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SCIENTIFIC ARTICLE - CASE REPORT

ESOPHAGEAL SCHWANNOMA: A CASE SERIES

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ABSTRACT

INTRODUCTION: Schwannoma is a neoplasm, usually benign, originating from Schwann cells, responsible for the formation of the myelin sheath in peripheral nerves. It is most frequently found in the peripheral and central nervous systems; however, it can rarely arise in the gastrointestinal tract. It presents an intramural location. Objective: To report a series of cases of Esophageal Schwannoma (ES).

METHOD: Review of the results of 65,484 upper digestive endoscopy (UDE) exams, performed at a Private Clinic in Goiânia-GO, and their respective biopsies. Results: Of these exams, 12 (0.018%) cases of ES were diagnosed, with a mean age of 43.4 ± 5.2 years, ranging from 17 to 82 years. Eight (66.6%) were female. The most common site of involvement was the distal esophagus (76.9%).

CONCLUSION: ES is a rare tumor in the digestive tract. In the present study, it was found in 0.018% of UDE exams. We observed a higher prevalence in females (66.6%). The distal esophagus was the most affected segment (76.9%).

Keywords: schwannoma; Esophageal Schwannoma; mesenchymal tumors of the gastrointestinal tract.

INTRODUCTION

Mesenchymal neoplasms involving the gastrointestinal tract (GIT) are typically intramural, and they are divided into two main groups: gastrointestinal stromal tumors (GISTs) and tumors similar to those occurring in soft tissues.[1, 2,3,6].

The subgroup of GISTs encompasses the most common mesenchymal tumors of the GIT [2]. These neoplasms are most frequently located in the stomach and proximal small intestine but can occur in any part of the digestive tract and occasionally in the omentum, mesentery, and peritoneum[2]. They are primarily identified by the expression of the KIT protein and often exhibit activating mutations in the KIT or platelet-derived growth factor receptor alpha (PDGFRA) genes [2,5].

The other, rarer subgroup is composed of tumors identical to those that can arise in soft tissues throughout the rest of the body [2]. This group of neoplasms includes lipomas, liposarcomas, leiomyomas, true leiomyosarcomas, desmoid tumors, schwannomas, and tumors of the peripheral nerve sheath [2].

Schwannomas, on the other hand, are solitary neurogenic tumors, with the majority being benign 6, arising from cells of the neural myelin sheath [1]. They are most commonly found in the peripheral and central nervous systems; however, they can rarely arise in the gastrointestinal tract [1,2,4,8,10,11]. The stom-

ach is the most common site for schwannomas, but they can also be rarely found in the retroperitoneum, esophagus, colon, and rectum[4,5].

Intra-abdominal schwannomas occur equally in men and women, without a peak incidence by age group[4]. Diagnosis typically occurs after excision of the lesion with histopathological study[8,9]. Definitive treatment involves complete resection of the lesion with negative margins[8].

Given the scarcity of data on the prevalence of Schwannomas involving GIT, we conducted a study to evaluate the prevalence of esophageal schwannoma in a private Gastroenterology and Endoscopy clinic in Goiânia-GO.

METHOD

This is a retrospective prevalence study, where we reviewed the results of upper gastrointestinal endoscopy (UGIE) exams and their respective biopsies performed at a private Gastroenterology and Endoscopy clinic in Goiânia-GO.

During the study period, 65,484 UGIE exams were performed. We reviewed the medical records of these patients and evaluated age, gender, endoscopic findings, and histopathological findings. We included in the study all patients who underwent UGIE during the period and excluded exams performed on the same patient and those where the presence of Schwannoma was not confirmed by histopathology.

RESULTS

Out of the 65,484 exams performed, we diagnosed 17 cases (0.03%) with endoscopic suspicion of esophageal Schwannoma (ES). Of these, three cases were repeated exams in the same patient, and in two cases, the histopathological exam was compatible with leiomyoma. The remaining 12 patients (0.02%) had a histopathological diagnosis of Schwannoma and composed the study population.

Of these, eight patients (66.6%) were female. The mean age of the group was 43.4 ± 5.2 years, ranging from 17 to 82 years. In 11 patients (91.6%), the lesions were solitary, and in one patient (8.4%), two lesions were identified. The size of the lesions ranged from 2 to 20 mm. Regarding the site of the lesions, the most common site of involvement was the distal esophagus, where ten lesions were found (76.9%), with two lesions (15.4%) in the middle esophagus and one (7.7%) in the proximal esophagus. In all cases, endoscopic treatment was possible without complications.

DISCUSSION

Benign primary tumors of the esophagus account for 2% of cases; of these, 80% are leiomyomas and only 1% are Schwannomas [2]. Schwannomas are tumors, in most cases, with benign behavior [4,6], derived from Schwann cells, responsible for the formation of the myelin sheath of nerves [4]. The most common location is in the central and peripheral nervous system [4], being rarely found in the gastrointestinal tract, where the most common site is the stomach [4,5]. They can also rarely be seen in the esophagus, colon, rectum, and retroperitoneum [1,2,3,4,5].

The ES was first described by Chatelin and Fissore in 1967 [7,8]. It is a rare type of benign mesenchymal tumor of the esophagus, with a higher incidence in Asia [8]. The average age of incidence is around 50-60 years[5]; however, in this study, the mean age was slightly lower (43.4 ± 5.2 years). The ES appears to have an equal distribution in both sexes4; however, in the present study, it was twice as prevalent in females.

Patients with ES can be asymptomatic or present with a wide variety of symptoms, including ab-

dominal pain, intestinal constipation, nausea, and vomiting [3,5]. Some patients may experience dyspnea due to compression of the trachea by the tumor [9,10]. However, the most common clinical manifestation is dysphagia [3,10].

Diagnosis is typically suspected through EGD 3,10, with confirmation achieved through histopathological study of the lesion and immunohistochemistry [1,3,4,7,8,10]. Biopsy findings commonly include cellular pleomorphism, presence of lymphoid follicles, rare mitotic figures, and occasional areas of necrosis [4,8]. Tumor cells are immunohistochemically positive for S100 [1,3,4,6], a characteristic marker of Schwann cells[8,9], and negative for smooth muscle markers such as SMA, CD34, and CD117 [3,4,10].

The treatment strategy for gastrointestinal Schwannomas is based on size, location, and association with surrounding tissues; available treatment modalities include endoscopic and surgical resection.[11] Surgical resection remains the standard and most effective treatment, involving complete tumor removal with a margin of safety.[4,5,8,10] Endoscopic resection can serve as an alternative method for selected patients and may be attempted in gastrointestinal schwannomas with a diameter <3 cm and no signs of malignancy.[11] Radiation therapy and chemotherapy seem to have no benefit in treating this condition.[3,10] After complete resection, the prognosis is usually favorable, as tumor recurrence is rare [4,8].

CONCLUSSION

ES is a rare mesenchymal tumor of the upper digestive tract. In the present study, we observed a prevalence of 0.018%. Although there is no distinction between sexes in the literature, there was a higher prevalence in females in our population (66.6%). The mean age of our population was slightly lower than reported. The distal esophagus was the segment most affected (76.9%). All cases were treated endoscopically. Larger studies are needed to outline the epidemiological profile of this rare tumor.

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