RECURRENT LIPOMATOUS TUMOR OF HYPOPHARYNX: CASE REPORT AND LITERATURE REVIEW

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SUMMARY – Liposarcoma is one of the most common soft-tissue sarcomas in adults, but head and neck are rarely involved, especially regions of the larynx and hypopharynx. According to Enzinger and Weiss, liposarcoma can be divided into 5 subtypes: well-differentiated, myxoid, round cell, pleomorphic and dedifferentiated. We present an unusual case of well-differentiated liposarcoma of the hypopharynx in a patient with previous three procedures of endoscopic removal of hypopharyngeal tumor classified as benign lipoma. Well-differentiated liposarcoma is a tumor of low-grade malignancy, which frequently recurs locally, but does not metastasize. Wide tumor resection with free margins is mandatory. Immunohistochemistry is a useful diagnostic tool. We also discuss recently published literature on this unusual presentation of well-differentiated liposarcoma.

Key words: *Hypopharyngeal neoplasms; Lipoma; Liposarcoma; Differential diagnosis; Case reports*

Introduction

Liposarcoma is one of the most common soft tissue sarcomas in adults, with the reported incidence varying from 5% to 30% of all soft-tissue sarcomas[1]. Different tumor sites have been reported including unusual presentation such as primary mesenteric liposarcoma[2]. The head and neck region is involved in only 3% to 6% of all liposarcoma cases[3]. Well-differentiated liposarcomas are tumors of low-grade malignancy that may recur locally but do not metastasize. We present a surgically treated male patient with well-differentiated hypopharyngeal liposarcoma with previous endoscopic removals of hypopharyngeal tumors.

Case Report

A 63-year-old male patient was admitted to our Department with symptoms of acute severe dyspnea. In the last 3 months, he had experienced progressive dysphagia. Available medical records revealed that the patient had undergone endoscopic removal of the recurrent hypopharyngeal tumor on three occasions in the past 23 years. These procedures were performed in other clinical institutions, thus complete documentation was not available. According to the available documentation and histologic revision of specimens, the tumor was classified as lipoma (Fig. 1).

After surgical tracheostomy, direct microlaryngoscopy revealed a massive pink-yellowish tumor that filled both piriform sinuses and veiled laryngeal aditus. Because endoscopic removal of the tumor was impossible due to its large size, lateral pharyngotomy was performed. During the surgery, a cluster-like tumor that filled both piriform sinuses was seen. The tumor had attachment (1.5x2 cm) to the lateral wall of the right piriform fossa. Complete removal of the tumor was done. The hypopharyngeal mucosa defect was primarily reconstructed.

The excised tumor was lobulated, gray-yellow tumor, 4.6x3.2 cm in size. Histologic examination of

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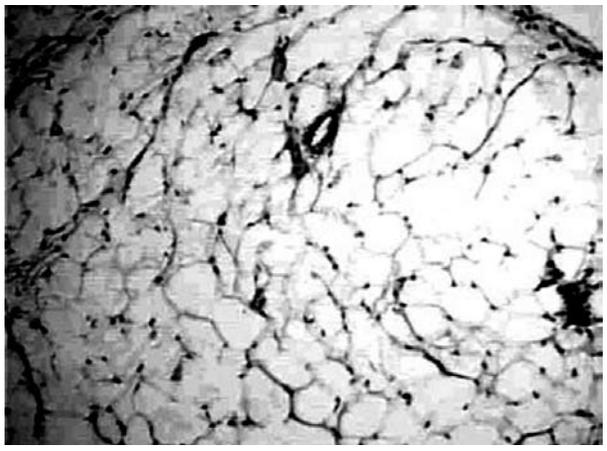

Fig. 1. Lipoma (H&E, X100): mature fat cells showing only slight variation in cellular size and shape.

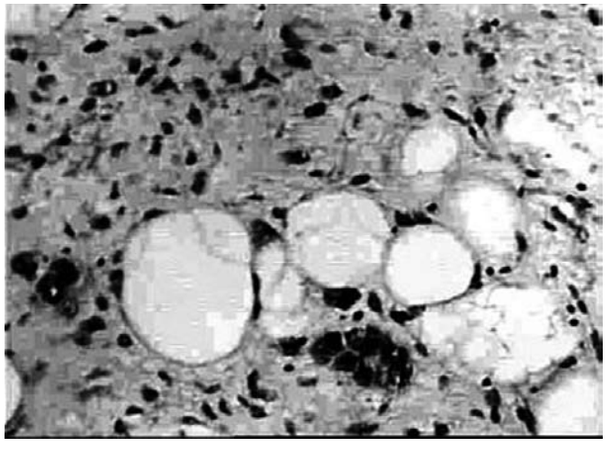

Fig. 2. Liposarcoma (H&E, X200): well-differentiated liposarcoma with pleomorphic areas.

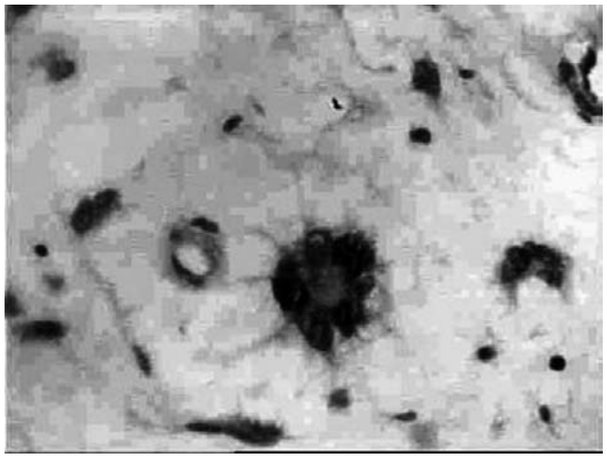

Fig. 3. Liposarcoma (H&E, X400): characteristic multivacuolated and multinucleated lipoblast.

the specimen revealed that the tumor was composed of lipoblasts and rare immature lipocytes. Lipoblasts were mainly multivacuolated with large irregular hyperchromatic nuclei (Figs. 2 and 3). Squamous nonkeratinizing epithelium overlying the tumor had normal appearance. Immunohistochemistry was positive for S-100 protein, vimentin and p53. This tumor was classified as well-differentiated liposarcoma with scattered pleomorphic areas.

Consequently, the patient was decannulated. Five years later, there are no signs of local or regional recurrence of the disease.

Discussion

Liposarcoma of the hypopharynx is a very rare tumor difficult to diagnose. According to Enzinger and Weiss, liposarcoma can be divided into 5 subtypes: well-differentiated, myxoid, round cell, pleomorphic and dedifferentiated, or in three subtypes according to its clinicopathologic and molecular characteristics[1,4].

Until 1954, there was no single case of liposarcoma reported above the clavicles[5]. In 1975, Miller *et al*. reported the first case of liposarcoma of the larynx[6]. Most patients with head and neck liposarcoma are adults in the 4th to 6th decade of life. The youngest patient (28-year-old) with laryngeal liposarcoma has been reported by Shah and Lowry[7]. Male patients account for approximately 55% to 60% of all head and neck region liposarcomas[3,8].

Due to the presence of a slow growing tumor in the hypopharynx, liposarcoma can cause progressive dysphagia and dyspnea. Wenig *et al*.[3] have described similar clinical features in patients with laryngeal liposarcoma. Baj *et al*.[9] describe a case of giant liposarcoma of the hypopharynx that encroached the larynx causing airway obstruction, so the patient had to lean forward in order to be able to breathe.

Macroscopic and histologic appearance within well-differentiated liposarcoma may vary. Within lipoma-like liposarcoma, areas of well differentiated adipocytes can be seen. This suggests the necessity of wide tumor resection with free margins. Histopathologic examination of a large number of specimens and immunohistochemical analysis provide definitive diagnosis and distinction from benign lipoma.

In the majority of cases, both lipoma and liposarcoma have a tendency to recur even after surgical

removal. Recurrent tumor may have a histologic appearance that differs from the primary tumor.

In recently published literature, atypical lipomatous tumor/well-differentiated liposarcoma is associated with amplification of the MDM2 gene. Although the diagnosis of this tumor is based on histologic aspect, this molecular genetic alteration allows for differentiation from benign lipomatous tumors. Sampling, immunohistochemistry (IHH) and fluorescence in situ hybridization (FISH) for MDM2 and CDK4 are useful for diagnosis[10].

Well-differentiated liposarcomas frequently recur locally[11]. The risk of lymph node metastasis is very low, so neck dissection is not indicated. There are no reports on distant metastases of well-differentiated liposarcoma. Metastases are signs of the dedifferentiated component[10].

Surgical therapy remains the mainstay of treatment[12,13]. Reed and Vick report that the average time between surgery and recurrences was 69 months[14]. Hurtado *et al.* report on endoscopic (direct laryngoscopy) removal of a well-differentiated laryngeal liposarcoma 18 mm in diameter[15]. There are controversial opinions on postoperative radiation therapy, although it may be beneficial, especially after excision of well-differentiated liposarcoma recurrence[8,13,16].

We presented an unusual case of a patient with recurrent pharyngeal tumor. Our patient had transcervical removal of the well-differentiated liposarcoma tumor with 3 previous procedures of endoscopic removal of slow growing hypopharyngeal tumor classified as benign lipoma. Histologic appearance of these two tumors can be similar. This underlines the significance of complete surgical removal of the tumor and IHH.

Cervical approach proved successful in this case. In selected cases, endoscopic approach is possible, but in most large tumors wide excision with surgical margins of healthy tissue is a key to complete surgical resection[17]. Both techniques have indications for use, which depend not only on the size of the tumor, but also on the tumor site, accessibility and anatomical relations or limitations of each patient.

Conclusion

Lipoma and liposarcoma are rare tumors in the region of hypopharynx. Both these tumors have a tendency to recur. Complete removal is essential for local control of the disease, which can be achieved through lateral pharyngotomy or through less invasive endoscopic approach. IHH is useful for correct histopathologic diagnosis. Long term follow up is necessary.

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Sažetak

OPETOVANI LIPOMATOZNI TUMOR HIPOFARINKSA: PRIKAZ SLUČAJA I PREGLED LITERATURE

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Liposarkom je jedan od najčešćih mekotkivnih sarkoma u odrasloj dobi. Usprkos tome, zahvaćenost glave i vrata je rijetka, osobito u regijama larinksa i hipofarinksa. Prema Enzingeru i Weissu, liposarkom se može podijeliti u 5 podtipova: dobro diferenciran, miksoidni (miješani), tumor kružnih stanica, pleomorfni i nediferencirani. Mi predstavljamo neobičan slučaj bolesnika s dobro diferenciranim liposarkomom u hipofarinksu, koji je prije bio tri puta endoskopski operiran, a svaki je put odstranjeni tumor bio dijagnosticiran kao benigni lipom. Dobro diferencirani liposarkom je tumor niskog malignog potencijala, koji često nakon toga opetovano raste, ali ne daje metastaze. Široka resekcija tumora sa slobodnim rubovima je metoda prvoga izbora. Imunohistokemijska analiza je od osobitog dijagnostičkog značenja. Također, u radu se raspravlja o podacima iz novije literature glede neobičnih prikaza dobro diferenciranih liposarkoma hipofarinksa.

Ključne riječi: *Hipofaringealni tumori; Lipom; Liposarkom; Diferencijalna dijagnoza; Prikazi slučaja*