‘Isolated’ Appendix - An Unusual Presentation

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Abstract
Appendix is blind muscular tube with all layers of intestine. Morphologically it is the underdeveloped end of Caecum and is considered as a vestigial organ. It arises as a diverticulum from the post-arterial segment of the midgut loop. The caecum and the appendix are formed by enlargement of this bud. But due to differential growth of the walls of this bud, the lateral (right) wall growing more rapidly than the medial (left) wall causes the point of attachment of the appendix to lie on the medial side of caecum, resulting in a tubular structure of varying length from 6 to 9 cm. Rotation of the appendix commonly results in a retroperitoneal but intraabdominal structure. In approximately one quarter of the cases, rotation of the appendix is incomplete resulting in pelvic, subcaecal or paracaecal position. Mesoappendix arises from lower surface of the mesentery of terminal ileum and contains appendicular artery at its free end. Occasionally accessory artery may be present.

Introduction
Normal variations of appendix are mainly with respect to the position of its tip to the caecum while base of appendix is constant. They are:

- Retrocaecal (74%)
- Pelvic (21%)
- Paracaecal (2%)
- Subcaecal (1.5%)
- Preileal (1%)
- Postileal (0.5%)

These anatomical variations have significant clinical importance in the context of acute appendicitis. Unusual positions of the caecum including those where it lies in the right hypochondrium or in the left iliac fossa (in situs inversus) causes diagnostic difficulties if appendicitis develops.

Unusual presentations of appendix are very rare. They are:

- Duplication of appendix either at the tip or complete 0.004.1
- Diverticuli are noted in 0.014.2
- Complete absence of appendix is seen in 0.0009%.3

However, ‘Double blind’/ ‘isolated’ appendix with complete mucosal discontinuity is not documented in the literature.

Case Report
A 65 year old female came to the OPD with a classical history of acute appendicitis one and half month back (viz. pain at right iliac fossa, fever, vomiting) that lasted for 3 to 4 days that settled after admission and conservative management in a private hospital. There was no associated bladder or bowel complaint. There was no history of anorexia, wt loss or evening rise in temperature. Patient was menopausal since 15 years. No operative interventions in past. Patient had come for elective appendicectomy after one and half months. She was a known diabetic and was on regular oral hypoglycaemic medications. Diabetes was under control over the period.

On physical examination patient’s vitals were stable. Per abdomen there was minimal tenderness in the right iliac fossa and no palpable lump. Systemic examination was normal.

X-ray abdomen was not contributory. USG abdomen and pelvis showed single tubular structure
in right ileac fossa s/o chronic appendicitis with no other abnormality. Barium meal follow through showed non visualization of appendix. Rest of the bowel was normal. Blood investigations were normal (HB 11 gm%, CBC 8000/cmm) and blood sugars were under control. Patient was posted for elective appendicectomy.

Intraoperative findings revealed a single tubular structure in vicinity of caecum, 6 cm in length, separate from the caecum but attached to it with single fibrous strand (Figs. 1 and 2). The tubular structure representing appendix was found to be blind at both ends with complete mucosal discontinuity, with a visible appendicular artery in mesoappendix supplying it.

The fibrous strand and the appendicular artery was ligated separately and cut and the appendix was removed. The fibrous band was so thin that it ruled out any possibility of a lumen within it and hence no attempt was made to transfix it or cauterize the stump. The structure was sent for histopathological examination. Rest of the bowel, mesentery and pelvic organs were normal.

Patient’s histopathological examination revealed chronic obliterator appendicitis.

Patient was discharged and her post op course was uneventful.

**Discussion**

Congenital anatomical disorders of appendix are very rare. The only anatomical variations listed in literature and standard text books include duplication of appendix, diverticuli, complete absence of appendix and disorders of rotation of caecum and appendix with it. The exact incidence of each anomaly is debated.

True ‘isolated’ appendix i.e. one without any luminal continuity with caecum has not yet been documented. In our case the appendix in this 65 year old female was found in pelvic (preileal) position with only a fibrous strand connecting it with the caecum. The mesoappendix delivering the appendicular artery was thinned out demonstrating the vessel clearly as evident in the picture. The possibility of the proximal end of the appendix to be fibrous as sequelae to repeated infection and obliteration of the mucosa and submucosa, leaving the distal part intact can also be argued.

Efforts to find out any similar anatomical and pathological variant in literature were met with no results, which we therefore believe the case as very unusual, worth documenting.

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REDUCING EXACERBATIONS IN PATIENTS WITH COPD

The most important general health promotion strategy when managing patients with COPD is smoking cessation, which is the only intervention proven both to reduce decline in FEV1 and extend life expectancy.

As with all individuals with chronic lung disease, annual influenza vaccination is recommended for patients with COPD and pneumococcal vaccination is also suggested in light of the higher incidence of bacterial infections.

Formal exercise programmes (pulmonary rehabilitation) have been shown to improve walking distance activity level and wellbeing in patients with COPD and regular exercise should be encouraged.

Weight management, is also important, as patients with COPD who are underweight die earlier and those who are overweight are more breathless as they have to work harder to move.

Interventions to increase life expectancy are limited to smoking cessation and the provision of long-term oxygen therapy for patients who have chronic hypoxia.

Patients with more severe disease and ongoing symptoms should take both a long-acting anticholinergic and a combination long-acting beta-agonist/inhaled corticosteroid.

There is little evidence that delivery of drugs by a nebuliser is superior to delivery by an inhaler.

Ambulatory oxygen should be considered for patients with COPD who have exertional breathlessness and a significant fall in oxygen level on exercise.

Mucolytics may lead to a reduction in exacerbation frequency in patients with regular exacerbations.

Exacerbations can be treated with a 5-7 day course of a broad-spectrum antibiotic such as an aminopenicillin or macrolide. Alternatives include an oral cephalosporin or newer quinolone.