








CASUISTIC PAPER

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Neuroendocrine tumor of appendix located Spiegel hernia – case report and review of the literature

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ABSTRACT

Introduction. Appendix, located within the Spiegel hernia is a rare condition. Few cases have been reported to date. Although it is generally asymptomatic, patients can apply with strangulation findings. Along with the physical examination findings, imaging methods play an important role in diagnosis and definitive diagnosis is made intraoperatively. Per-operative surgical method is determined according to the condition of the structures in the hernia sac. If an appendix is detected in the hernia sac, appendectomy is often preferred regardless of symptoms. Postoperative pathology is mostly benign but malign appendix pathologies should be kept in mind.

Aim. Here, we aimed to present our case undergoing emergency surgery due to incarcerated hernia as it is the first case of appendix neuroendocrine tumor in the Spiegel hernia sac according to our literature review.

Description of the case. A 77-year-old female patient who was admitted to the hospital with complaints of nausea and vomiting was evaluated as an emergency. In the clinical evaluation of the patient, we detected ileus due to hernia. We operated on the patient and found the appendix and cecum in the spiegel hernia. We did appendectomy and hernia repair. Histopathological examination of the appendix revealed a well-differentiated neuroendocrine tumor.

Conclusion. Detection of the appendix in a Spiegel hernia is a rare condition. This is the first case of appendiceal malignancy in a Spiegelian hernia.

Keywords. appendix, carcinoid tumor, hernia, neuroendocrine tumor, Spiegel

Introduction

Spiegel Hernias (SH) are occur on the lateral border of the rectus abdominus muscle and weak fascia on the linea semilunaris. Originally introduced by Josef Klinsch in 1764, this hernia consists of Adrian van den Spieghel whose name defines linea semilunari. It oc-

curs congenitally or acquired and accounts for 1-2% of all hernias.^{1,2}

In the literature, we detected only 22 cases of appendix located the hernia sac of patients operated for SH. As a method, the English language literature based on the MEDLINE (PubMed) database was reviewed. The

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keywords spiegel, hernia and appendicitis were used. None of these cases reported malignancy as a result of histopathological evaluation.

Aim

In this article, we presented the patient, who was operated for SH because of appendix within the hernia sac, in the light of the literature. This case, as far as we know, is the first case in the literature in which a NET was detected in the appendix located in SH.

Description of the case

The patient is an obese women 77 years old. She was admitted to the emergency room with a complaint of abdominal pain, nausea and vomiting. In her past medical history, she had abdominal pain at intervals for the last 1 year. She had a previous history of cholecystectomy and hysterectomy operation. She had no chronic diseases other than hypertension. Body mass index (BMI) was calculated as 36,1. On physical examination, there was minimal distention in the abdomen. There was sensitivity and defense, especially in the lower right quadrant. No mass was detected during palpation. She had gas and out-flow. Intestinal sounds were detected normoactive. Gastric fluid came from the nasogastric tube which was about 100 cc. In laboratory tests, WBC: 16.100×10^3 U/L, CRP: 0.4 were detected. Computed tomography (CT) examination showed that the cecum and appendix were herniated from the 3 cm defect in the lower right side of the rectus muscle in the lower right quadrat (Fig. 1A, B).

Proximal loops were dilated due to compression of the cecum in the hernia sac. Upon this, the patient was operated with a pre-diagnosis of strangulated hernia. The hernia sac was reached through an oblique incision made from the right iliac region. The appendix was found to be adherent in the pouch (Fig. 1C). Appendectomy was performed and the hernia sac was excised. Hernia repair was performed with prolene mesh

placed in the preperitoneal area. The patient was discharged from the hospital on the second postoperative day without any problem.

Histologically the tumor was located at the tip of the appendix invading the muscularis propria with a diameter of 5 mm (Fig. 2A, B).

It was composed of solid islands and small groups of round monotonous cells. As there were no mitosis and Ki-67 proliferation index was lower than %1 the tumor was evaluated as well differentiated grade 1 neuroendocrine tumor of the appendix. The tumour cells were diffusely positive for chromogranin and synaptophysin immunohistochemically (Fig. 2C). Mesoappendix and surgical margin were free of tumour.

No recurrence or metastasis was detected during the 2-year follow-up of the patient.

Discussion

SH is a lateral ventral hernia or also called semilunar line hernia . SH arises from the weak area in the Spielian fascia between the rectus muscle and the internal and transverse oblique muscles. Increased intra-abdominal pressure, abdominal wall trauma, and weakening of the fascial layers with age are the reasons included in the etiology.^{1,3} Obesity, chronic obstructive pulmonary disease, diabetes, coronary artery disease and peripheral vascular diseases are the most common comorbidities. In addition, 50% of these patients have a history of abdominal surgery.⁴ Omentum (39.1%), small intestine (33.7%) and colon (13.5%) are frequently located in the hernial sac. Rarely, the stomach, gallbladder, ovary and appendix can be located in the hernia.⁴ Clinical findings may differ according to the structures in the hernia. Patients can usually apply only with pain without palpable swelling, and this makes diagnosis difficult.⁵ Our patient also presented with a non-specific abdominal pain without swelling as stated in the literature. Ultrasound (US) and CT help in diagnosis. In the series of 81 cases by Larson et al., 74%

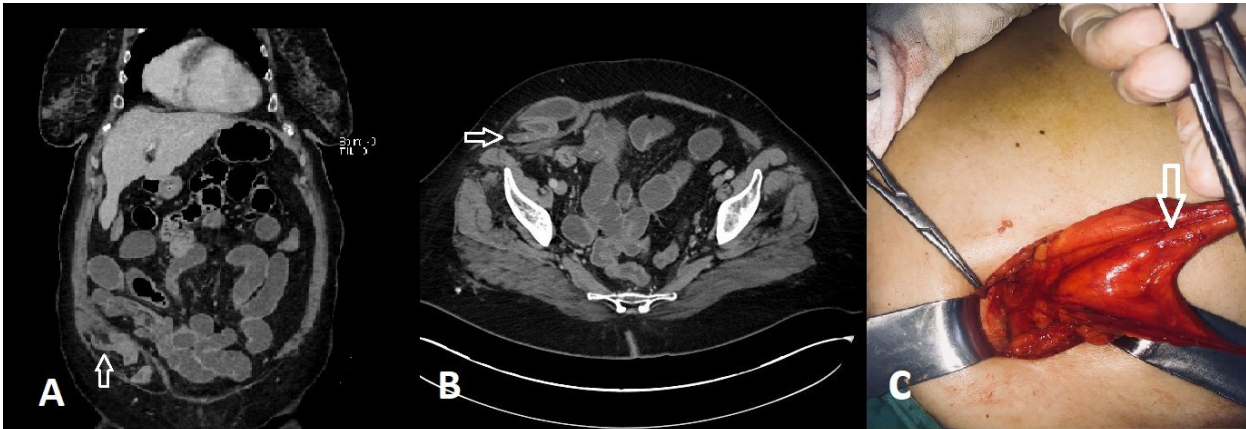


Fig. 1. The coronal CT (A) and axial CT (B). The arrows indicate the insert within the SH sac. Peroperative appendix and mesoapandix (C). The marked arrow shows the appendix inside the opened hernia sac

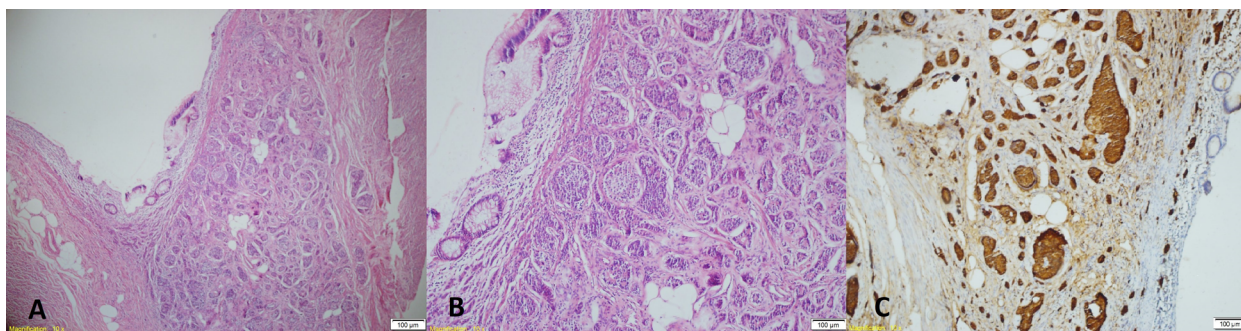


Fig. 2. Histopathologically solid islands and small round uniform cell groups (A: H&E×4, B: H&E×10), Commonly positive tumor cells for chromogranin and synaptophysin (C: Chromogranin×10)

of the patients were diagnosed only by physical examination, while the rest was diagnosed by imaging methods. In this study, incarceration rates in SH were between 17–24%, and 10% of patients were taken to emergency operation.³ In our patient, diagnosis was made with CT an emergency operation was planned.

Amyand's hernia is the placement of the appendix in the inguinal hernia and named by Claudius Amyand in 1735. De Garengeot hernia is the placement of the appendix in the femoral hernia and it was defined by Rene Jacques Croissant de Garengeot in 1731. The incidence of both is less than 1% of inguinal and femoral hernias.^{1,6} The presence of appendix in SH is much more rare also and has not yet been specifically named in the literature. There are only 22 cases.

The purpose of the surgical operation is appendectomy and hernia repair. Appendectomy according to the findings of intraabdominal sepsis and ischemia; it can be done by transabdominal or herniotomy. Open and laparoscopic techniques using primary suture or mesh are applied for SH repair. A number of authors state that the open approach is appropriate in the case of incarceration.² In the absence of appendix inflammation in SH, the use of mesh in prophylactic appendectomy and hernia repair remains controversial. There are not enough studies on this subject yet. In the prospective multicenter study of Malazgirt et al., it has been revealed that in open or laparoscopic methods, preperitoneal repair reduces the length of hospital stay and provides better patient compliance.⁷ Since our patient had strangulation findings, laparoscopic method was not preferred.

NET which is the most common malignancy of the appendix, is mostly asymptomatic and is detected incidentally after appendectomy. The rate of NET detection in patients undergoing appendectomy is 0.3–0.9%.⁸ The vast majority are less than 1 cm and appendectomy is sufficient. In the pathological examination of our case, the treatment was completed in accordance with the 2017 NCCN recommendations, due to the detection of a well-differentiated NET with a diameter of 0.5 cm at the tip of the appendix.⁹

Conclusion

Detection of appendix within the SH sac has not been named before. Our patient is the first case with appendix malignancy in SH. The possibility of malignancy should be kept in mind in these patients. Appendectomy should be performed routinely in case of appendix detected in SH pouch. Primarily laparoscopy should be preferred according to strangulation status. It would be appropriate to remove the appendix using an endobag.

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