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Case Report

Appendiceal neuroma: an uncommon entity

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ABSTRACT

Appendiceal neuroma or neurogenous hyperplasia of the appendix is an uncommon entity. We report an incidental finding of an appendiceal neuroma in a 35 year male patient who underwent appendectomy. On microscopy showed thickened appendicular wall with well circumscribed submucosal nodules of proliferating spindle cells in myxoid areas, eosinophilic cell infiltration and obliterative appendicitis. It was reported as appendiceal neuroma. We are presenting this case for its uncommon entity, clinical and histopathological finding.

Keywords: Histopathology, Neurogenic hyperplasia, Obliterative appendicitis, Pain abdomen

INTRODUCTION

Masson in 1928 first described the findings of neurogenous hyperplasia of appendix.[1] It is characterized by obliteration of the lumen of the appendix by proliferation of neural tissue. It represents hyperplastic proliferation of the enterochromaffin like endocrine cells and non myelinated nerves and Schwann cells.[2] The repeated subclinical inflammation of appendix are considered to trigger this proliferation.[3] Appendiceal neuroma is not a very well-known entity and histopathological evaluation plays an important role to diagnose this condition.

CASE REPORT

A 35 year male patient presented with a history of right lower quadrant abdominal pain, generalized malaise, vomiting and anorexia of 3 months, with sudden increased in last 2 days. On physical examination showed localized tenderness in the right lower abdominal quadrant. Patient was having mild fever 3 days. There was no significant past or family history. No history of Neurofibromatosis type I, Cowden syndrome, MEN 2B syndrome, Ganglioneuromatosis etc. On USG abdomen pelvis showed distended appendix with narrowed lumen and periappendiceal mesenteric fat infiltration was noted. No other systemic disease was detected. Patient underwent appendectomy. On gross showed appendix with attached mesoappendix measures 4 cm in length and 0.5 cm in diameter. External surface was grey white to grey brown and showed congestion. Cut section showed obliterated lumen and fibrotic changes at the tip.

On microscopy showed wall of appendix, the mucosa was flatten at places with prominent submucosal circumscribed, nodular lesion composed of proliferative spindle cells arranged in short fascicles (Figure 2).

The cells were elongated spindle having wavy nuclei and scant cytoplasm (Figure 3).

Focal myxoid change was noted. There was increased in eosinophilic infiltrate in wall. Areas of adipose tissue and fibrosis were noted (Figure 4). Serosal surface was unremarkable.

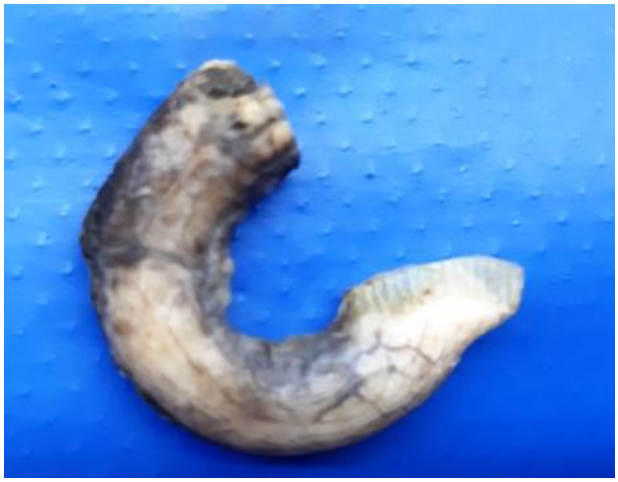

Figure 1: On gross showed appendix 4 cm in length the external surface was grey white to grey brown and showed congestion.

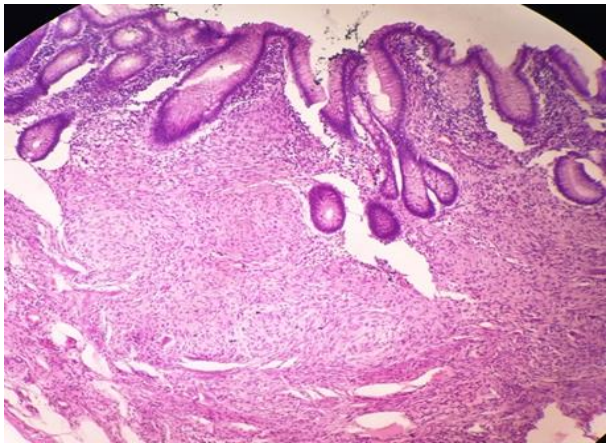

Figure 2: Microscopy showed wall of appendix, with prominent submucosal circumscribed, nodular lesion composed of proliferative spindle cells arranged in short fascicles (H&E stain, 40x).

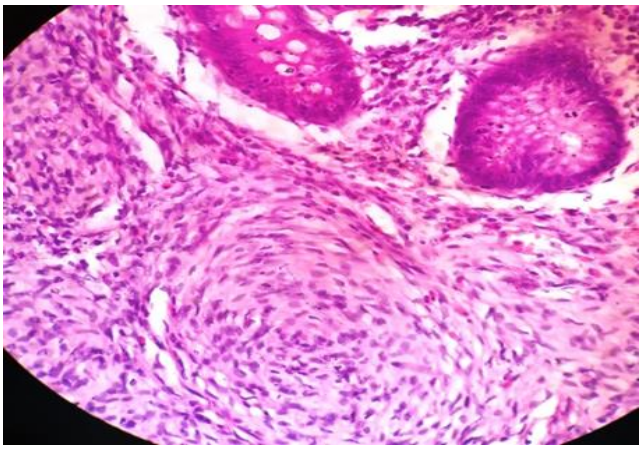

Figure 3: Microscopy showed wall of appendix, with prominent submucosal circumscribed, nodular lesion. The cells were elongated spindle having wavy nuclei and scant cytoplasm(H&E stain,100x).

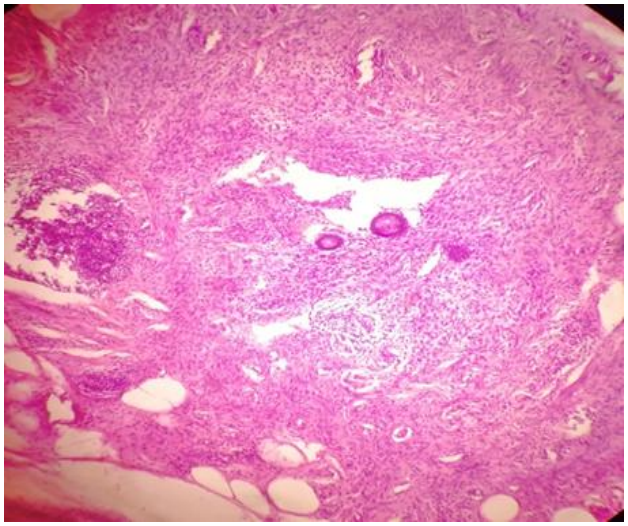

Figure 4: Microscopy showed wall of appendix, submucosal circumscribed nodular lesion with obliterative changes (H&E stain, 40x).

DISCUSSION

Appendiceal neuroma or neurogenous hyperplasia of appendix is a proliferative lesion and not to be considered as tumor. The pathogenesis of the process remain unknown. Various study stated it is secondary to inflammation giving rise to hyperplasia of neuroendocrine cells. Many time neuroma shows endocrine cells within hypertrophied nerve bundles. On clinical presentation patient present with pain abdomen, vomiting or repeated attacks of acute appendicitis. It is reported more in male than female, within adolescents and adults.[4,5] On gross lesions are mostly of obliterative type with fibrosis of appendix. Others are intramucosal lesions with patent appendiceal lumen. The fibrotic specimens are considered as end-stage of this process. Repeated subclinical attacks of inflammation are thought to trigger this lesion. On microscopic examination shows submucosal hyperplasia consists of proliferation of spindle cells, elongated cells arranged in fusiform or nodular pattern. The background may contain myxoid material, adipose tissue, fibrosis, mononuclear cell infiltrate or predominant eosinophilic infiltrate.[6] The fibrous obliteration is predominant in advance stage of disease. The various condition causing neurogenic appendicopathy related to intestinal nerve lesion like Ganglineuromatosis, Schwannoma, Perineuroma, Well differentiated neuroendocrine tumor Neurofibromas of Von Recklinghausens disease, Mucosal neuroma of MEN 2B syndrome, etc should be carefully look for.[7,8] Appendiceal neuroma is consider to be potentially a precursor to carcinoid. The lesion shows immunoreactivity to S-100 protein and Neuron specific enolase.[7] The treatment is surgical excision of appendix which is adequate. A complete surgical resection by open appendectomy or laparoscopic procedure is done. Follow-up care with regular screening may be recommended.

CONCLUSION

Appendiceal neuroma is an uncommon entity and it requires proper histopathological evaluation in an appendectomy specimens as most of these cases present as fibrous obliteration.

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Ethical approval: Not required

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