Solitary fibrous tumor of the larynx treated with transoral endoscopic CO2 laser surgery – a case report and literature review

Samotny guz włóknisty krtani usunięty metodą endoskopową przy użyciu lasera CO2 – opis przypadku i przegląd literatury

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ABSTRACT:

Introduction: Solitary fibrous tumors of the larynx are very rare mesenchymal tumors most frequently located within the upper part of the larynx. They are usually benign, slowly growing, and not prone to infiltrate the surrounding tissues. Distant metastases are rarely reported. The diagnosis is based on histopathological examination with immunohistochemical assays. The treatment of choice consists in surgical removal of the lesion within the resection margin of macroscopically and microscopically healthy tissues. Acceptable surgical approaches include either endoscopic or external access surgery in cases of larger lesions.

Case report: A case of a 46-year-old male patient with left vestibular tumor of the larynx is presented. Complaints at presentation included hoarse throat and globus sensation. Imaging studies were performed. CT angiography revealed a richly vascularized tumor supplied from the upper laryngeal artery. Embolization of the upper left laryngeal artery was performed prior to the procedure. Total endoscopic resection of tumor was performed using a CO2 laser with good laryngeal function being maintained. The final histopathological diagnosis was that of solitary fibrous tumor. Since negative resection margins were achieved, the patient did not require further treatment and was qualified for periodic laryngological follow-up alone.

KEYWORDS:

benign neoplasm of the larynx, CD34 (+), laser CO2, mesenchymal tumor, solitary fibrous tumor

STRESZCZENIE:

Wstęp: Samotny guz włóknisty (SFT) krtani jest bardzo rzadkim nowotworem mezenchymalnym, najczęściej lokalizującym się w górnym piętrze krtani. Zwykle ma łagodny charakter, wykazuje powolny wzrost i nie ma skłonności do naciekania okolicznych tkanek. Rzadko daje przerzuty odległe. Rozpoznanie stawiane jest na podstawie badania histopatologicznego z oceną immuno-histochemiczną. Leczenie z wyboru to chirurgiczne usuwanie zmiany w marginesie makroskopowo i mikroskopowo zdrowych tkanek. Akceptowana jest zarówno chirurgia z dojścia zewnętrznego w przypadku zmian o dużych rozmiarach, jak i chirurgia endoskopowa.


SŁOWA KLUCZOWE: CD34 (+), laser CO2, łagodny nowotwór krtani, nowotwory mezenchymalne, samotny guz włóknisty

ABBREVIATIONS

HPF – high power fields
SFT – Solitary fibrous tumor
TORS – transoral robotic surgery
WHO – World Health Organization
INTRODUCTION

Solitary fibrous tumor (SFT) is a rare mesenchymal tumor [1–3], first described by Klemperer and Rabin in 1931 [1–10]. Most frequently, it derives from serous membranes, particularly pleura [11–15]. Although extrapleural locations are less common, reports on SFTs located e.g. within the upper gastrointestinal tract, genitourinary system, thyroid, nasopharynx, paranasal sinuses, orbits, salivary glands, lacrimal sacs, trachea, liver, lungs, mediastinum, and larynx can be found in the literature [1, 2, 6, 7, 11, 12, 14–17].

SFTs are mostly benign and well-differentiated from their environment. Sometimes, local recurrences may be encountered; this pertains particularly to large-sized tumors with malignant features or cases in which R0 resection had not been achieved. Distant metastases are rarely reported [6].

The diagnosis is based on histopathological examination with immunohistochemical assays. In most cases, laryngeal SFTs are benign and require surgical treatment alone [4, 5, 12]. Laryngeal SFTs are most frequently located within the upper part of the larynx [3]. The surgical access route depends on the size and location of the tumor. Available approaches include endoscopic surgery, external access surgery, and transoral robotic surgery (TORS) [13].

CASE REPORT

A 46-year-old male patient presented at the local Laryngological Clinic in November 2020. Main complaints included globus sensation and periodical hoarse throat. In the past, the patient underwent a left vocal fold polyp removal procedure. The patient had quit smoking three years before presentation; prior to quitting, he had used to smoke about 15 cigarettes per day; he also reported his alcohol consumption to be occasional.

Laryngological fiberoscopic examination revealed smooth tumor, approximately 3 × 3 cm in size, dislocating the left vestibular fold. Rima glottidis was respiratorily efficient. No significant abnormalities were observed in anterior rhinoscopy, otoscopy, and laryngeal endoscopy. Cervical lymph nodes were impalpable.

A CT scan performed in November 2020 revealed an oval, well-delineated and strongly enhancing structure sized 2.1 × 2.8 × 2.7 cm, located tangentially to the posterior surface of the thyroid cartilage on the left, and dislocating the left vestibular fold and the laryngeal pouch to the right. A vascular peduncle supplying the tumor lesion was reported on its anterior surface. No infiltration of laryngeal cartilages was observed. Isolated lymph nodes, sized from 1.1 × 1.2 cm, were visualized within the soft tissues of the neck. Otherwise, no pathological findings were reported.

The patient was referred for an angio-CT scan to rule out the vascular nature of the lesion.

A lesion as reported in the previous imaging study was described in the examination report, with very good tumor vascularity as an additional finding (Fig. 1.–3.). Due to the significant risk of bleeding, the patient was referred for a procedure involving embolization of the tumor-supplying vessel prior to surgery.

Selective angiography of the left superior laryngeal artery was performed at the 2nd Voivodeship Specialist Hospital in Jastrzębie Zdrój to reveal extensive pathological vascularization of the tumor. Due to the tortuosity and the small size of the vessel (1.5 mm in diameter), a 1.5F MARATHON microcatheter was introduced for proximal insertion of embolization coil. Follow-up angiography revealed the rate of the flow within the tumor-supplying superior laryngeal artery...
Malignancy is observed for about 13–23% of tumors in pleural locations. These tumors are much more likely to cause local recurrence (9–19% of cases) as well as more likely to form distant metastases (19%). Non-pleural SFTs are characterized by a statistically slower rate of growth, and low likelihood of malignant transformation; distant metastases are formed in only 6–10% of cases [1].

Differential diagnostics should include other tumors such as schwannomas, angiomas, paragangliomas, pericytic hemangiosarcomas, neurofibromas, histiocytic fibromas, leiomyosarcomas, fibrosarcomas, and desmoid tumors [5, 14].

Head and neck SFTs account for about 6% of all diagnosed SFTs. The most common primary tumor sites within the head and neck region include the nasal cavity and paranasal sinuses, orbits, mouth, salivary glands, and deep soft tissues of the neck [18]. Solitary fibrous tumor is extremely rare in laryngeal locations [4, 5, 15]. Most frequently, it develops within the upper floor of the larynx [1, 16, 18].

Symptoms presented by patients are not specific, and depend on the size and the location of the tumor. Patients can report hoarse throat, globus sensation within the larynx or throat, distorted voice, cough, and shortness of breath [4, 11, 13, 15–17].

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The final histopathological diagnosis was that of the SFT. R0 resection was achieved during the surgery. In addition, immunohistochemical assays revealed the following statuses: CD34(+), bcl2(+), S100(−), SMA(−), VIM(+), CK(−), Ki67 = 5%. The mitotic index was 1MF/10 HPF. The lesion was shown to have a low risk of metastasis.

Consequently, no adjuvant treatment was required and the patient has since then remained subject to periodic laryngological follow-up alone in local outpatient setting.

The interior of the larynx was assessed during a follow-up visit. Fiberoscopic examination revealed proper healing, with vocal folds presenting with smooth surfaces and proper motility upon phonation and breathing. The patient feels good, declaring complete resolution of complaints.

DISCUSSION

Primary spontaneous fibrous tumors of the head and neck are rare spindle cell tumors of mesenchymal origin [18]. According to the World Health Organization (WHO) classification, they belong to the group of fibroblastic/myofibroblastic neoplasms [3, 7].

The diagnosis of SFT is based on histopathological examination with immunohistochemical staining. Upon microscopic assessment, SFTs present with unarranged spindle cells with elongated nuclei. The tumors are positive for the expression of CD34, vimentin, and bcl-2 while being negative for the expression of S-100 and alpha-SMA [6, 7, 9, 10, 12, 14, 16].

SFTs are most often characterized by slow growth, not accompanied by infiltration of surrounding tissues, with distant metastases being rarely reported [5, 17]. However, some SFTs may be malignant, infiltrate the surrounding tissues, cause metastases and/or local recurrences [6]. The malignant lesions are usually larger and present with increased mitotic activity, necrosis or ecchymosis, and high degree nuclear atypia in histopathological examinations [1, 3, 9, 10].

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In endoscopic testing, a nodular bulge within the larynx, covered with smooth, unremarkable mucosa, is usually observed. In all cases, diagnostic examinations should be extended to include contrast-enhanced imaging studies to assess the extent of the tumor and potential infiltration of the surrounding tissues [5, 9, 13, 16].

SFTs are more common in male as compared to female subjects (M:F ratio of 6:1) [1, 15].

The treatment of choice involves surgical resection of the tumor with R0 margin and possibly best conservation of laryngeal function [5, 9, 11–13, 15, 17]. Endoscopic access is recommended if the inside of the larynx and the tumor can be well visualized, if no laryngeal sphincter involvement is present, and if the lesion does not extend beyond the anatomical boundaries of the larynx [1, 13, 18].

In some cases, when the histopathological result is suggestive of a potentially malignant character of the lesion or when positive resection margins are obtained, adjuvant chemo- or radiotherapy should be considered [10, 14] However, adjuvant therapy is rarely indicated in cases of solitary fibrous tumors of the head and neck region [12].

Long-term, periodic follow-up is required following surgery [1, 4, 6, 17, 18].

SUMMARY

Presented is a case of an exceptionally rare laryngeal tumor removed by means of endoscopic surgery using a CO2 laser following earlier embolization of the left superior laryngeal artery. From the patient’s perspective, it is extremely important that the procedure be performed in a possibly least invasive manner so that no laryngeal or pharyngeal dysfunctions develop as the consequence thereof.

Due to the total nature of the resection with negative (R0) resection margins being achieved, and due to the histopathological examination being suggestive of the benign nature of the lesion, no adjuvant treatment was required in the reported case.

Due to the sporadic incidence of SFTs within the laryngeal region and the incompletely studied biology of these tumors, long-term follow-up in the setting of an outpatient laryngological clinic is advisable.

REFERENCES

Competing interests: The authors declare that they have no competing interests.

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