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Case Report

Pathology Section

Carcinoid Tumour of the Vermiform Appendix Mimicking Acute Appendicitis: A Case Report and Review of Literature

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ABSTRACT

Although carcinoid tumours are relatively rare, they are the most common neoplasms of the vermiform appendix where they are often discovered incidentally. In symptomatic patients, however, these tumours may produce signs and symptoms that closely mimic those of acute appendicitis and thus require adequate histopathological assessment to resolve the diagnostic challenges. We review the case of a 21-year-old Nigerian male university undergraduate who presented with right-sided lower abdominal pain and vomiting a few hours before coming to the hospital. Clinical evaluation suggested a diagnosis of acute appendicitis for which an emergency appendicectomy was performed. The patient recovered and was subsequently discharged home. Histopathological examination of the appendiceal specimen, however, revealed a carcinoid tumour at the tip of the specimen. It, therefore, becomes imperative that all appendicectomy specimens be subjected to appropriate histopathological assessment even if the sample appears innocuous to the naked eye.

Keywords: Acute appendicitis, Carcinoid tumour, Incidental, Mimic

CASE REPORT

A 21-year-old university undergraduate presented with sudden onset of abdominal pain and vomiting of recently ingested food substances 8 hours before presentation to our hospital. The pain was colicky and located in the right lower abdomen. It became worse about an hour before presentation and this necessitated his search for medical attention. There were no other complaints. Physical examination revealed that he was not febrile but was in painful distress. His blood pressure was 120/80mmHg and he had a pulse rate of 80 beats per minute. Abdominal examination revealed tenderness in the right iliac fossa with rebound tenderness and guarding. There was no organomegaly. Rectal examination revealed a mild tenderness in the right lower quadrant of the abdomen but no blood or any papable mass. Examination of other systems revealed no abnormality.

A clinical diagnosis of acute appendicitis was made. The patient subsequently had an emergency appendicectomy done and was discharged three days after the surgery. The appendicectomy specimen was sent to the histopathology department for histological examination.

Gross anatomic findings

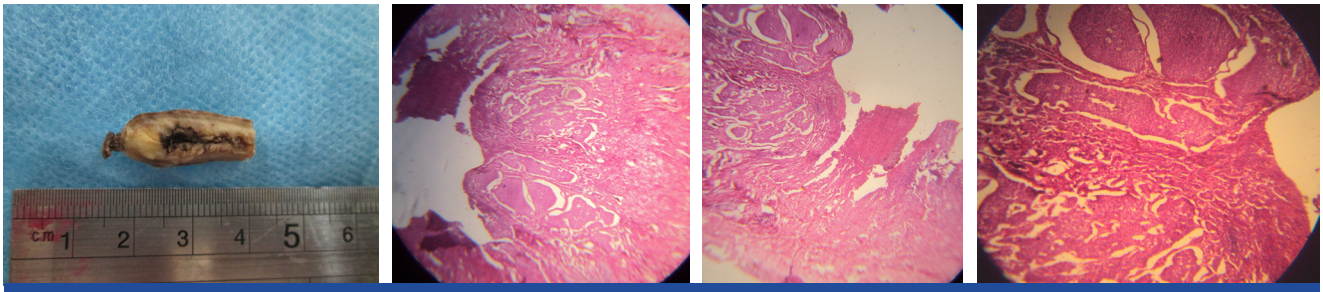
The appendicectomy specimen was received preserved in 10% buffered formal saline. The specimen measured 9cm long and 1cm in its widest diameter. The cut surfaces showed a greyish-white tissue with a narrow lumen. A firm round yellow coloured mass with a diameter of 0.5cm was also seen at the tip of the specimen [Table/Fig-1].

Histological findings

The tissue sections showed the appendix with isolated areas of mucosal ulceration as well as hyperplastic mucosal lymphoid follicles with germinal centres. The lamina propria and submucosa were infiltrated by nests, cords and trabecules of neoplastic neuroendocrine cells [Table/Fig-2,3]. These cells were fairly monomorphic and had central vesicular nuclei and scanty eosinophilic cytoplasm [Table/Fig-4]. Their nuclei had a speckled chromatin pattern giving them a salt and pepper appearance. Areas of clefting from the surrounding connective tissue stroma are also seen.

DISCUSSION

Carcinoid tumours of the vermiform appendix are relatively rare neoplasms although they represent the most common



[Table/Fig-1]: The yellowish poorly circumscribed mass can be seen at the tip of the appendix just beneath the mucosa
[Table/Fig-2,3]: Low power light microscopic view of the appendiceal tip showing the nests of tumour cells within the lamina propria, There is extensive ulceration of the mucosa. (H&E X 80)
[Table/Fig-4]: Higher power view showing nests of monotonous cells with fairly uniformly sized speckled nuclei within the lamina propria, (H&E, X 200)

primary tumours of this organ being found in 0.3%-0.9% of patients undergoing appendicectomy [1,2]. Interestingly, this is the first case of carcinoid tumour of the appendix diagnosed at our centre over a three year period. Although they affect all age groups, carcinoid tumours of the appendix are commoner in female patients with the average age at diagnosis being about 42 years [3,4].

In most instances, the clinical presentation of carcinoid tumour of the appendix is indistinguishable from that of the typical acute appendicitis [5]. This is because the tumour frequently causes intermittent partial obstruction of the appendiceal lumen which then initiates the series of events similar to those of typical acute appendicitis. Therefore, in most cases where carcinoid tumour is eventually diagnosed, the initial clinical impression is usually that of acute appendicitis. It is well-known that tumours located at the tip of the appendix and measuring less than 1cm usually mimic acute appendicitis, while those measuring more than 2cm and located at the base of the appendix may present with clinical signs of peritonitis [6]. This was the case in our index patient who presented with a 0.5cm tumour located at the tip of the appendix and presenting with fever and a sudden onset of pain in the right iliac fossa. In fact, it is well-known that carcinoid tumours of the appendix are often asymptomatic and found incidentally in appendicectomy specimens including those removed as part of other intra-abdominal surgical procedures [7-9]. On the contrary, symptoms of carcinoid syndrome such as flushing, diarrhoea and cardiac disease, which are typically associated with liver or retroperitoneal metastasis, have been rarely reported [10,11]. In such cases increased urinary excretion of 5-HIAA may assist in the diagnosis and monitoring of the disease. It is, therefore, vital that all appendicectomy specimens be subjected to histopathological assessment irrespective of the degree of clinical and/or surgical convictions.

Although carcinoid tumours rarely metastasize, the site and size of the tumours are the most significant parameters for pathological assessment. In fact, acceptable indications for more aggressive surgical re-intervention after its incidental discovery in appendicectomy specimens include lesions greater than 2cm, histological evidence of mesoappendix involvement, tumours located at the base of the appendix and showing surgical margin involvement or involvement of the caecum, high grade tumours and goblet cell adenocarcinoids [12]. The tumours are located at the appendiceal tip in approximately 75% of cases while in 20% and 5% of cases respectively, they are located in the mid portion and the appendiceal base [12,13]. Tumours smaller than 2cm very rarely metastasize and are treatable by radical appendiceal resection while those greater than 2cm in diameter are usually associated with widespread metastasis requiring more aggressive treatment options [14]. Tumours less than 1cm in size demonstrate a risk of metastatic disease of almost 0% and require only simple appendicectomy [15]. Almost all appendiceal carcinoid metastases are restricted to regional lymph nodes and are found at the time of first tumour presentation [15]. Our patient seems to have good prognostic indices since she presented with a tumour 0.5cm in size and located at the tip of the appendix. Although he did not present with symptoms of metastatic disease, the involvement of the mesoappendix by the tumour may portend an unfavourable outcome.

From a histogenetic viewpoint, the tumour cells originate from the subepithelial neuroendocrine cells located mainly at the tip of the appendix unlike in other parts of the gastrointestinal tract, thus lacking any anatomic relationship with the overlying appendiceal mucosa [16]. They also show weak to moderate expression of low-molecular weight cytokeratins although they are typically closely associated with scattered, interstitial S-100 positive sustentacular cells just like those seen in phaechromocytomas [16].

CONCLUSION

This report shows that it is imperative that all appendicectomy specimens be subjected to appropriate histopathological assessment irrespective of the circumstances under which

they were excised since majority of carcinoid tumours are discovered incidentally and the tumours are known to mimic acute appendicitis in symptomatic patients. This is especially important in a resource poor setting like ours where available histopathological services are not only limited and inadequate but grossly underutilised [17].

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