Case report

Incidental diagnosis of a low-grade mucinous appendicular neoplasm: A case report

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ABSTRACT

Introduction and importance: Low-Grade Appendiceal Mucinous Neoplasms (LAMN) are noninvasive epithelial tumors of the appendix. Symptomatology is unspecific and can simulate appendicitis. The multidisciplinary approach for the correct diagnosis and management of this kind of tumor is important, as the prognosis depends on it.

Presentation of the case: A 70-years-old man with an appendiceal picture is presented. McBurney and Blumberg’s signs were found and the Echography has reported an acute appendicitis. He underwent laparoscopic surgery, founding a Tumor in the Cecum of 10 × 5 cm. We decided to perform an appendectomy with a total cecectomy. The pathological diagnosis was a LAMN. Ten months later, a Computed Tomography (CT) Scan control was performed reporting no evidence of Tumoral activity or Distance Metastasis, neither pseudomyxoma peritonei (PMP) evidence.

Discussion: LAMN has been a very controversial tumor. The definitive management is appendectomy. There are controversies about the laparoscopic and open approach, and when there are positive margins. The stage was a pTis (LAMN) pNx according to the 8th edition of the American Joint Committee on Cancer (AJCC). The prognosis depends on the tumoral stage and the presence of PMP. In our case, we have performed a CT scan follow up and there was no Tumoral activity or Distance Metastasis, neither PMP evidence.

Conclusion: LAMN is a very interesting and rare tumor. The diagnosis of this case was a real challenge. Our management was simple due to the tumoral stage. Pathology played a very important role in treatment and prognosis.

1. Introduction

LAMNs are noninvasive epithelial tumors of the appendix [1]. The classification of the World Health Organization (WHO) (2010) and the 8th edition of the AJCC Cancer Staging Manual, are the most used and it has not been well established a definitive agreement between them [2–4]. Symptomatology is unspecific, it can be diagnosed incidentally after an appendectomy (23%) [5]. The diagnosis can be made by a CT scan [3]. In pathology, the finding is acellular mucin inside the wall and pushing invasion [2]. Tumor markers levels utility is still unclear [2]. The management is an appendectomy with or without right hemicolectomy [5]. The prognosis depends on the tumor stage, symptoms, perforation, the surgical approach, and the evidence of tumoral cells on surgical margins [2,6]. The most feared complication is PMP [6].

We present a 70-years-old man with an incidental trans-surgical diagnosis of an Appendiceal Mucinous Tumor and his evolution. This case was reported in line with the SCARE criteria [7].

2. Presentation of the case

A 70-years-old man has come to the Emergency Room (ER) on December 23th 2019, with a history of 24 h of -constantly and intensity progressive- abdominal pain, that initiated unspecific and soft [5/10 of Visual Analog Scale] on the Mesogastric Abdominal Region and after 12 h since the beginning, the pain was localized on the Abdominal Right Lower Quadrant (RLQ). It was associated with nausea and hyporexia and increased intensity with physical activity. No diarrhea, fever, or constipation was associated and no analgesic had been taken. There was no relevant surgical, family, drug, allergic, medical or psychosocial history.

On physical examination, we found normal vital signs [Heart Rate: 82, Respiratory Rate: 18, Temperature: 37.7°C, BP: 125/80 mmHg, Oxygen Saturation: 98%, Body Mass Index: 36%], a moderate abdominal
adipose panicle, with low peristalsis, muscular resistance to palpation in RLQ and a positive McBurney and Blumberg signs, with no tumor palpation. The blood test had the presence of Neutrophilia (69.1%) and an increased C-Reactive Protein (CRP) (46.02 mg/L). Abdominal Echography found an acute appendicitis with complex, voluminous, fluid-containing peri-appendicular collection, extending from the Cecum to Ileocecal valve. Due to the clinical features, the Laboratory and Echography findings, we diagnosed the case as an Acute Appendicitis and we decided to take the patient to the operating room.

A Laparoscopic Appendectomy was performed by an Advanced Laparoscopic Surgeon of the Vivian Pellas Hospital; pneumoperitoneum was made (12 mmHg of CO$_2$) with the Veress Needle Technique in the umbilicus through a 10 mm incision. The surgical findings were a Tumor in the Cecum of $10 \times 5$ cm, proximal to the Ileocecal-valve; cystic in appearance, soft, with lateral parietal peritoneum and mesentery adhesions and omentum in the posterior wall (Fig. N1). We decided to perform a tumor resection in the proximal cecum, 2 cm distal to the tumor and below the ileocecal-valve, with 60 and 45 mm blue ENDOGIA mechanical suture. The piece was removed in an Endo Bag enlarging the wound of the hypogastrium trocar to 3 cm for his extraction, the complete tumor was successfully extracted without spilling liquid into the peritoneal cavity or subcutaneous tissue (Fig. N2). The liquid contained in the tumor was clear and mucinous. The patient adhered and tolerated the provided intervention. He also, was discharged with no exceptional medication requirements either complications the next day.

The pathological diagnosis (January 2nd, 2020) was a LAMN with a pTis (LAMN) pNx stage, using the pTNM, Eighth Edition of the AJCC, with free margins of tumoral disease. On October 14th, 2020, a CT scan control was performed, reporting no evidence of Tumoral activity or distance metastasis, neither PMP evidence. The patient has accomplished with all follow up appointments and expressed satisfaction and happiness about the management and prognosis.

3. Discussion

The LAMN’s has been a very controversial tumor in the literature due to the different clinical and histopathological features [3,5]. In this kind of neoplasms, the average age of diagnosis is above 50-years-old, predominantly in females [1,2,5,8–10], the clinical presentation usually can confuse the diagnosis process due to its non-specific symptomatology [5,9]. The most frequent symptoms are RLQ abdominal pain, a palpable mass, weight loss, change in bowel habits [2,8] and an inguinal hernia can be present in male patients [8]. In fact, in many cases (23%), it can be diagnosed incidentally after an appendectomy, as in our case. On the other hand, it can be asymptomatic and diagnosed incidentally by imaging studies [2]. In this sense, a preoperative abdominal echography was performed that confirmed the initial diagnosis, and no tumor was seen. Yang, JM. et al. presented a case report of a 63-years-old postmenopausal woman with LRQ pain and mass, the echography reported “a huge cystic mass of 7 cm $\times$ 13 cm in the right ilceccal-appendix region, which had a blurred border with the surrounding tissue” [9]. The abdominal echography usually reports “a mass with small echo spots and/or a concentric echo layer (known as the ‘onion skin’)” [9], however, it depends on the number of layers [11] and only less than 50% of the cases can be identified [3].

During the diagnosis process of LAMNs, the utility of laboratory tests is still a controversial issue. Elevated tumor markers CEA, CA 19-9 and 125 can be associated with mucinous cystadenoma and cystadenocarcinoma, however, they are not specific [2,5,8,9,11]. Gündogar et al. (2018) made a five years (2011–2016) small case series and literature review, they have concluded that tumor markers “were not
significant in terms of follow-up, treatment, prognosis, and recurrence” [2]. In our case, there was no justification for the use of tumoral markers.

The definitive management of LAMN is the surgical approach nonetheless, there still are discrepancies in the extent of surgical resection and the laparoscopic vs laparotomy procedure [3]. Some reports affirm that the laparoscopic approach is a safe procedure and provides a better evaluation of the abdominal cavity, although, there is the risk of rupture of the neoplasm that can complicate the patient with an eventual PMP [2,3,11], for this reasons some authors advise -according to the surgeon experience- not lose sight of the possibility to convert and perform a laparotomy, especially, if there is a larger lesion or with more adhesions that could increase the risk of tumor rupture and injury [9].

The surgical margins were negative and we considered not to perform a right hemicolectomy. In the LAMN cases where the tumor is localized, many publications are agreed that simple appendectomy is sufficient to be curative [2,5,9–11], but there are still no guidelines about the appropriate management when the surgical specimen has positive surgical margins [2,3,12]. During our review we found a case report of Givalos, N. et al. affirming that there are reports of experts suggesting a cecectomy in mucinous neoplasms with positive margins with negative lymph nodes [12].

The definitive diagnosis was LAMN with a pTis (LAMN) pNx stage according to the 8th edition of the AJCC. As we mentioned above, the classification of the Appendiceal Mucinous Neoplasms (AMNs) has been changed many times, in fact, today, there is not a definitive consensus [2,6,9]. This tumor “constitutes a very broad diagnostic spectrum ranging from adenoma to mucinous adenocarcinoma” for this reason its classification has been so difficult [2]. The most used and actualized classification is the one proposed by the WHO, in 2010, in the 4th edition of the World Health Organization Digestive System Tumor Classification [9], where it classified the AMNs in Adenoma, Low-grade appendiceal neoplasm, and Mucinous adenocarcinoma [2,6]. On the other hand, in 2017, the AJCC in its 8th edition, made changes in the staging of the LAMN creating the Tis stage that consists of a tumor that invade the lamina propria, with or without muscularis mucosa, but not through the muscularis propria[2], occasionally with a “pushing” pattern as a diverticulum-like growth into the muscularis [11]. In this classification there is no T1 and T2 stages, this is due to a replacement of the muscularis propria by fibrosis[11], and the involvement of the subserosa is T3 stage and T4 category is when the tumor or the acellular mucin involves the surface (T4a) or invade other organs (T4b)[2].

The prognosis of AMNs will depend on the tumoral stage and the presence or not of PMP. In most of the cases, in pTis-LAMN, the patient is cured by the resection of the appendix, with no risk of recurrence [4,11,13], however, it has been reported that 2% can evolve to PMP [11]. In our case, we have performed a CT scan follow up and there was no Tumoral activity or Distance Metastasis, neither PMP evidence.

4. Conclusion

The LAMN is a very interesting and rare tumor. In our case, the management was simple due to the tumoral stage and the absence of peritoneal mucin, however, it was a very challenging diagnosis. In this sense, the pathology report played a very important role to determine the definitive treatment and prognosis. The approach has to be multi-disciplinary and cooperative between the surgery, radiology, and pathology departments.

In surgical management, we have to be sure if the laparoscopic procedure is a feasible approach according to the tumoral characteristics and the surgeon experience, as well as we have to give a very well detailed data to the pathologist. Even if the pathology department reports an initial stage (pTis-LAMN) without positive margins, a radiological follow-up must be well done to discard recurrences or the presence of PMP.

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Ethical approval

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Consent

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Author contribution

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The authors Gabriel A. Guzmán, Isaías Montealegre and Alejandro M. Obando declare no conflict of interest.

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