Case of oesophageal gastrointestinal stromal tumour

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Gastrointestinal stromal tumours (GIST) are tumours of the mesenchymal origin most often found in the stomach (60–70% of cases), in the small intestine (20–25%), and colon (5%). The oesophagus constitutes the rarest location of GIST (2–3%), and with uncommon clinical symptoms it is possible for the tumour to grow large [1, 2]. In this letter, we would like to share our own insights in the treatment of oesophageal GIST.

A male, 63 years old, was admitted to the department due to a smooth pathological mass located in the lower thoracic part of the oesophagus. For 2 years the patient had complained of difficulties in swallowing, which had intensified in recent months. X-ray examination of the oesophagus with contrast showed the modelling of the oesophagus at a level of 33 cm from the incisors (Figure 1 A). Computed tomography scan of the chest revealed a smooth pathological mass with the dimensions of 30 × 30 mm located at the level of the lower pulmonary vein, without mediastinal lymph node enlargement (Figure 1 B). The patient underwent a biopsy with the EUS technique with the diagnosis of GIST. The patient was subjected to surgical treatment using the Ivory-Lewis technique and side-to-side anastomosis. Postoperative care was uncomplicated and the patient was discharged on day 9 after the surgery. Histopathological examination confirmed the earlier diagnosis of GIST, and an immunohistochemical examination showed positive reactions for CD 117 and CD 34, negative for S100 and actins of smooth muscles. The mitotic index was 0/50 HPF (Figure 2). The patient did not receive adjuvant treatment after surgery (0/50 HPF). Four-year follow-up did not show any recurrence.

In 1983 Mazur and Clark were the first ones to use the term GIST for mesenchymal tumours. The definition is general and defines GIST as the group of non-epithelial gastrointestinal tumours composed of spindle cells derived from interstitial cells of Cajal [3]. This tumour is usually found in men aged 40–50 years, and its main symptom is gradually increasing dysphagia, the severity of which depends on the location and size of the tumour [4, 5].

Diagnosis is based on the gastrofibroscopy, transoesophageal ultrasound, and computed tomography (CT) of the chest [5]. It is controversial to perform the biopsy of the tumour using the EUS technique. It is believed that GIST has a brittle consistency, and the biopsy can increase the risk of bleeding and spreading the cancer. At the same time, pathologists do not guarantee the final diagnosis of small tissue fragments obtained with a thin needle. The biopsy also increases the risk of leakage if the sub-mucosal resection is considered [1]. In our patient biopsy with thin-needle EUS technique was performed, obtaining the GIST diagnosis, which allowed the selection of the appropriate strategy of surgical treatment.

In the immunohistochemical examination of preparations, as much as 95% stated the activity of antibodies CD 117: in 60–70% for CD 34, 30–40% for SMA, and in 5% for S 100 [1, 6]. In negative CD117 cases, the immunohistochemical analysis should be based on the presence of the DOG-1 antibody. It allows avoidance of false negative results [6]. In differential diagnosis we should exclude the most common benign tumour of the mesenchymal origin – leiomyoma [1].

The GIST, depending on the size of the tumour and mitotic index, are divided into four groups: very low, low, intermediate, and high risk. As adverse factors include the diameter of the tumour > 9 cm and the mitotic index > 5/50 HPF [1, 4, 5]. In our patient, the diameter of the tumour was 3 cm and the mitotic index was 0/50 HPF, which is favourable in the prognosis.

Metastases are usually found in malignant forms of GIST, and their percentage is estimated at 10-30%. The cancer is spreading mostly through blood to the liver (50–60%), peritoneum (20–43%), and to lungs and bones (10%). Metastases in the lymph nodes are found

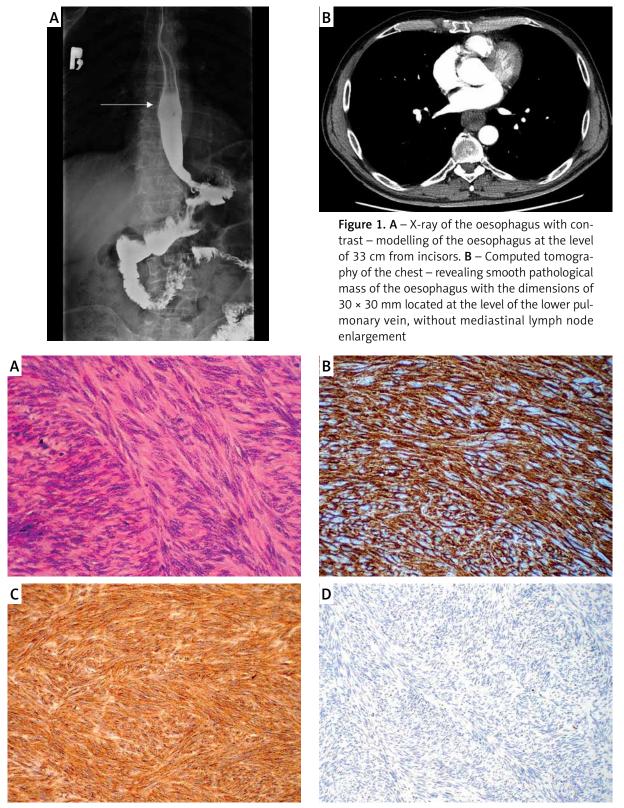


Figure 2. Histopathological examination. A – In microscopic examination monomorphic, hypercellular, spindle cells were noticed. The tumour was located in the submucosal layer and was well-bounded with fibrous capsule. The cells showed no atypia and no mitotic activity counted in 50 high-power fields. B – Immunohistochemistry: strongly positive CD34. C – Immunohistochemistry: strongly positive CD117. D – Immunohistochemistry: uniformly negative S100

in < 10% in malignant form [4]. In our patient, there were no metastases.

Surgery is the treatment of choice for GIST located in the oesophagus. The selection of the surgical technique determines the size of the tumour and its location [2, 4]. The endoscopic enucleation with sub-mucosal removal using minimally invasive techniques is recommended for small tumours (2–5 cm) [3, 4]. In the case of tumours with diameter > 9 cm there is a high risk of malignant nature of the tumour, and the treatment of choice is resection of the oesophagus. Similarly, such a technique should be used when the mucosa covering the tumour is ulcerated, not allowing for the safe conduct of the sub-mucosal enucleation [1]. In cases of damage to the mucosa in the long margin of the oesophagus with the enucleation using VATS technique, the main indication is performance of conversion to thoracotomy [3]. In cases when the tumour is located in the lower part of the oesophagus or in the cardia, the largest numbers of GIST recurrences are observed. Jiang et al. showed 80% of tumour recurrence in this location, although in all cases resection of the oesophagus was performed [1]. In the case of our patient the tumour had 3 cm diameter and was located 33 cm from the incisors. Based on the above results, we resigned from performing video-assisted thoracoscopic surgery procedure and considered the resection of the oesophagus with the cadria in the open approach.

In cases of advanced tumours, imatinib is recommended for adjuvant treatment [1, 4].

Conflict of interest

The authors declare no conflict of interest.

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