Appendico-Umbilical Fistula

We report two rare cases of fistulae from the vermiform appendix to the umbilicus and discuss the underlying etiopathogenesis.

Case 1

A full term male baby was admitted at 2 months of age for treatment of what was thought to be a patent omphalomesenteric duct (POMD). There was redness and feculent discharge per umbilicus starting 25 days of life. Fistulogram delineated a track towards right iliac fossa ending into a globular structure, cecum getting partially delineated in later films. Exploration through an umbilical incision with right transverse extension revealed a track communicating with the lateral aspect of vermiform appendix. Rest of bowel and intra abdominal structures was normal. The appendix and the track were removed and the patient made good post operative recovery. Histopathology confirmed a POMD opening into vermiform appendix.

Case 2

A 3-year-old boy with a normal birth history was referred for recent onset feculent discharge per umbilicus for 20 days. Child had an episode of right lower abdomen pain associated with vomiting and fever 2 months prior which was treated with intravenous antibiotics. A fistulogram done through umbilical opening revealed an irregular track extending towards right iliac fossa. No clear distal communication with the gut could be seen.

Exploration revealed dense adhesions in the right iliac fossa and the track was found communicating with the tip of the thickened vermiform appendix. Omentum was found wrapping the junction and the adjoining structures. Appendectomy and excision of the track was performed. Child had smooth recovery and is doing well at 8 months follow up.

Histopathology revealed a fibrous track with granulation tissue communicating with appendix. Appendix showed moderately dense acute and chronic inflammatory cell infiltrate forming focal foamy aggregates.

Reports of fistulous connection between appendix and umbilicus are rare. Collins(1) reported only 3 cases in a review of 50,000 specimens of the human vermiform appendix (0.006%), but did not describe their etiology as congenital. POMD usually arises from ileum but may unusually arise from cecum, ascending colon or appendix leading to lateral appendico-umbilical, terminal appendico-umbilical or ceco-umbilical fistulae(2). Our first case represents a type of lateral appendico-umbilical fistula (AUF). Delay of involution of the omphalomesentric duct and appendico-umbilical connection of POMD has been postulated as the underlying etiology(3).

Our second patient developed AUF secondary to appendicular inflammation. Acquired AUFs are described as a result of injury; either from a clamp to the tip of the appendix within the cord, or ligature of a unrecognised exomphalos(4). Appendico-cutaneous fistulae may also occur as a result of inflammatory processes(5), associated tumors or subsequent to surgical or percutaneous drainage of appendicular abscesses(6).

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Kabuki Syndrome and Diaphragmatic Defect

Kabuki syndrome is a rare genetic disorder with characteristic facial features. Other common findings are mental retardation, postnatal progressive growth retardation, skeletal, cardiac and dermatoglyphics abnormalities(1).

A child, 13 months old, sixth in birth order, product of non consanguineous marriage, presented with failure to thrive and developmental delay. He had characteristic facial dysmorphism with high arched eyebrows that were sparse in the lateral part, long palpebral fissures, prominent and protruding ears and lip pits on the lower lip. (Fig.1). A detailed dermatoglyphic study showed an absent digital triradius ‘c’ and ‘d’. Detailed cardiovascular examination revealed an ostium secundum atrial septal defect. The CECT upper abdomen showed diaphragmatic eventration on the right side with a mediastinal shift. The X-ray of bilateral hands showed clinodactyly. The abdominal ultrasound examination was normal. Karyotype done on peripheral lymphocytes was normal. A