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Case Report

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Tumor carcinoide de apéndice en pediatría. Revisión de la literatura a propósito de un caso.

Carcinoid appendix tumor in pediatrics. Review of the literature about a case.

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Keywords:
Carcinoid tumor, appendicitis, appendectomy, appendiceal neoplasm, neuroendocrine tumor, pediatric tumors.

Abstract

Resumen

Los tumores neuroendocrinos aparecen con mayor frecuencia en el apéndice cecal, son de baja incidencia en la población general. Los tumores carcinoides apendiculares son asintomáticos, por lo que la mayoría de las ocasiones es un hallazgo en las biopsias de la pieza postoperatoria de las apendicectomías. Su tratamiento es quirúrgico y tienen un buen pronóstico. Se presenta el caso de un paciente de 8 años de edad con cuadro clínico de apendicitis aguda, a quien se le realizó apendicectomía. El resultado de estudio anatomopatológico confirmó tumor neuroendocrino bien diferenciado grado 1, con estudios de imagen complementarios sin evidencia de metástasis. Integrándose diagnóstico definitivo de tumor carcinoide apendicular. Por lo inusual de estos tumores en edades tempranas, se presenta este reporte de caso, subrayando la importancia del seguimiento del reporte del estudio histológico de pacientes sometidos a apendicectomía.

Summary

Neuroendocrine tumors appear more frequently in the cecal appendix, are of low incidence in the general population. The appendiceal carcinoid tumors are asymptomatic, so most of the time it is a finding in the biopsies of the postoperative part of the appendectomies. Its treatment is surgical and have a good prognosis. We present the case of an 8-year-old patient with a clinical picture of acute appendicitis. The result of anatomopathological study confirmed well-differentiated neuroendocrine tumor grade 1, with complementary imaging

studies without evidence of metastasis. Definitive diagnostic integration of appendiceal carcinoid tumor. For example, in the present article a case report is presented, highlighting the importance of the follow-up of the study of the histological study of patients in an appendectomy.

Introduction

The first description of the characteristics of the carcinoid is attributed to Lubarsh, in 1888. In 1890 the term was introduced as an attempt to highlight the "benign" behavior of these tumors and no importance was attached to their malign nature, until the end of the 1940.[1]

Gastrointestinal carcinoid tumors are a group of neoplasms that are derived from the neuroendocrine, subepithelial cells of the lamina propria and the submucosa of the digestive tract, in general they are slow growing, trabecular, glandular or in rosettes but well differentiated[1,2,3].

They are the most frequent neuroendocrine tumors (NETs) in childhood. Family history conditions four times more risk of carcinoid tumors. They occur with an incidence of 1.14: 1,000,000 cases in children per year according to Parkes et al (1993), although D'Aleo (2001), reported 1: 100,000 cases in children per year.[2,3]

They occur throughout the gastrointestinal tract, the most common sites are cecal appendix, small intestine, mainly in the ileum, rectum, stomach, colon, although it can also be located in the pancreas, bladder and bile ducts, bronchi and lung, ovary , thyroid, parathyroid, thymus and urogenital tract. Appendiceal tumors represent 0.5% of intestinal neoplasms and are found in approximately 0.08% of appendectomies [3,4,5,6.]

The clinical presentation of the appendix carcinoid is similar to acute appendicitis and is, in most cases, an incidental postoperative finding.[5, 6] It mainly affects the female sex, in a ratio of 2-4: 1.[3,4,5]

In this case report, the clinical review of a patient and the successful resolution of the condition are presented, without complications or metastasis until now.

Clinical Case

Male schoolchild of 8 years of age, originally from Colima, Mexico, antecedent of asbestos exposure during 4 years, with no other antecedents of importance for the current condition. In August 2015 he started with an enteral chart of 72 hours of evolution, upon admission to the emergency service at the Naval Hospital of Manzanillo, he presented with a globose abdomen, hyperbaralgesia, abdominal muscular resistance, generalized palpation pain, positive rebound sign, data Clinical parameters of peritoneal and paraclinical irritation with hemoglobin, 12.3 g / dl; hematocrit 36.6%; leucocytes 16,000 / mm3; neutrophils 89%; 6.2% lymphocytes and 4% monocytes; platelet count 418,000 / mm3; prothrombin time (TP) of 12.4 seconds, with International Normalized Ratio (INR) of 1 and partial thromboplastin time (PTT) of 26.8 seconds. Serum electrolytes, blood chemistry, partial urinalysis within parameters for age, simple radiography of the standing and decubitus with hydro-aerial levels, peripheral dilatation of the handles, without gas in the rectal ampulla, without pneumoperitoneum and ultrasound data. abdomen with a report of dilation of the handles, thickening of the wall of the colon, with little free fluid in the right parietocolic slide, suggestive of intestinal obstructive process.

For clinical and paraclinical data of acute abdomen, exploratory laparotomy was performed with prophylactic appendectomy and superior mesenteric ganglion biopsy, without eventualities, finding a perforated appendix with localized abscess as a trans-surgical finding.

It required postoperative intrahospital management with a double scheme of intravenous antibiotics based on cefotaxime (50 mg / kg / day) and metronidazole (30 mg / kg / day) for 7 days, as well as ketorolac-tromethamine-based analgesia (0.5 mg / day). kg / dose) alternated with acetaminophen (15 mg / kg / dose) for 3 days. During his clinical evolution, he tolerated assisted walking at 24 hours and channeled gases at 48 hours, so that enteral feeding was restarted at 72 hours postoperatively without eventualities. He completed 7 days of treatment, without complications and he left home with a diagnosis of complicated appendicitis.

During follow-up, histopathological report of cecal appendix with well-differentiated neuroendocrine tumor grade 1 located at the tip of the appendix, less than 1 cm, with extension to submucosa, with positive immunohistochemical tests for chromogranin and Grimelius stain, stage I was received.

Ambulatory studies, laboratory studies and extension cabinet were requested, with reports of tumor markers within normal parameters, as well as simple tomography of the abdomen, thorax and pelvis without evidence of metastasis. Therefore, definitive diagnosis of appendiceal carcinoid tumor was integrated.

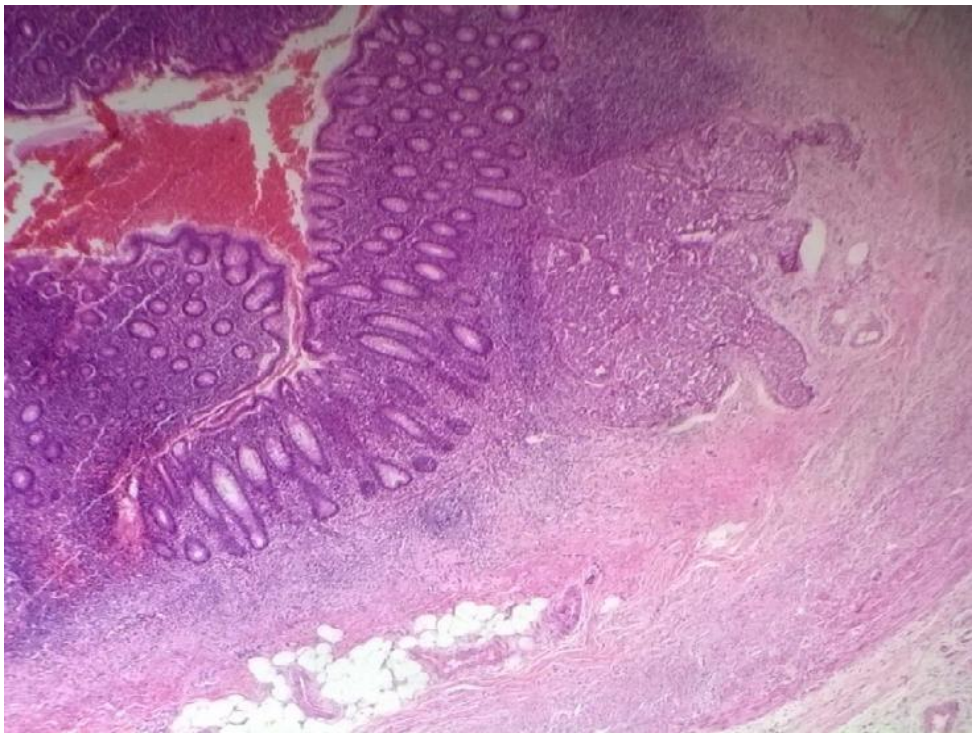

Fig. No. 1. Microscopic photograph stained with hematoxylin-eosin, mucosa of cecal appendix with reactive lymphoid tissue and a lesion of 2 mm located in the submucosa of neuroendocrine cells, which respects the muscular layer itself.

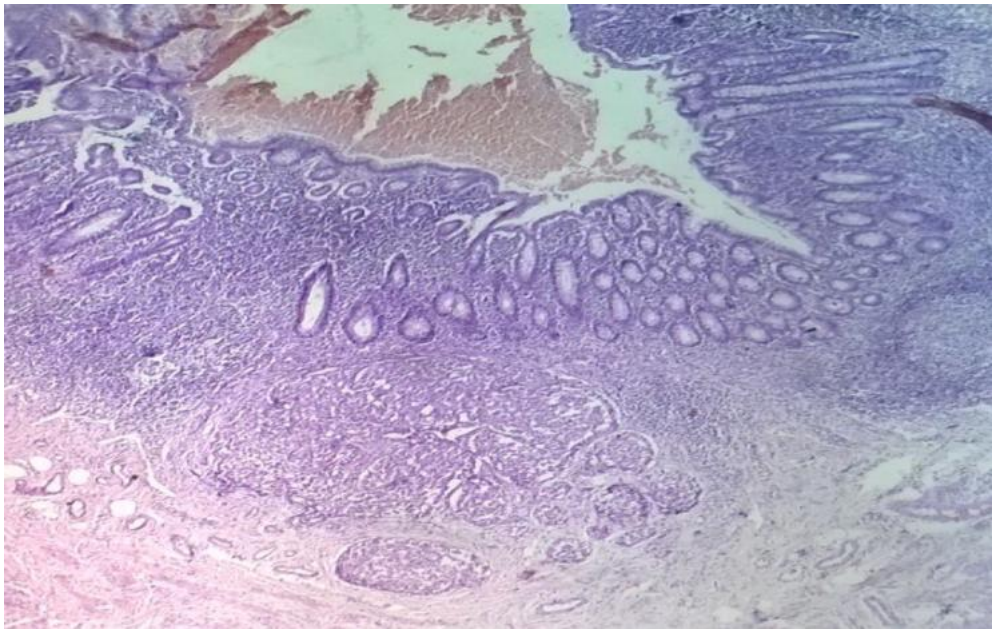

Fig. No. 2. Microscopic photography. Submucosa of cecal appendix stained with hematoxylin - eosin, with neoplastic proliferation of neuroendocrine cells.

Discussion

The clinical presentation of the NET of the appendix is similar to acute appendicitis and is, in most cases, an incidental postoperative finding [1,4,5].

Abdominal pain, nausea and vomiting are the most common presentation symptoms. The symptomatology observed in our case was compatible with the of the carcinoid tumor since they are the same as for a process of acute abdomen with suspicion of appendicitis. Carcinoid symptoms occur as a consequence of the release into the systemic circulation of vasoactive amines (histamine and dopamine), substance P and prostaglandins, with metabolic degradation directly from locally invasive or retroperitoneal small intestine carcinoid tumors by metastasis and manifested as facial flushing , abdominal pain and diarrhea, including bronchoconstriction and carcinoid heart disease, however only isolated reports are described in patients with metastatic disease. [6,7] Gastrointestinal carcinoids can present as linked to hereditary syndrome such as multiple endocrine neoplasia type 1 and neurofibromatosis type 1 and in sporadic presentations, some chromosomal alterations have been found by comparative analysis of genomic hybridization, the most common in chromosomes 5 , 11 (11q), 14, 17 (17q), 18 and 19. The most commonly reported gene mutation in gastrointestinal carcinoids is β-catenin (CTNNB1) .7 In the above case, the location of the appendix NET corresponded to the most frequent location according to the study by Moertel et al. being its most common location the distal portion in 71%, in the middle portion 22% and in the proximal portion 7%. 80% are <1 cm in length, only 6% are> 2 cm.1 Infiltration through the layers of the appendix wall in periappendiceal fat or mesoappendix has been reported in up to 63% of children.6 It is known that the cells of most carcinoid tumors have well-formed secretory granules and that most contain chromogranin A, synaptophysin and neuron-specific enolase, as well as serotonin. [3, 9, 10,11] and that these are considered by the World Health Organization (WHO) as markers for the differentiation of neuroendocrine neoplasms and at the same time they are the most common messengers of these cells.[10] In Prommegger et al. (2002) showed that serotonin is the substance that in most percentages of carcinoid tumors is positive, followed by chromogranin.[11,12] For treatment, appendectomy is considered curative, being the laparoscopic technique, currently the most used. The NET of the appendix was not suspected at the time of surgery in all patients so no samples were taken from the lymph nodes during surgery. [6,9,11] The German Society of Pediatric Oncology and Hematology recommends hemicolectomy if the tumor size is> 1.5 cm, since they are more likely to spread to lymph nodes. However, with completely resected tumors, hemicolectomy is not indicated.[6,11] The rarity and lack of definite natural history of these tumors in children contributes to a shortage of recommendations in this population. Evidence suggests that appendiceal carcinoids in pediatric patients may be associated with a less malignant phenotype than that seen in adults. Although metastases are reported in regional lymph nodes in several reports of pediatric cases, patients have satisfactory results despite not receiving additional surgical treatment.[13] The size of the tumor, location, depth of local infiltration, lymphatic infiltration, presence of metastasis, histological type and age of the patient are considered prognostic variables to define the aggressiveness of the approach, however, treatment and follow-up evaluations have not been standardized, although serum markers can be used to identify residues or metastases such as serum chromogranin A and serotonin and 24-hour urinary 5-hydroxyindole acetic acid.[1,5,8,14] Has a good prognosis with an average survival at 5 years from 85.9 to 100%. In cases of unresectable metastasis, the average survival at 5 years is 21-42%. Most authors have monitored their patients for 5-10 years without recurrence, and therefore consider long-term follow-up studies unnecessary.[5,14,15,16]

Conclusion

The carcinoid tumor of the appendix is a rare neoplasm, but it is very likely that the pediatrician must face first-hand a picture of appendicitis, its diagnosis has not been standardized, due to the rarity of this tumor in the pediatric population and the lack of centralized studies. A constant genetic marker for the prognosis of gastrointestinal carcinoids has not yet been identified. The classic form of the carcinoid is the most frequent and benign, it is not necessary to follow postoperatively in small tumors (<1 cm) completely resected, also in absence of persistent carcinoid syndrome, postoperative examinations and serum biomarkers are not useful, however, there is no consensus on how to follow tumors Andes. Being an incidental finding, in histopathology reports, the physician should know the final histopathological diagnosis of the piece, since it depends on the patient's adequate channeling for follow-up.

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