The Lipomatosis of the Parapharyngeal and Retropharyngeal Space: A Case Report

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SUMMARY
Introduction Lipomas are the most common benign mesenchymal tumors, which account for almost 50% of all soft-tissue tumors.
Case Outline The case of a 75-year-old patient with a slow growing lesion of para- and retropharyngeal space was reported. The patient was suffering from progressive dysphagia, respiratory obstruction and sleep apnea.
Conclusion An external surgical approach is the treatment of choice. Etiology, differential diagnosis and therapy of head and neck lipomas has been discussed.
Keywords: lipoma; parapharyngeal space (PPS); retropharyngeal space

INTRODUCTION

Lipomas are slow growing expansive benign tumors. They are the most common benign mesenchymal tissue tumors accounting for almost 50% of all soft tissue tumors with an incidence of almost 1% of the population [1]. They occur more often subcutaneous or submucosal, and rarely intramuscular or in the abdominal cavity organs. Only 13% of them are localized in the head and neck region. Unusually, lipomas affect internal auditory meatus, oral cavity and cerebellopontine angle [2]. Upper respiratory tract lipoma is uncommon. These tumors can cause dysphagia, respiratory distress as well as obstructive sleep apnea [3].

CASE REPORT

A 75-year-old male was admitted to the ENT Department of Wroclaw Medical University with a few years history of slow growing tumor of para- and retropharyngeal space. Progressive dysphagia and respiratory obstruction was present. The tumor caused sleep apnea. Head and neck examination with fiberoptic laryngoscopy revealed a soft tissue mass in the pharynx involving hypopharynx (laryngopharynx) on the right side. The tumor was causing a bulge of the lateral right side of the neck (Figure 1). Head and neck computed tomography (CT) scans revealed a smoothly marginated fat density mass (from -100 to -50 HU) localized forward and laterally from the right long neck muscle. The upper part of the tumor within the nasopharynx reached just below the fold of the Eustachian tube, in the lower part ranged the angle of the jaw with total dimensions 85×58×72 mm. The tumor had a mass effect with displacement of adjacent anatomical structures, especially cervical vessels (internal jugular vein and internal carotid artery) shifted forward and outward. The lesion expanded prevertebral space and reached on the left side about 1 cm behind the median line of the body. There was no evidence of bone destruction in the tumor vicinity. There were no pathologically changed lymph nodes (Figure 2). The lipoma with an expanding type of growth was suspected. The differential diagnosis between lipoma and liposarcoma favored lipoma as CT scan did not prove soft tissue invasion. The patient was qualified for surgery by the interdisciplinary team of ENT surgeon and maxillofacial surgeon.

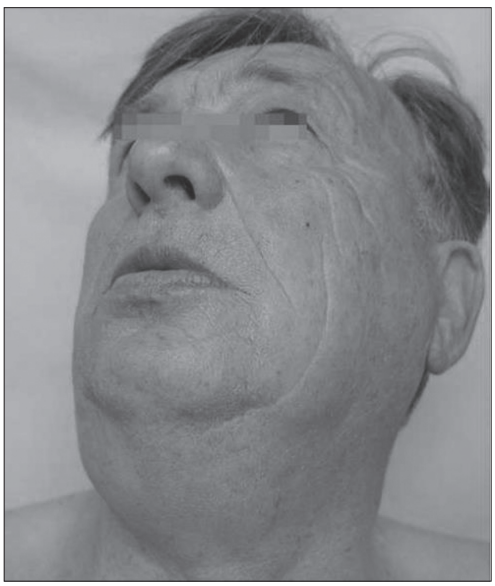

Figure 1. The size of the neck lipoma in a 75-year-old man

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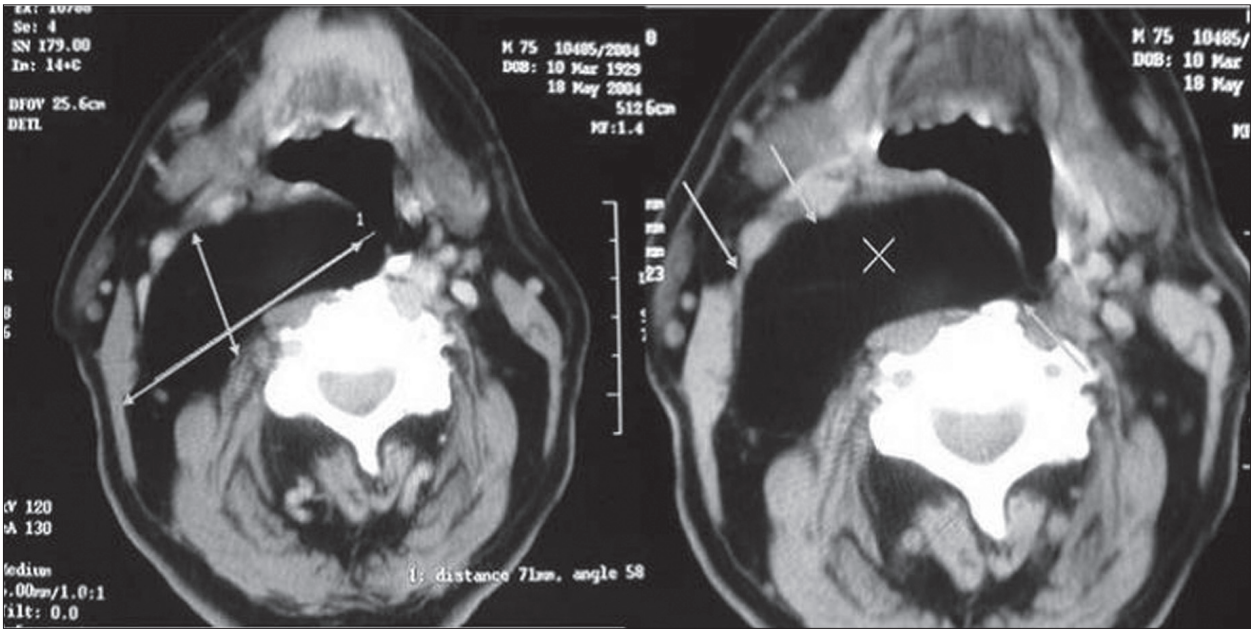

Figure 2. CT scans showing lipoma with total dimensions 85×58×72 mm

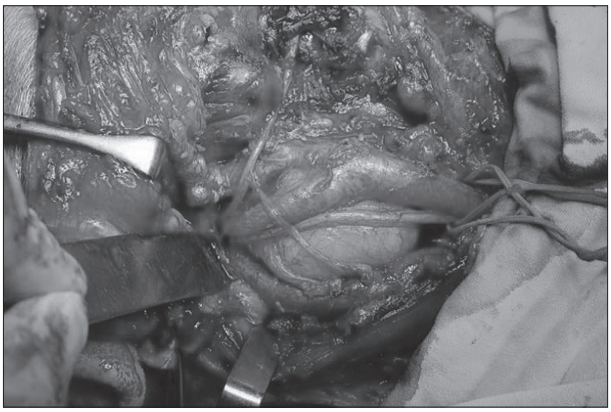

Figure 3. The surgical view of lipoma

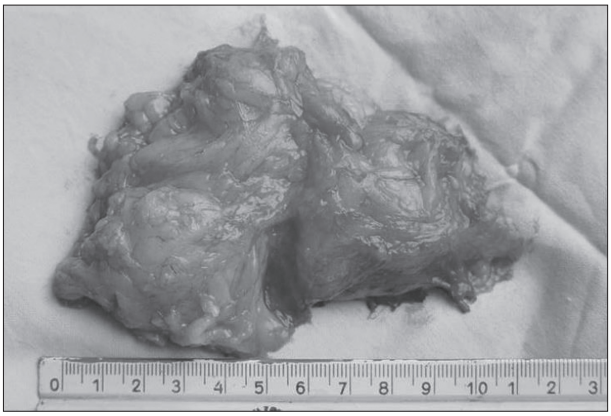

Figure 4. The size of an excised lipoma

In general anesthesia, in a natural skin crease, lateral cervical approach was used to expose the jugular vein, common, external and internal carotid artery, the vagus and the sympathetic trunk nerves: IX, XI, XII (Figure 3). A large tumor was located in the parapharyngeal space, inferior to the level of the hyoid bone and superior to the base of the skull, involving submandibular gland, under named above structures and between cervical muscles. The non-capsulated mass tumor was excised totally (Figure 4). The submandibular gland was also removed due to tumor invasion (so-called infiltrative type of lipoma). The pathological examination revealed lipoma in the tumor, and lipomatosis in the submandibular gland. The postoperative course was uneventful. There was no need for nasogastric tube nor temporary tracheostomy. On the 14th month the clinical follow-up revealed no signs of the disease. The long term result of the surgical treatment is excellent without any morphological or functional impairments.

DISCUSSION

Lipomas are composed of mature adipocytes. All cells of the tumor are well-differentiated and seem to contain abundant fatty material. They are usually well-circumscribed lesions surrounded by a thin capsule separating them from the remaining tissue. They have a greasy consistency and are yellow in color. Clinically, pain starts to appear with tumor growth and pressure on angioneurotic structures. Single lipomas are more frequent in females aged 40–60 years. However, multiple lipomas are more often in males. They can also occur in syndromes: hereditary multiple lipomatosis, familial multiple lipomatosis (FML), benign symmetric lipomatosis (BSL), also known as Madelung's disease or Launois–Bensaude syndrome, Gardner's syndrome or Dercum's disease [4]. The presence of benign, encapsulated, restricted, subcutaneous tumor(s) is usually sufficient for suggesting a diagnosis. Management of lipomas does not usually pose a problem. However, problems arise in rare cases of multiple lipomas and lipomatoses which develop sporadically or in association with other

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diseases and syndromes. These present as enormous fatty masses, non-capsulated, penetrating and infiltrating the adjacent structures, often painful and involving an extensive body area (benign symmetric lipomatosis / BSL, Dercum's disease, Gardner's syndrome). Lipoma should be differentiated with BSL, liposarcoma, FML, simple obesity or obesity caused by steroids. The treatment of choice is the complete surgical excision. Lipectomy or liposuction can be especially helpful for patients with severe cosmetic deformity, especially in case of any airway compromise. Complete excision is important to avoid local recurrence, which is present in 5% of cases [4, 5, 6]. In the study of Oliai et al. [7] from Baltimore, only one out of 24 neoplasms in the parapharyngeal space was lipoma. The treatment of choice in case of laryngeal lipoma seems to be an external surgical approach for complete surgical removal of a large lipoma. In our case only this approach allowed the radical dissection of the tumor. Lipomas of the oral cavity are usually single. However, multifocal lipomas of tongue are incidentally reported [8]. In the literature there are few reports on the malignant neoplasia of lipoma converting into liposarcoma [9, 10, 11]. The onset of multiple lipomas coinciding with cytotoxic chemotherapy has been described by Cronin et al. [12].

In the diagnosis of primary tumors of the parapharyngeal space, the growths of connective tissue and fat should be considered. In the presented patient clinical symptoms and tumor location excluded BSL.

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Липоматоза парафарингеалног и ретрофарингеалног простора: приказ болесника

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КРАТАК САДРЖАЈ

Увод Липоми су најчешћи бенигни мезенхимални тумори, који чине скоро 50% свих тумора меког ткива.

Приказ болесника Приказан је 75-годишњи болесник са спорорастућом лезијом парафарингеалног и ретрофарингеалног простора. Болесник је патио од прогресивне дисфагије, опструкције дисајних путева и апнеје у сну.

Закључак Метода избора је спољашњи хируршки захват. Разматране су етиологија, диференцијална дијагноза и лечење липома главе и врата.

Кључне речи: липом; парафарингеални простор; ретрофарингеални простор

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