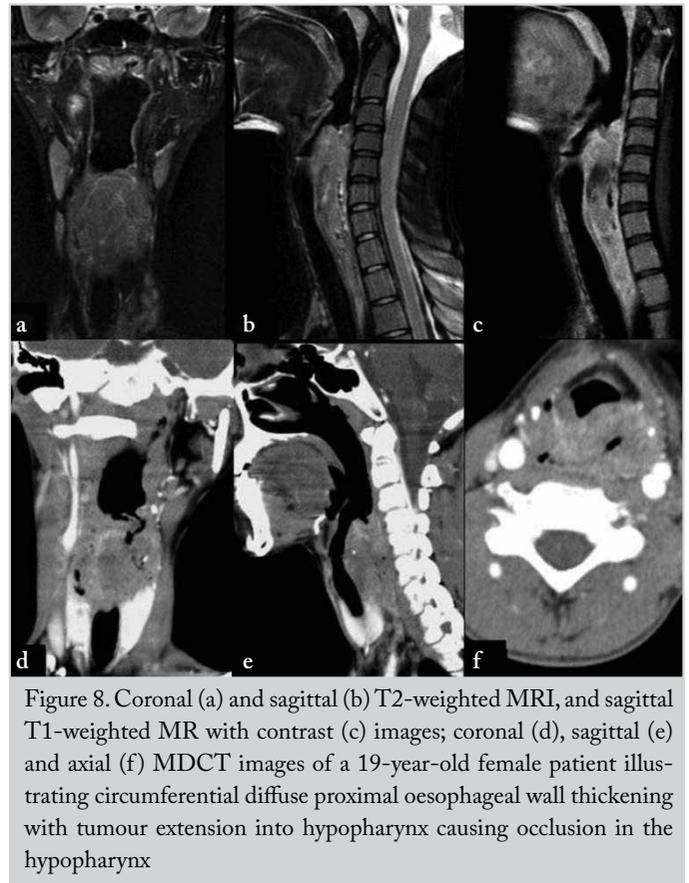


Figure 8. Coronal (a) and sagittal (b) T2-weighted MRI, and sagittal T1-weighted MR with contrast (c) images; coronal (d), sagittal (e) and axial (f) MDCT images of a 19-year-old female patient illustrating circumferential diffuse proximal oesophageal wall thickening with tumour extension into hypopharynx causing occlusion in the hypopharynx



Figure 9. Lateral cervical spine radiography (a), sagittal (b), axial (c) MDCT and sagittal MR (d) images of a 64-year-old male patient. Lateral cervical graphy (e) and sagittal MR (f) images of a 69-year-old male patient. Sagittal (g) and axial (h) MDCT images of a 67-year-old female patient showing giant cervical osteophytes in the ventral portion of the cervical spine and contiguous calcification of anterolateral cervical vertebral bodies. Large ‘crow-beak’-shaped osteophytes protruded anteriorly and impinged upon the posterior wall of the pharynx, causing dysphagia

develop at the bifurcation of the common carotid artery and arise from normal carotid body tissue. Glomus vagale tumours are located in the infratemporal region and along the course of the cervical vagus nerve (14). Two glomus caroticum cases in our study were located on the left side and were treated surgically.

**Table 2.** The clinical results and the lesion sizes of the patients

No.	Diagnosis	Gender	Age	Clinical results	Size (cm)
1	Soft Plate-Uvula Pleomorphic Adenoma	F	59	Operated	4.45x3.44x2.53
2	Right Parapharyngeal Glomus Vagale	M	45	Operated	6.18x4.80x2.55
3	Left Glomus Caroticum	M	37	Operated	8.53x6.85x4.04
4	Left Glomus Caroticum	F	55	Operated	4.8x4.1
5	Bilateral Glomus Caroticum	F	34	Lost to Follow-up	on right: 7.25x5.55x2.72 on left: 6.50x4.87x4.06
6	Ectopic Lingual Thyroid	F	46	Operated	3.70x2.69x2.64
7	Aberrant Internal Carotid Artery	F	30	Under Follow-up	-
8	Aberrant Internal Carotid Artery	M	67	Under Follow-up	-
9	Aberrant Internal Carotid Artery	F	78	Under Follow-up	-
10	Laryngeal Haemangioma	M	38	Operated	3.51x3.15x2.33
11	Proximal Oesophageal Squamous Cell Carcinoma	F	19	Radiotherapy	-
12	Diffuse Idiopathic Skeletal Hyperostosis (DISH)	M	64	Under Follow-up	-
13	Diffuse Idiopathic Skeletal Hyperostosis (DISH)	F	69	Under Follow-up	-
14	Diffuse Idiopathic Skeletal Hyperostosis (DISH)	F	67	Under Follow-up	-
15	Diffuse Idiopathic Skeletal Hyperostosis (DISH)	M	61	Under Follow-up	-
16	Fibrous Dysplasia in the C 2	F	50	Operated	-
17	Fibrous Dysplasia in Maxilla	F	16	Lost to Follow-up	-
18	Interarytenoid Stalked Polyps	M	32	Operated	1.6x2.1
19	Killian Jameson Diverticula	F	72	Operated	7.7x3.7
20	Extending To Oropharynx Angiofibroma	M	15	Operated	2.6x2.8
21	Mass of the Oropharynx	M	80	Lost to Follow-up	2.91x2.26

One case of glomus caroticum in our study was bilateral. They were very seldom and were large. The upper poles of the masses extended up to the level of the skull base. This case was lost to follow-up.

Large anterior osteophytes of the cervical spine are seen in diffuse idiopathic skeletal hyperostosis (DISH), which is also known as Forestier's disease. It is one of the rare aetiologies of oropharyngeal dysphagia. Anterior cervical osteophytes occur in 20-30% of the elderly population and 17% of patients with DISH are characterized by dysphagia (15). There are several mechanisms of dysphagia caused by osteophytes. These mechanisms include mechanical blockage, inflammatory reaction due to osteophytes and neuropathy owing to osseous impingement. Dysphagia of DISH may be treated either conservatively or surgically (16). Four cases of DISH causing dysphagia in our study only received follow-up.

Lingual thyroid, a developmental congenital anomaly, occurs as a result of abnormal embryological development of the thyroid tissue, with no thyroid tissue in the neck. The true incidence of lingual thyroid may never be known because approximately 1/18,000 to 1/100,000 live births are associated with ectopic thyroid tissue involving the tongue. The female:male ratio is 3/1-7/1 (17). One of our cases had ectopic thyroid tissue located in the tongue and causing dysphagia. There was no thyroid tissue in the neck and this case was only followed-up.

The congenitally tortuous and ectopic internal carotid artery is a very rare variation but it is an important anomaly for the otolaryngologist to recognize. Its symptoms may be dysphagia and malaise, and oral examination reveals a reddish, pulsatile, protruding mass in the lateral wall of the oropharynx. Computed tomography and magnetic resonance imaging of the neck provide spatial information about the adjacent pharyngeal anatomy, and demonstrate a vascular appearance and the ectopic portion of the ICA. These procedures are less invasive than an angiogram. The ectopic ICA poses a risk in major oropharyngeal tumour resection and other less extensive procedures, such as tonsillectomy, adenoidectomy and uvulo-palato-pharyngoplasty (18, 19). Our three cases were tortuous and ectopic internal carotid artery.

Killian-Jameson diverticulum is a paraoesophageal diverticulum originating from the Killian-Jamieson space in the anterolateral wall of the proximal cervical oesophagus (20). The incidence of Killian-Jameson diverticulum ranges between 0.01% and 0.11%; however the prevalence of Zenker's diverticulum is four times higher (21). Paraoesophageal diverticula are usually seen in the seventh and eighth decades (22). The most common symptom of paraoesophageal diverticula is dysphagia. Food regurgitation, halitosis, aspiration, oesophageal obstruction, ulceration, haemorrhage, trachea-oesophageal fistula and squamous carcinoma are among the other symptoms and complications. One of our cases was very seldom Killian-Jamieson diverticulum that caused serious dysphagia. It was treated surgically.

Laryngeal haemangiomas are benign vascular tumours that may be of two types, pediatric and adult. The pediatric type is more common and its incidence in infants is 4-5% (23). Laryngeal haemangiomas are very rare in adults, with an unknown incidence. Whereas pediatric haemangiomas usually occur in children younger than 2 months old, it can occur in adults at any age. It is usually located in the supraglottic region. Symptoms associated with laryngeal haemangiomas are dysphonia, dyspnoea and dysphagia. The haemangioma case in our study presented with dyspnoea and dysphagia. It was treated surgically.

Fibrous dysplasia is a skeletal bone developmental anomaly of unknown aetiology in which normal bone is replaced by abnormal fibro-connective tissue proliferation secondary to a defect in osteoblastic differentiation and maturation. It represents about 2.5% of all bone tumours and over 7% of benign tumours. One in three craniofacial fibrous dysplasias is localized in the craniofacial region. Regardless of gender, it usually manifests before the third decade of life. There are three types: monostotic, polyostotic and McCune Albright syndrome (24, 25). Our two cases were of the monostotic type. One of them was localized in the maxillary bone and the other was localized in the second cervical vertebra. They caused dysphagia secondary to mass effect in the pharyngeal region. They were treated surgically to relieve symptoms.

The other cases causing dysphagia were laryngeal interarytenoid stalked polyps (one case), antrochoanal polyps (one case), angiofibroma (one case) and squamous cell carcinoma (one case). All caused dysphagia with mass effect. Some of them were localized in the pharynx and some in the parapharyngeal spaces and extending to the pharynx. The squamous cell carcinoma was localized in the proximal oesophagus and infiltrated the hypopharynx. The interarytenoid stalked polyps, antrochoanal polyps and angiofibroma were treated surgically. The squamous cell carcinoma was treated with radiotherapy.

## Conclusion

The pharynx is an anatomically and functionally complicated segment of the gastrointestinal tract. Dysphagia is very common and is currently considered as an alarm symptom that requires immediate evaluation. Dysphagia affects the quality of a patient's life and may lead to dehydration, malnourishment and aspiration pneumonia. A detailed history and examination along with endoscopic, radiological and other investigations are crucial for early diagnosis and management. Radiologists and clinicians should be familiar with normal and abnormal findings as well as postoperative radiographic findings of the pharynx. They should also be aware of the rare anatomical variants. Knowledge of the differential diagnosis of these lesions and proper preoperative investigations will help early diagnosis and prevent patients from being subjected to unnecessary surgical procedures.

## Conflict of interest

No conflict of interest was declared by the authors.

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