BJMHR
British Journal of Medical and Health Research
Journal home page: www.bjmhr.com

A case of appendiceal neuroma (fibrous obliteration of appendix)

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ABSTRACT

Acute appendicitis is the one of the most common appendiceal disease and abdominal surgical condition. One of the lesions that are found after removal of the appendix is fibrous obliteration or appendiceal neuroma. Nearly 30% of the resected specimens were observed to have this lesion. Herein we report one case of appendiceal neuroma in a woman with classical appendicitis like symptoms.

Keywords: Appendiceal neuroma, neuromata, fibrous obliteration.

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Received 12 August 2016, Accepted 23 August 2016

Please cite this article as: Hairon SM *et al.*, A case of appendiceal neuroma (fibrous obliteration of appendix). British Journal of Medical and Health Research 2016.

INTRODUCTION

Appendiceal neuroma is a relatively common entity characterized by obliteration of the lumen of the appendix by profileration of neural tissue. In one study, 7.1% of the appendixes removed from the liver donors were observed to have fibrous obliteration. In one retrospective study stated that 30% of the appendix specimens are diagnosed with fibrous obliteration [1].

Case report

History and physical examination

The case that we presented in this case report is about a 22-year-old lady who presented with abdominal pain for five days prior to admission. The pain is classical appendicitis-like pain which is first noted at the epigastric region and spread to the right iliac fossa. She also complaint of nausea, vomiting, diarrhea and poor appetite for the same period of the pain. Patient however remains afebrile. The full blood count and renal profile were unremarkable. Ultrasound examination shows no evidence of acute appendicitis and no fluid collection in the peritoneal and pelvic cavities. Intraoperatively, the appendix appears to be normal. There is no adhesion noted among the bowel.

Histological finding

Grossly, the appendix measures 30mm in length and 5mm in widest diameter. There is an attached meso-appendix measuring 25x4mm. The appendix appears unremarkable but the lumen is obliterated by whitish material. The mucosal lining of the appendix, especially at the body and the tip is diminished. It is replaced by extensive fibrosis with occasional fatty tissue. Within the fibrotic area, there are neural differentiated cells showing wavy-spindle nuclei. They show reactivity toward neuron-specific enolase (NSE) immunostaining (figure 1-3).

DISCUSSION

Appendiceal neuroma was described by P. Massonin an article published in American Journal of Pathology [2]. In the article, he analysed 1200 specimen of appendix that include normal and pathological specimens. Some of the pathological specimens are appendiceal neuroma (neuromata). The pathological specimen which are diagnosed with neuromata or appendiceal neuroma, when examined under the electron microscopy, it is found out that the S100 protein and neuron specific enolase positive cells have somatostatin and serotonin secretory granules within the cytoplasm. It is also observed that there are two types of cells which make bundles of cells in appendiceal neuroma – i.e. the schwann cells and the cells with neurosecretory granules.

Somatostatin containing cells are widely found in the gut and pancreas [3]. They have paracrine mode of action. Somatostatin positive cells are seen to have foot processes that wraps around the target cells. When the granules is released, it will act on the target cells in a paracrine manners.

Serotonin also act as a paracrine messenger. It is mainly found in entero-chromaffin cells. It acts on both extrinsic and intrinsic neuron of the gut causing peristaltic movement.

It is unclear why appendiceal neuroma presented with appendicitis-like symptoms. These symptoms is persistently similar in some case reports [4]. Fibrous obliteration of the appendix (or appendiceal neuroma) is an unusual case. In one study, after the author examined 2752 cases of appendectomy, 157 cases was diagnosed with unusual pathological diagnosis. Out of the cases, 80.2% was diagnosed with fibrous obliteration of the appendix [1].

Even though some of the appendiceal neuroma cases give rise to appendicitis-like symptoms, the other remain silent. Upon examination of appendix specimen taken from 338 living donor for liver transplant, it is found out that nine of them was diagnosed with fibrous obliterated appendix and one was diagnosed with appendiceal neuroma [5]. Neuroendocrine tumor is another tumor that arises from enterochromaffin, serotonin positive cells. Some carcinoid-adenocarcinoma cells also expressed the similar granules (serotonin). In view of the similar cell that giving rise to neuroendocrine tumor and appendiceal neuroma, the possibility of neuroma transforms into neuroendocrine tumor could be postulate.

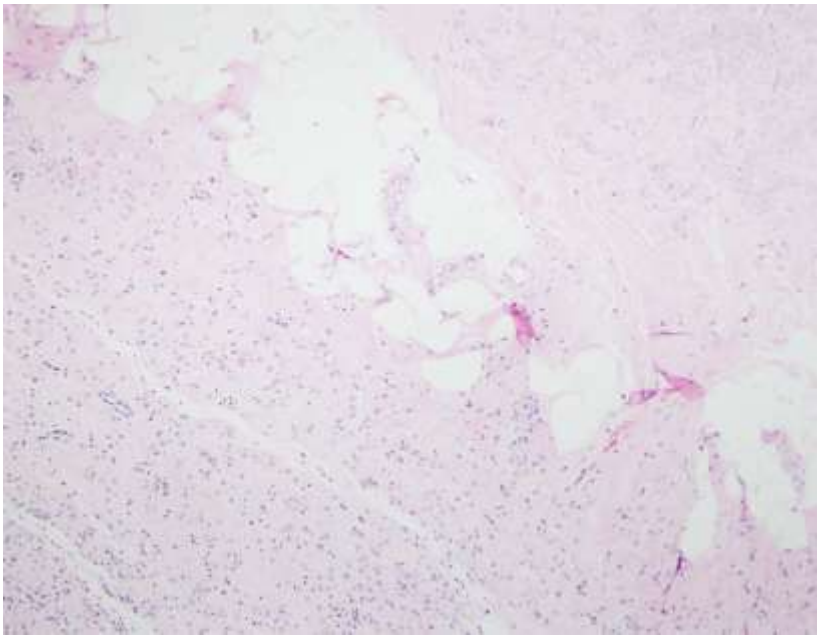

Figure 1: The appendiceal wall is replaced by fibrous and fatty tissues. (hematoxyline and eosin, original magnification x40).

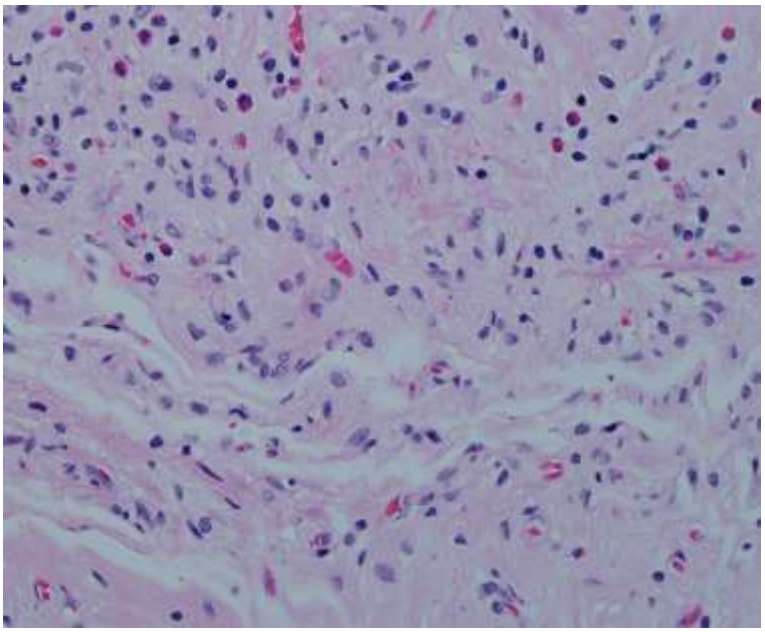

Figure 2:There is a mixture of fibrocytes and neural differentiated cell – wavy and spindly nuclei. (hematoxyline and eosin, original magnification x400).

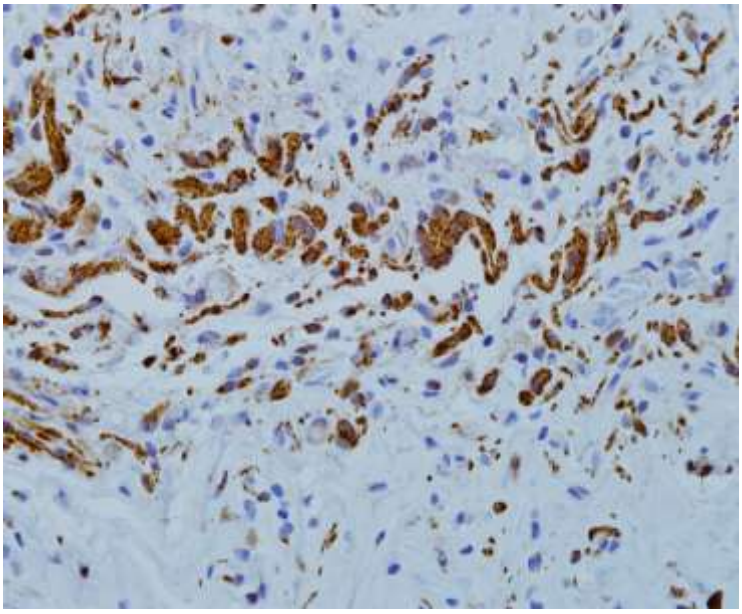

Figure 3: Immunohistochemical staining with neuron-specific enolase (NSE) highlighted and confirmed the wavy and spindle cells are of neural in origin. (NSE immunohistochemical staining, original magnification x400).

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