Neuroendocrine Tumor of the Appendix
(A Case Report)

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ABSTRACT

Neuroendocrine tumours (NETs) are characterised by their ability to secrete hormones. There is a wide variety of NETs that can develop in any organ of the digestive system. NETs of the appendix are rare with an estimated incidence of 0.15 to 0.6 cases/year per 100,000 people and are often seen in young adults with a male predominance. Most NETs of the appendix are asymptomatic and usually discovered incidentally during appendectomy. Treatment is by surgical removal of the tumour to a healthy margin in accordance with the principles of carcinoid tumour.

We report on a case of a 24-year-old female patient. She was admitted with generalized abdominal pain originating in the right iliac fossa with a B.P. of 0 and a fever of 38°C. The abdominal and pelvic CT scan showed acute appendicitis with a discrete agglutination of the loop around the appendix and an effusion of the right iliac fossa and pelvis of medium abundance. The biological work-up showed a hyperleukocytosis with a predominance of neutrophils. The patient underwent ileo-caecal resection, ileo-colostomy in double gun barrel after an exploration which revealed a mass of appendicular cystic appearance "suspicion of appendicular mucocele?" with a purulent peritoneal effusion and false membranes in the inter-anses. Pathological examination showed a proliferation of organoid architecture suggestive of a neuroendocrine tumour measuring 4 mm in length.

Keywords: Appendix, carcinoid tumour, neuroendocrine tumour, right ileo-haemicolecotony.

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I. INTRODUCTION

Neuroendocrine tumors (NETs) are mainly caused by enterochromaffin cells present in the gastrointestinal tract and the bronchopulmonary system. Neuroendocrine tumors of the appendix are the third most common gastrointestinal neuroendocrine tumor after the small intestine and the rectum. Appendiceal neoplasms are rare. They are mostly asymptomatic. Tumors of the appendix are mostly incidentally identified in patients operated on for acute appendicitis. They are detected in about 1% of appendectomy specimens on histopathological examination [1]. Current guidelines suggest that a simple appendectomy is an adequate treatment for NETs if they are less than 1 cm in size, and a right hemicolectomy is recommended for those larger than 2 cm. However, the ideal treatment for tumors between 1 and 2 cm is still controversial [2].

We report on a case of a 24-year-old female patient. She was admitted with generalized abdominal pain originating from the right iliac fossa with a P.S. of 0 and a febricula of 38 °C. Abdominal examination revealed generalized abdominal tenderness. In the paraclinical work-up, the abdomino-pelvic scanner showed acute appendicitis, a stercolith with discrete agglutination of the loop around the appendix and an effusion of the right iliac fossa and pelvis of moderate abundance. The biological work-up showed a hyperleukocytosis with a predominance of neutrophils. The patient underwent ileo-caecal resection, ileo-colostomy in double gun barrel after an exploration which revealed a mass of appendicular cystic appearance "suspicion of appendicular mucocele?" with a purulent peritoneal effusion and false membranes in the interrines. Pathological examination showed a proliferation of organoid architecture suggestive of a neuroendocrine tumor measuring 4 mm in length.

II. MEDICAL OBSERVATION

The patient was 24 years old, with no particular pathological history, and was admitted to the emergency room for generalized abdominal pain originating in the right iliac fossa with vomiting and no other associated signs.

On general examination, the Status of Performance was zero and the temperature was 38 °C. Abdominal examination showed generalized abdominal tenderness.

The abdominal and pelvic CT scan (Fig. 1) showed acute appendicitis with a stercolith measuring 37x35 mm, with discrete loop agglutination around the appendix and a moderate amount of effusion in the right iliac fossa and pelvis.

The patient had undergone ileo-caecal resection, double gun barrel ileo-colostomy, peritoneal cleansing with saline and drainage of the right iliac fossa and CDS of Douglas by 2 redon drains.

On examination, a medium-sized peritoneal effusion of purulent fluid was found in the right parietal-colic gutter and pelvis, false membranes in the inter-anal area and a cystic appendicular mass "suspicious appendicular mucocele" (Fig. 2 and Fig. 3).

Pathological examination showed a proliferation of organoid architecture suggestive of a neuroendocrine tumor measuring 4 mm long axis infiltrating the mucosa, submucosa and muscularis propria without extending beyond it and no lymph node metastases "0N+/29N": classified p T1N0.

The clinical evolution judged on regular controls was favorable without recurrence over a period of 26 months.

III. DISCUSSION

Primary neuroendocrine neoplasia (NEN) can develop in any organ of the digestive system. According to the most recent data from the North American Epidemiology Registry (SEER), the most common digestive NENs are those arising in the small intestine or rectum (age-standardised incidence rate of about 1.2/100,000/year each), pancreas (about 0.8/100,000/year) and stomach or appendix (about 0.4/100,000/year each). Other locations, such as the oesophagus, liver or bile ducts, are exceptional [3]. Neuroendocrine tumour of the appendix is a rare entity [4].

The annual incidence of neuroendocrine tumours of the appendix is estimated to be 0.15 up to 0.6 cases/year per 100,000 people [5]. Their prevalence is estimated to be between 0.3 and 0.9% of appendectomy patients; 95% of appendiceal carcinoid tumors are less than 2 cm in size and 75% of them are located in the distal third of the appendix [6]. The predominance of male patients in our series - four to
three - is unusual because of the small series [7].

The age of the patient at diagnosis is also unique in appendiceal carcinoid tumors, which are often seen in young adults [8]. There are no specific clinical signs or symptoms associated with appendiceal carcinoids. Most NETs of the appendix are asymptomatic and usually discovered incidentally during appendectomy or other benign pelvic surgery. More than 50% of carcinoids mimic acute appendicitis with the classic right lower quadrant abdominal pain, which leads to appendectomy [9].

Carcinoid syndrome is very rare (<1%) [5]. In the vast majority of cases, the diagnosis of appendiceal NET is made incidentally postoperatively in appendectomy specimens that were performed due to either acute appendicitis or recurrent abdominal pain. As most appendiceal NETs are diagnosed postoperatively [10].

The 8th Edition of the UICC TNM classification (Table II) appears to be questionable for appendiceal NETs as invasion of the subserosa or mesoappendix and the size cut-off of 4 cm have not been valid [3].

TNM stage according to the ENETS 2007 classification [3]:

- T1, tumor ≤1 cm invading the submucosa and muscularis propria;
- T2, tumor ≥2 cm invading the submucosa, muscularis propria and/or locally invading (up to 3 mm) the subserosa/mesoappendix;
- T3, tumor ≥2 cm and/or extensive invasion (more than 3 mm) of the subserosa/mesoappendix;
- T4, tumor invades the peritoneum or other organs.

The risk of lymph node extension is correlated with the size of the lesion: 0% for a lesion smaller than 1 cm, around 2-3% for lesions between 1 and 2 cm, and 30% for a lesion larger than 2 cm [6].

In particular for tumors larger than 2 cm, a CT scan and somatostatin receptor scan (SRS) are recommended at 6 months and 12 months postoperatively and then annually. Colonoscopy is recommended for the early detection of large bowel tumors that are synchronous or metachronously developed [10].

The use of plasma chromogranin-A levels as a tumor marker contributes to the differential diagnosis of caliciform cell carcinoma, early detection of recurrence and long-term follow-up of metastatic disease. All patients should be examined 6 and 12 months after surgery and then annually, while follow-up should be lifelong [10].

Hematological investigations, biochemical or urinary investigations are only indicated in rare carcinoid syndrome to monitor disease progression [9].

Treatment involves surgical removal of the tumor from the healthy margin. The extent of the resection is determined by the risk of lymph node metastasis. Tumors less than 1 cm in diameter are considered to be at low risk of lymph node involvement, and appendectomy is sufficient. Tumors larger than 2 cm in diameter are considered to have a high risk of metastasis (64% lymph node metastasis) and should undergo oncologic right colectomy.

However, the consensus of the European Neuroendocrine Tumor Society (ENETS) and the National Comprehensive Cancer Network (NCCN) is to perform appendectomy for low-risk tumors and right colectomy for high-risk ones. Risk is defined by the presence of tumor invasion of the mesoappendix, vessels, uncertain surgical margins, mixed histology or G2 grade [4].

The management of metastatic tumors and carcinoid syndrome has focused on cytoreductive chemotherapy and pharmacological control of the bioactive substances produced by these tumors. It is not the first choice of treatment for patients with carcinoid tumors. Its use should be discussed in a multidisciplinary approach linked to other available treatments [11].

The prognosis of appendiceal carcinoid tumor is much better than that of midgut carcinoid tumor [11]. The overall 5-year survival rate for all carcinoid tumors, regardless of site, was 67.2% [12].

Post-operative oncological follow-up is not necessary for tumors smaller than 1 cm. On the other hand, it is essential for high-risk tumors and involves an abdominal scan at 3 months post-operatively and then every 6 to 12 months for 10 years. A colonoscopy should also be performed at a distance from the operation to look for a synchronous tumor [4].

IV. CONCLUSION

Neuroendocrine tumors of the appendix are rare and can be discovered by chance. Its clinical picture is unrecognized, often misleading and non-specific. It is mistaken for acute appendicitis.

Although neuroendocrine tumors of the appendix are frequently rare, knowledge of the therapeutic management of these tumors is very important as they have a poor prognosis.

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