

Appendiceal Hypoplasia - An Extremely Rare Appendiceal Anomaly

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ABSTRACT

Appendiceal anomalies are extremely rare malformations that are usually found incidentally. Agenesis and duplication of the appendix has been well documented, however the reported cases of appendiceal hypoplasia are very limited, as only few cases have been reported so far. Here, we report a 31-year-old male who underwent appendectomy after clinical presentation and diagnosis of appendicitis. The appendicular specimen showed all histomorphological features of appendiceal hypoplasia. Appendiceal hypoplasia is often missed and usually difficult to be diagnosed in the preoperative workup. The chief concern in this case is to focus on the classical histo-morphological features of appendiceal hypoplasia, especially in view of its clinical course.

KEYWORDS: Hypoplasia, Presentation, Absence mucosa.

INTRODUCTION

Congenital malformations have traditionally been divided into categories of "major" and "minor", based on the clinical significance of different conditions. Hypoplasia is generally defined as a condition of arrested development in which an organ or part of an organ remains below the normal size or in an immature state.¹

Hypoplasia is a rare congenital appendiceal anomalies and only a few data are available about the appendiceal hypoplasia and development of all appendiceal layers.² Lazar J. et al (1997) describes on the commonest congenital appendiceal anomalies in the clinical scenario, briefly describing different types of anomalies e.g. hypoplasia, complete absence (agenesis), appendix multiplex, and horseshoe anomalies.³

Arnold G. et al (2012) describes some valuable anatomical information of the appendix in adult age group, which are needed to verify the diagnosis of appendiceal hypoplasia and agenesis. The stated

average length of the appendix is 8cm, ranging from 2 to 20cm. The described caliber of normal appendix ranges from 5 to 10mm (Lazar J. et al (1997).

Case presentation

A 31-year-old male presented to the emergency room with abdominal periumbilical pain followed by anorexia and minimal nausea. The pain was radiated to his right- lower quadrant (RLQ). Upon physical examination, he had point tenderness in his right lower quadrant; without rebound tenderness, guarding or rigidity. His white blood cell count was mildly elevated without leukemoid shift. A urine analysis did not reveal any abnormalities. An ultrasound was performed, which visualized very thick hard mesentery at distal part of the appendix with no obvious inflammatory changes.

Operation finding reveals very small caliber appendix with regards to his age.

Grossly, the vermiform appendix is thin, tall, firm and worm-like tubular structure measuring 3.5x0.2cm, embedded within congested lobulated / minimal fatty tissue. Cut section reveals small caliber with inconspicuous lumen. No definite lymph node is included. Figure 1.

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Fig-1: the appendix is thin, tall, firm worm-like tubular structure, measuring 3.5x0.2cm, embedded within congested lobulated minimal fatty tissue.

Histological examination: section of the vermiform appendix showed tubular structure with extremely narrow and patent lumen, which is lined by inconspicuous flattened epithelium with a few red blood cells. This is surrounded by layers of smooth muscularis propria with thinner outer one. No

definite characteristic colonic glandular mucosa lining or other lymphoid tissue components were detected. No significant inflammation is noted. No infectious agents or granuloma detected. No evidence of atypia or malignancy seen (Fig 2)

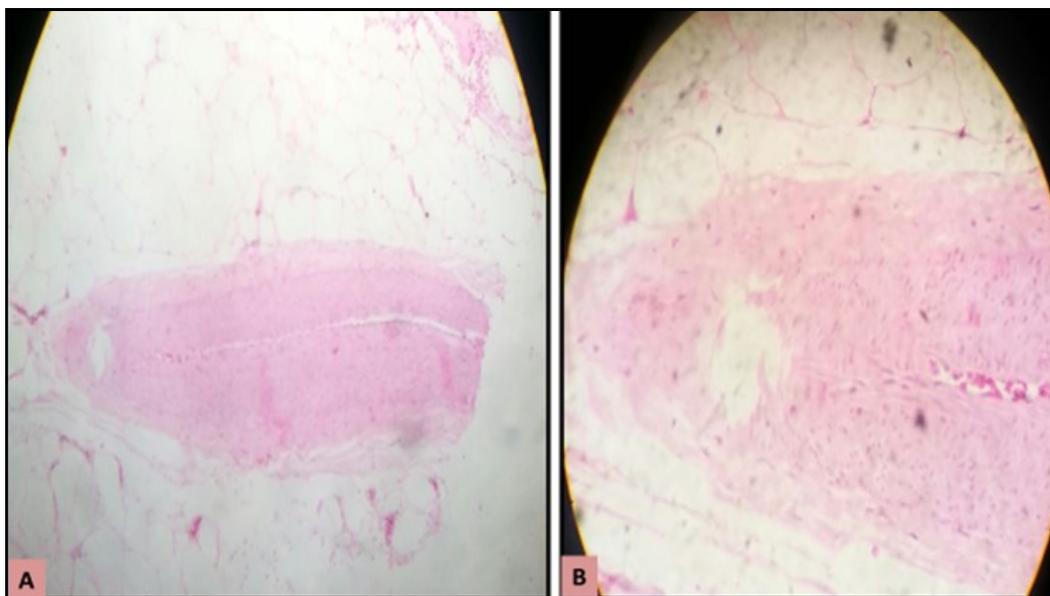


Fig-2: Appendix. A and B photomicrograph (Low and high power respectively) show transverse section of the vermiform appendix with blind-ended tubular structure.

The lumen is extremely narrow and patent, which is lined by inconspicuous flattened epithelium with a few red blood cells. No definite characteristic colonic glandular mucosa lining or other lymphoid

tissue components detected. No significant inflammation is noted. No infectious agents or granuloma detected (Fig 3).

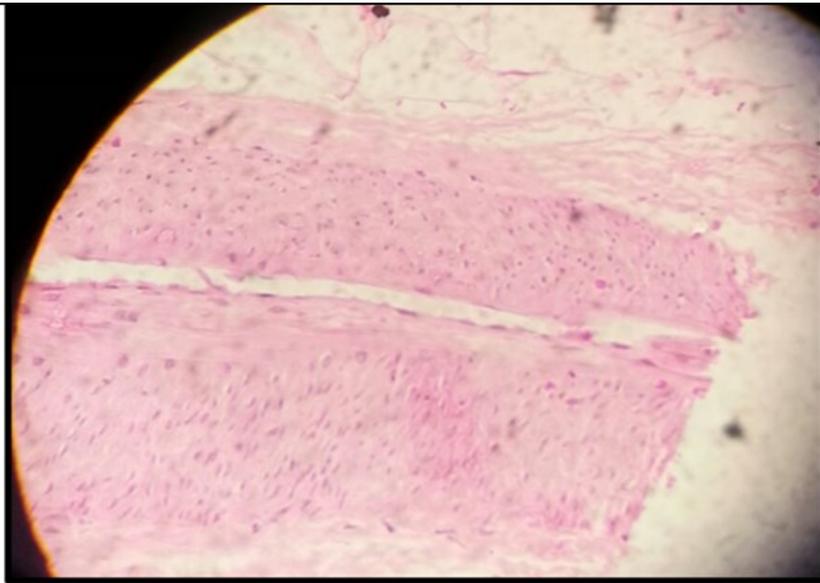


Fig-3: the appendix showed extremely narrow and patent lumen, which is lined by inconspicuous flattened epithelium. Absence of the characteristic colonic glandular mucosa lining and other lymphoid tissue components.

The appendix (Fig 4) surrounded by muscular coat, which is composed of two layers of muscularis propria with inner thick layer and thinner outer one.

The above mentioned histomorphological features are in keeping with appendiceal hypoplasia and are not typical of normal histology of the appendix.

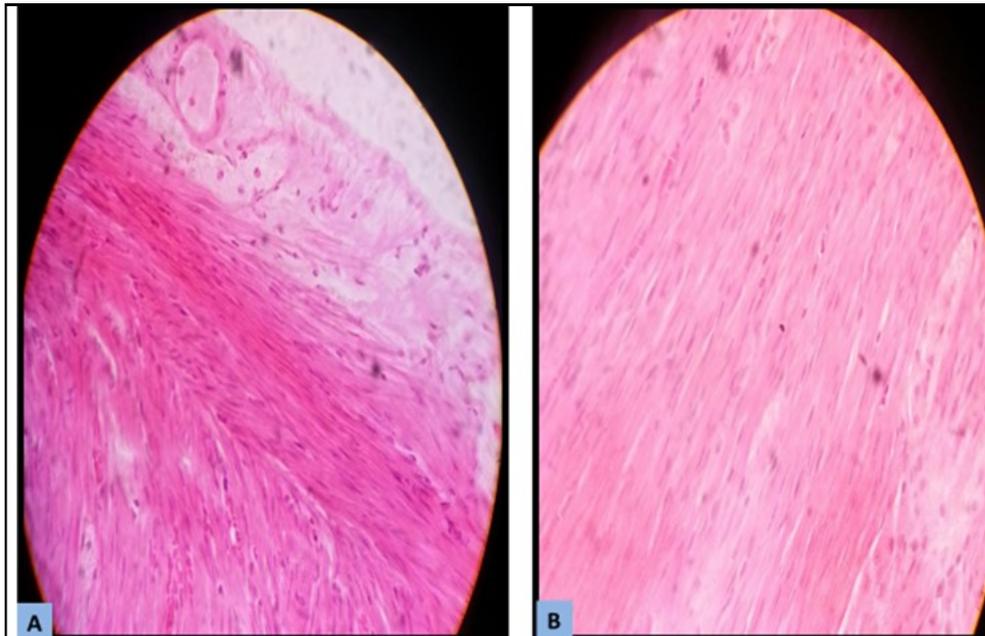


Fig-4: Appendix. A, B photomicrographs of transverse section of the appendix, exhibiting muscularis propria composed of inner thick circular muscular layer and outer thin longitudinal muscularis layer.

DISCUSSION

Congenital disorders of the appendix are rare conditions when compared to other gastrointestinal tract anomalies. In this aspect appendiceal hypoplasia is also not well described and only a few scientists have commented on the subject. Collin (1955) describes and analyzes 50,000 appendectomy specimens excised from different

age groups and he reflects how rare appendiceal congenital anomalies are. No evidence of appendiceal hypoplasia were reported among the 50,000 specimens, while only four (4) conditions of appendiceal agenesis were detected (0.008%) and two (2) conditions of appendiceal duplication (0.004%).⁴

General observation from most of the routine appendectomy specimens received in our histopathology department is that the clinical impression of appendicitis is based on the clinical examination, imaging and diagnostic laboratory investigations.⁵ There is high percentage of consensus between the final histopathology diagnosis and the preoperative impression for almost all cases, except in very few cases found in chronic condition where at most only very few congenital disorders are recorded.

In our case the patient was observed and confirmed clinically as a case of acute appendicitis and all preoperative work-up was done as usual; but the histopathology examination reported congenital malformed appendix with no features of inflammation in the background. Microscopic studies of the longitudinal and cross sections of the appendix strongly support the diagnosis of appendiceal hypoplasia rather than any other acquired medical condition. The possibility of having acute inflammation or any other type of inflammation was totally excluded.

Hypoplasia of appendix, like other congenital disorders of appendix is usually impossible to detect in the preoperative period.⁶ Further medicolegal issues should be avoided by analyzing different sources of the typical symptoms of abdominal pain.⁷

Categorization of appendiceal hypoplasia needs detailed histomorphological description with exclusion of any obvious source of acquired conditions. This can be achieved by documentation of the morphological changes and variation noticed in comparison with the normal appendix as a reference. The following points can reflect some of those differences as shown in our case:

- In hypoplastic appendix, the normal histomorphological architecture including all the layers of normal appendix are disturbed (mucosa, submucosa, muscularis and serosa).⁸
- The mucosa along with crypts and the argentaffin and nonargentaffin endocrine cells are absent. Also the submucosa was totally absent, while four well-organized layers are seen in the normal appendix.
- The submucosal lymphoid aggregate was absent. However, prominent lymphoid aggregate was found in the normal appendix.

There is a well-defined slightly thickened inner circular smooth muscularis layer with inconspicuous absent outer longitudinal muscular layer, whereas normal appendix shows both layers with adequate thickness like other portion of the gastrointestinal tract.⁸

Histopathology features of appendiceal hypoplasia can be clustered in the following points: [a] Extremely thin and elongated appendix [b] The lumen will be patent and rather narrowed; and as shown in this case, the average diameter is 2mm (< 5mm). No other elements of obliteration or occlusion can be seen along the whole length of the appendix. [c] The mucosal lining is not well developed and no obvious submucosal lymphoid tissue is seen. [d] There will be thick prominent inner circular layer and rather thin inconspicuous outer longitudinal muscular layer.

CONCLUSION

Appendiceal hypoplasia is a rare congenital anomaly of the appendix and it has not been well described in current literature.

The chief concern in this case report is to address the presentation and the classical histomorphological features of appendiceal hypoplasia in relation to its clinical presentation.

Conflict of interest

The authors have no conflict of interest to declare.

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