Gastric Schwannoma–Surgery or Endoscopic Treatment?

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Abstract: Schwannoma is a tumor formed of spindle cells that arise from the nerve plexus Schwann sheath cells of the gastrointestinal tract wall. It appears to be rare among the tumors of the gastrointestinal tract (GIT) and occurs mainly in the stomach, representing only 0.2% of all cancers of the stomach. It is usually a benign and slow growing tumor, and malignant transformation is very rare. For a long time it may remain clinically silent. Endoscopy of the upper gastrointestinal tract is considered to be the basic diagnostic tool. At this stage, the differential diagnosis is very difficult, based mainly on the exclusion of the presence of GIST (gastrointestinal stromal tumors), which appear more commonly in this location.

This study presents the case of a 44-year-old man with non-specific epigastric pain whose gastroscopy revealed a sub mucosal polyp of the body of stomach which was subsequently removed by endoscopic polypectomy. Based on histopathological and immunohistochemical findings, an Schwannoma Antoni type A was diagnosed.

Keywords: Gastric schwannoma, endoscopy, polypectomy.

1. INTRODUCTION

Schwannoma (neurinnoma) is a rare mesenchymal tumor of the gastrointestinal tract (ca.2-7% of cases), this category including gastrointestinal stromal tumors (GIST), fibroids (leiomyoma) and leiomyosarcoma. Among these neoplasms, GISTs are the most common and 60-70% of them are located in the stomach [1]. Schwannomas account for only 0.2% of all benign, and 4% of all malignant tumors of the stomach [2]. Considering all gastrointestinal neoplasms derived from Schwann cells of peripheral nerves, schwannoma is the most common tumor [1]. It occurs predominantly in middle-aged (40-50-year-old) women [3]. It is usually a benign and slow growing tumor that rarely undergoes a malignant transformation. After complete surgical resection, further prognosis is excellent [4]. For a long time it may remain clinically silent or manifest itself in the form of non-specific pain or discomfort in the abdomen. Due to an indolent growth pattern, these tumors are often discovered incidentally. Diagnostic methods comprise endoscopy, EUS (endoscopic ultrasound), fine needle biopsy or computed tomography (CT). However, the final diagnosis must be based on biopsy tissue histology with further confirmation by immunohistochemical markers, where the tumor cells show a positive expression of protein S-100 and vimentin, and negative expression of smooth muscle actin (SMA) and c-KIT [5].

2. CASE REPORT

We present a 44-year-old man with a history of left testicle seminoma and status post orchiectomy who was admitted to the Department of Gastroenterology, PUMS, because of intermittent epigastric pain. Physical examination revealed tenderness in the upper-abdomen without a palpable mass or pathological resistance. No weight loss, loss of appetite, or weak-ness were presented. Routine blood investigations and a CEA test were unremarkable.

An abdominal CT scan showed a well-defined homogeneous submucosal lesion arising from the body of the stomach. The surrounding lymph nodes were unremarkable. There was no evidence of any other abnormalities.

Gastroscopy confirmed the presence of a polyp along the lesser curvature in the body of the stomach with a diameter of approx 2cm, covered with macroscopically normal gastric mucosa. In addition, there were numerous small (3-4mm) mucosal elevations, of which two were sampled for a histopathological examination. The antral mucosa was also biopsied as it was slightly congested.

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Microscopically, fundic gland endometrial polyps were found in the gastric mucosa (WHO: Fundic gland Polyps). Histopathological examination of fragments of the antrum showed no significant changes. The presence of Helicobacter pylori was ruled out in both specimens.

The patient was referred for re-gastroscopy in the operating theater with the aim of performing an endoscopic polypectomy. The lesion, which measured 2.5cm, was removed entirely with a diathermic loop and submitted for a histopathological and immuno-histochemistry examination.

The histopathological examination of the removed lesion revealed the presence of spindle cells forming palisades, which indicated a Neurinoma, Antoni type A (according to WHO: Schwannoma, Antoni type A). The diagnosis was confirmed by the result of hematoxylin and eosin (H and E) staining. The margins of the endoscopically removed tumor were negative and the patient did not require additional surgery after the procedure.

The patient is still followed-up, with two follow-up endoscopies and imaging studies (computed tomography and magnetic resonance imaging of the abdomen and pelvic) showing no deviations.

3. DISCUSSION

Digestive schwannomas were defined by Daimaru *et al*. in 1988. This tumor grows slowly and is non-symptomatic on clinical examination or has uncharacteristic symptomatology, producing non-specific abdominal pain, as in our patient. Much less often, it can be felt as a palpable mass in the upper abdomen. If symptomatic, the most common presenting symptom is upper GI bleeding, because of ulcers formed in the mucosa overlying the tumor. They develop secondary to ischemia in the mucous membranes of the tumor, which are formed due to a reduced tolerance to the gastric acidity or as a result of pick-up of blood by growing tumor cells [6]. If the change is located in the esophagus or rectum, dysphagia or constipation are also possible [7]. At the stage of physical examination, setting the initial diagnosis is practically impossible due to the non-specific clinical symptomatology.

Preoperatively, for a gastric submucosal lesion, the main differential diagnosis is very important, but difficult at this stage, and is primarily based on distinguishing schwannoma from GIST (the most common tumors in this location) due to their similar morphology. However, at the stage of early diagnosis, most frequently the exclusion of GIST is clinically impossible. Schwannoma comprise enough small percentage of mesenchymal tumors of the stomach, that in everyday clinical practice, the diagnosis is not routinely considered.

Available imaging studies such as abdominal CT, gastrointestinal endoscopy or EUS, provide information on the nature of the lesion and its morphology. The final diagnosis is always based on histopathological and immunohistochemistry examination, because there is no pattern of presentation pathognomonic for schwannoma.

In 2005, Levy *et al*. found that in CT studies gastric schwannomas are uniquely different in that they present as homogeneously attenuating, well-defined, mural, hypodense mass that, unlike GIST, shows no evidence of degeneration, necrosis, polycystic lesions, calcifications or hemorrhage, which appear as heterogeneous enhancement [6, 8]. In our patient, a CT examination showed the presence of a submucosal lesion within the body of the stomach that was well-defined and homogenous. Endoscopic examination helps to determine the position of the tumor. When the lesion is submucosal, a needle biopsy can also be performed to obtain cellular material for histopathological examination. Gastroscopy, which was conducted in our patient, confirmed the presence of a small, submucosal polyp macroscopically covered with normal mucosa. Due to the easily accessible location, it was possible to conduct biopsy for histopathological examination. Additional information about the exact position of the tumor is provided by MRI studies. The overall MRI signal pattern of schwannoma is low on T1-weighted images and markedly elevated on T2-weighted images [9]. For the diagnosis of small lesions, the best method is EUS [10], and when the tumor reaches a larger size, a good alternative is abdominal ultrasonography [11].

Macroscopically, a schwannoma has a circular or elliptical shape and an average diameter of approx. 5cm. In some cases, yellowish nodules on the surface and bleeding may be present. The incisional surface is greyish to sallow [12].

Microscopically, based on the arrangement of schwannoma's spindle cells, two types are recognised,

namely Antoni A (AA) and Antoni B (AB). In type AA, spindle cells are densely packed, forming palisades. In addition, they form specific Verocay bodies (palisade structures) and formations resembling Meissner bodies. In contrast, type AB has a significantly looser histology and the cellular composition can be heterogeneous. In specific cases, the tumor cells display a certain degree of pleomorphism and a small number of mitotic figures (<4/10 HPF), but do not exhibit pathological mitotic figures or coagulation necrosis. Outside the tumor mass is surrounded by a thick fibrous capsule with single lymphocytes and foam cells; also single blood vessels are visible. Of high diagnostic value is the immunohistochemistry examination, which enables precise differential diagnosis. The nucleus and cytoplasm of tumor cells show a positive reaction to the presence of S100 protein, GFAP and vimentin, and negative to the presence of CD117, dog1, SMA and CD34 [12]. In our patient, histological examination of the removed lesions made by H&E staining revealed a characteristic pattern of spindle cells forming palisades, which clearly indicated a Neurinoma, Antoni type A (according to WHO: Schwannoma, Antoni type A). Thus further immunohistochemical studies were unnecessary.

Due to difficulty in diagnosing gastric schwannoma preoperatively and its insensitivity to chemotherapy and radiotherapy, the treatment of choice is complete surgical resection of the lesion. Gastric schwannomas rarely exhibit lymph node metastases, so it is believed that regional lymph node dissection is not required [12]. If the tumor has been completely removed along with a margin of tissue, the prognosis is excellent with a very low recurrence rate. The selection of the most appropriate treatment should take into account the location of the tumor, its size and relative position of the surrounding organs. To remove small, benign-looking tumors growing in the gastric lumen, laparoscopic or endoscopic resection is the best. Wedge resection, total or subtotal gastrectomy is performed if the lesion grows outside the gastric lumen or is malignant [7]. In our patient, the tumor of 2.5cm in size and was removed by endoscopic polypectomy, due to its small size and unremarkable histopathological examination of the material sampled during diagnostic gastroscopy. The final diagnosis was established postoperatively by histopathological examination.

So far, few reports have been published in the literature about schwannoma of the stomach. Because of the rare occurrence of schwannoma and the inability to collect a sufficiently large clinical group, in the literature, there are only a few large studies, single case reports mainly.

This study is an indication for physicians and clinicians to highlight the fact that in everyday practice and treatment of apparently benign-looking polyps of the stomach, it is important to consider also a rare mesenchymal tumors, including schwannoma as well.

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