Introduction

Intramural esophageal tumors (IET) is group of pathological lesions arising from vessels, nerves, smooth muscles, and mucous glands located beneath unchanged mucous membrane of the esophagus. These lesions may develop into both benign and malignant tumors or developmental disorders such as cysts. The most frequent malignant tumors are gastrointestinal stromal tumor (GIST) and esophageal cancer. Benign esophageal tumors are rare and account for less than 1% of esophageal tumors [1, 2]. The classification of benign intramural esophageal tumors includes leiomyomas, schwannomas, and lipomas.

Aim

The aim of the study is to present 5 years of experience of diagnostics and treatment of IET.
Material and methods

In the years 2011–2015 eleven patients with IET were treated in our clinic: 6 males and 5 females, aged between 28 and 65 years (mean age: 52). The main symptom reported by all patients was dysphagia progressing for several months. Every patient was diagnosed by means of computed tomography of the chest and gastroscopy. Computed tomography (CT) scan revealed various sized tumors (3–10 cm) located in the esophagus. In 1 case the lesion was located in the upper esophagus, in 3 cases the middle esophagus and in the other 7 cases the lower esophagus (Fig. 1). Gastroscopy revealed unchanged mucous tissue elevated by an intramural lesion. In all cases an endoscopic ultrasound (EUS) guided fine needle biopsy was performed. One patient was diagnosed with GIST and 1 with esophageal cancer, whereas in the other cases biopsy results were nondiagnostic. In those cases positron emission tomography (PET) was conducted, but the results also did not establish a final diagnosis (Tab. I).

Results

The patient with GIST was treated with partial resection of the esophagus using the Lewis-Tanner technique. The patient with esophageal cancer was treated with transternal excision of the esophagus using the Orringer technique. In the other cases with benign lesions surgical enucleation was conducted except for 1 patient with leiomyoma. In this case esophageal resection by Lewis-Tanner technique was performed. One case of esophageal cyst was treated with radical excision. Postoperatively there were no complications. Diagnosis was established on immunohistochemical examination of the excised lesion (Tab. I). When the diagnosis of malignant tumor was established, the patient was treated with adjuvant chemotherapy.

Discussion

Introduction of new techniques of chest imaging enabled more frequent diagnosis of intramural lesions of the esophagus. Immunohistochemical examination allowed these lesions to be differentiated. Two percent of all esophageal tumors are leiomyomas. They are the most common benign tumors. They are localized mostly intramurally and can reach considerable sizes [2–4]. Over a half of all patients in this study were diagnosed with leiomyoma.

| Type of tumor | N  | Location in esophagus | Size [cm] | Diagnosis | Treatment |
|---------------|----|-----------------------|-----------|-----------|-----------|
|               |    | Upper | Middle | Lower |              |           |
| Leiomyoma     | 6  | –     | 1      | 5     | 5–10        | RTG, CT, EUS-FNA, PET | Surgical enucleation 5 Lewis-Tanner operation 1 |
| Schwannoma    | 1  | 1     | –      | –     | 3           | RTG, CT, EUS-FNA | Surgical enucleation 1 |
| GIST          | 1  | –     | 1      | –     | 4           | RTG, CT, EUS-FNA | Lewis-Tanner operation 1 |
| Cysts         | 2  | –     | 1      | 1     | 5–10        | RTG, CT, EUS-FNA | Surgical excision 2 |
| Carcinoma     | 1  | –     | –      | 1     | 8           | RTG, CT, EUS-FNA, PET | Orringer operation 1 |
| Total         | 11 | 1     | 3      | 7     |             |           |

Fig. 1. A – Gastrointestinal stromal tumor of the esophagus, B – schwannoma of the esophagus
Schwannoma and GIST have rarely been diagnosed so far due to great similarity to leiomyoma both in clinical course and imaging [2, 5]. Esophageal cysts look distinctly different in EUS and CT scans than solid tumors; therefore they are easier to diagnose [6–8]. Esophageal cysts are often located circularly in the esophagus. It is believed they originate from epithelial cells forming mucous glands located intramurally [1, 9].

The clinical course of patients with IET is distinctive. Major complaints are progressive dysphagia, hemoptysis, dyspnea, and chest pain [2, 6, 10, 11]. In our experience patients were mainly complaining of dysphagia.

Esophageal cysts occur mainly in adults, less frequently in younger patients [6, 8]. Leiomyomas and schwannomas are more common in middle aged patients. Malignant lesions are characteristic for people beyond the age of 50 [5, 9]. Gender has not been associated with frequency of such lesions [4, 5, 9, 11].

The patient is initially diagnosed by means of chest X-ray and gastroscopy. Radiograms frequently detect lumps located in the posterior mediastinum around the esophagus. Gastroscopy often shows elevation of unchanged esophageal mucosa to esophagus lumen [3, 7]. Computed tomography allows one not only to precisely localize the tumor but also to determine its structure and size [2, 3, 10]. Magnetic resonance imaging allows one to differentiate paraeosophageal from paravertebral localization of the tumor [7]. The majority of IET are located in the lower third of the esophagus [3–5]. Positron emission tomography is helpful to verify the status of lymph nodes and possible metastases [12]. In our study PET examination did not prove helpful to verify the status of lymph nodes and possible metastases [1, 4, 13].

Tab. II. Immunohistochemical differentiation of the intramural esophageal tumors

| Type of tumor | CD 117 | CD34 | SMA | Desmin | S-100 |
|---------------|--------|------|-----|--------|-------|
| Leiomyoma     | −      | + (10–15%) | +   | +      | Rare  |
| Schwannoma    | −      | +    | −   | −      | +     |
| Fibromatosis  | +/−    | + Rare | +   | + Rare | −     |
| GIST          | +      | + (60–70%) | + (30–40%) | Very rare | + (5%) |

IET depend on the location, size and histopathological type.

Conclusions

Intramural esophageal tumors are a diverse group of neoplasms, both benign and malignant. In any case of IET we should seek a histopathological diagnosis. Treatment of IET depends on the location, size and histopathological type.

References

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