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The Inbred, Fascimile Crater-Duplication Cyst

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ABSTRACT

Duplication cyst is an exceptional, congenital lesion arising within the gastrointestinal tract. Of obscure aetiology, alimentary tract duplication is hypothesized to occur due to embryological aberrations arising between fourth week to eighth week of gestation. Duplication cyst can be subcategorized into cystic lesions or a tubular cyst and majority of clinically symptomatic duplication cysts are detected within first two years of life wherein cysts coated with heterotopic gastric mucosa or pancreatic mucosa manifest abdominal pain or gastrointestinal haemorrhage due to ulceration and haemorrhage within the adjoining gastrointestinal tract. Duplication cyst is characteristically layered with mucosa and a smooth muscle coat wherein the mucosal epithelium is concurrent with a segment of the gastrointestinal tract. Duplication cyst requires a segregation from Meckel's diverticulum, congenital segmental intestinal dilatation or pre-sacral tumefaction as sacrococcygeal teratoma, anterior meningocoele or dermoid cyst. Ultrasonography of classic, layered, duplication cyst wall exemplifies alternating hyperechoic and hypoechoic zones designated as the pathognomonic "gut signature" sign. Comprehensive surgical eradication of the cyst is associated with an excellent extended prognostic outcome.

Introduction

Duplication cyst is an exceptionally discerned, congenital lesion arising within the gastrointestinal tract and may be designated as alimentary tract duplication. Variation in clinical manifestation is amenable to cogent radiographic assessment and may engender ascertainment of appropriate clinical diagnosis challenging. Characteristically, the cyst is denominated by an epithelial lining concomitant to a segment of the gastrointestinal tract, a well defined layer of smooth muscle and an abutting section of gastrointestinal tract as a common wall [1]. Although exceptionally delineated within the oral cavity, heterotopic gastrointestinal cyst may be exemplified upon anterior segment of the tongue. The cyst wall is coated with epithelium lining of diverse segments of the gastrointestinal tract.

Heterotopic gastrointestinal cyst is additionally denominated as foregut duplication cyst, choristomatic cyst, ciliated epithelial cyst, lingual duplication cyst, cystic tumour of the tongue, unusual thyroglossal duct cyst, heterotopic large bowel cyst, enterocystoma or alimentary tract cyst.

Disease Pathogenesis

Alimentary tract duplication is hypothesized to occur due to embryological aberrations arising between fourth week to eighth week of gestation [2,3].

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Of obscure aetiology, several theories are posited to contribute to disease emergence. The "split notochord" theory described an aberrant separation of endodermal cells from an evolving notochord. The supposition enunciates concurrence of vertebral anomalies with duplication cysts. As per "recanalization theory", erroneous recanalization of gastrointestinal tract engenders the duplication cyst. Nevertheless, duplication cyst may arise within areas where gastrointestinal tract is devoid of recanalization during embryological development [2,3]. The "embryonic diverticula" theory delineates the existence of certain diverticula within the expanding embryo. Therefore, a persistent diverticulum may extend along with an expansive alimentary tract with consequent genesis of duplicated alimentary canal [2,3]. The "partial" or "abortive twinning" theory posits the occurrence of duplication cysts on account of an incomplete twinning of the alimentary tract. The hypothesis is supported by concurrence of anomalous genitourinary tract with colorectal duplication cysts [2,3]. Of obscure pathogenesis, heterotopic gastrointestinal cyst may arise from undifferentiated ectodermal lining of primitive stomodeum which is entrapped within the oral cavity during fourth week of foetal development [3,4]. Despite aforementioned postulates contributing to the emergence of duplication cysts, the phenomenon of layering "heterotopic mucosa" within gastrointestinal duplication cysts remains perplexing and aetiology of aforesaid cysts within diverse areas remains undetermined [3,4].

Disease Characteristics

The anomaly is associated with an incidence of around 1 in 4500 live births. Upon ultrasonography, cogent disease discernment is possible in nearly ~30% instances. A mild male preponderance is observed [3,4]. Majority of clinically symptomatic duplication cysts are detected within first two years although adults may exemplify the condition. Besides, majority of heterotopic gastrointestinal cysts are discovered within the neonatal period although few instances are asymptomatic up to adulthood [5,6]. Contingent to structural configuration, duplication cyst can be subdivided into a) cystic lesion or b) a tubular cyst. The frequently discerned cystic duplication is apparent in around ~80% instances and appears to be devoid of communication with the bowel lumen [5,6]. In contrast, the infrequent tubular duplication arises in connection with lumen of the bowel [5,6]. Duplication cyst can be subcategorized into foregut, midgut or hindgut duplications pertaining to adjoining segment of the alimentary tract. Contingent to embryological origin, duplication cysts of the foregut can be categorized into oesophageal, bronchogenic and neurenteric cysts. Ileum is the commonest location of emergence of duplication cyst followed by oesophagus [5,6].

Heterotopic gastrointestinal cyst of the oral cavity is exceptionally layered by gastrointestinal epithelium whereas aforesaid epithelial lining is frequently discerned within cysts arising within the duodenum, gallbladder, common bile duct, jejunum, Meckel's diverticulum, ileum, appendix, colon or rectum [6,7].

Clinical Elucidation

Majority of duplication cysts or heterotopic gastrointestinal cysts are clinically symptomatic within initial 2 years. Variation in clinical representation occurs due to factors such as divergent localization or magnitude of cysts or occurrence of concomitant layering with heterotopic mucosa [6,7]. Enlarged duplication cysts situated upon the tongue may engender airway obstruction [6,7]. Majority of alimentary tract duplication cysts manifest in childhood or may occur as an incidental discovery. Clinical symptoms are contingent to localization and category of cyst (cystic or tubular) along with occurrence of ectopic mucosal layer. Incriminated, symptomatic individuals exhibit abdominal pain [7,8].

Tubular duplication cyst with partial communication or singular continuation with adjacent bowel may engender chronic constipation due to accumulated intestinal contents within a blind loop [7,8]. Duplication cyst coated with heterotopic gastric mucosa or pancreatic mucosa may manifest with abdominal pain or gastrointestinal haemorrhage due to ulceration and haemorrhage within the adjoining, healthy gastrointestinal tract [7,8]. Foregut duplication cyst as an oesophageal duplication cyst can manifest symptoms such as dysphagia, vomiting, epigastric pain or haemorrhage of upper gastrointestinal tract. Compression of circumscribing anatomical structures may engender symptoms such as an enlarged tumefaction within the neck, cough, stridor or dyspnoea. Bronchogenic cyst manifests dysphagia, chest pain, cough or dyspnoea. The exceptional gastric duplication cyst represents with abdominal pain, abdominal mass, vomiting or associated features of gastric outlet obstruction [7,8]. Duodenal duplication cysts may represent abdominal pain, nausea, vomiting, jaundice or upper gastrointestinal haemorrhage. Exceptionally, features of acute pancreatitis may ensue [7,8]. Midgut duplication cysts of the small bowel arising in children is associated with nausea, vomiting, abdominal pain or a palpable mass. Upper gastrointestinal tract haemorrhage may ensue due to coated heterotopic mucosa. Acute abdominal pain may occur due to intussusception or volvulus [9,10]. Hindgut duplication cysts are exceptionally discerned within the colon and are usually layered by colonic mucosa. However, lining with ectopic mucosa may be observed. Hindgut duplication cysts manifest with abdominal pain, palpable mass, vomiting, gastrointestinal haemorrhage or features of intestinal obstruction. Malignant metamorphosis may ensue. Also, rectal duplication cyst can distinctively represent as a pre-sacral mass, imperforate anus or a rectovaginal fistula [9,10].

Histological Elucidation

Duplication cyst exhibits a characteristic mucosal layer and a smooth muscle coat. Layering mucosal epithelium is concurrent with a segment of the gastrointestinal tract. However, mucosal layer may be heterotopic and non-concordant with adjacent bowel [9,10]. Ectopic gastric mucosa may be delineated in nearly ~ 30% instances and is especially discerned in oesophageal and midgut duplication cysts. Pancreatic mucosa is frequently discerned

in gastric duplication cysts. Also, bronchogenic cyst exhibits respiratory epithelium, cartilage or bronchial sub mucosal glands [9,10]. Heterotopic gastrointestinal cyst can be hypoglossal and appears as a true cyst wherein the cyst wall is layered by ciliated, columnar, cuboidal, squamous or columnar, gastrointestinal-type epithelium or pseudostratified ciliated epithelium. The cyst may be imbued with viscous, haemorrhagic fluid and circumscribed by connective tissue [9,10].

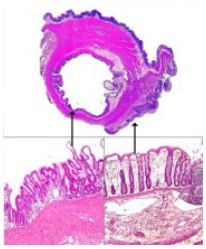

Figure 1: Duplication cyst layered by stratified and pseudostratified columnar epithelium enmeshed within fibrotic connective tissue (16).

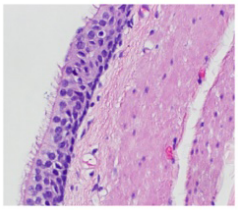

Figure 2: Duplication cyst layered by stratified and pseudostratified cuboidal epithelium surrounded by a connective tissue stroma (17).

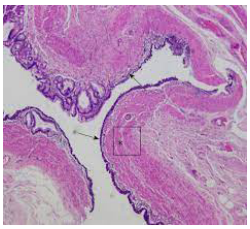

Figure 3: Duplication cyst exhibiting a lining of stratified and pseudostratified ciliated columnar epithelium and circumscribing fibrotic connective tissue (18).

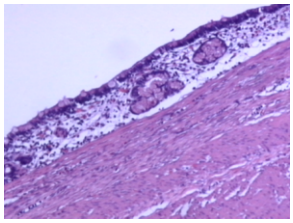

Figure 4: Duplication cyst exemplifying a layer of stratified and pseudostratified ciliated columnar epithelium enveloped by a dense fibrous tissue stroma (19).

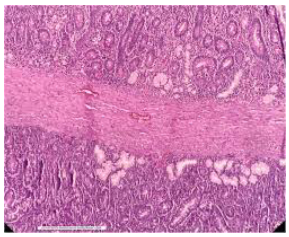

Figure 5: Duplication cyst delineating a layer of stratified and pseudostratified columnar epithelium encased within a fibrotic connective tissue stroma (20).

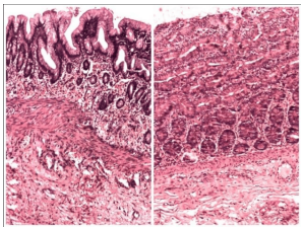

Figure 6: Duplication cyst demonstrating a lining of tall columnar epithelium with a subjacent zone of fibrous tissue stroma (21).

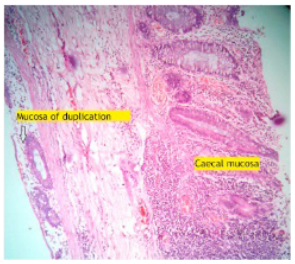

Figure 7: Duplication cyst displaying a cystic cavity lined by tall columnar epithelium resting upon a vascularized fibrous tissue stroma (22).

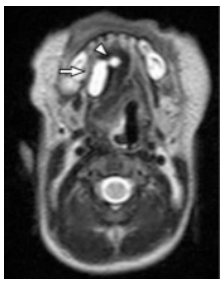

Figure 8: Duplication cyst depicting a layered cystic cavity with alternating hyperechoic and hypoechoic zones (22).

Differential Diagnosis

Duplication cyst of the alimentary tract requires a segregation from conditions such as

•Meckel's diverticulum which is situated upon the anti-mesenteric border and manifests with abdominal pain and upper gastrointestinal tract haemorrhage [2,4].

•congenital segmental intestinal dilatation characteristically depicts segmental dilatation of small or large intestine of up to ~ four times. Nevertheless, congenital segmental dilatation is devoid of cystic configuration adjacent to normal gastrointestinal tract [2 ,4].

•pre-sacral rectal duplication cysts requires distinction from diverse pre-sacral tumefaction as sacrococcygeal teratoma, anterior meningocoele or dermoid cyst [2,4].

Additionally, intrathoracic cysts as the pericardial cyst or thymic cyst are situated within middle or anterior mediastinum and necessitate differentiation. Besides, intra-abdominal cystic lesions such as mesenteric, omental, choledochal or ovarian cysts may require a segregation [2,4]. Also, heterotopic gastrointestinal cyst confined to the tongue requires a segregation from congenital cystic lesions of the tongue such as ranula, teratoma, thyroglossal duct cyst or lymphangioma [2,4].

Investigative Assay

Upon ultrasonography, the classic, layered, duplication cyst wall exemplifies alternating hyperechoic and hypoechoic zones. The aforesaid "gut signature" sign is pathognomonic for duplication cyst of the gastrointestinal tract [11,12].

Prenatal ultrasonography enunciates a sensitivity of discerning duplication cysts of around ~ 30%. Prenatal detection of heterotopic gastrointestinal cyst is extremely exceptional. Nevertheless, prenatal ultrasonography employed for detection of duplication cysts is essential in order to predict possible emergence of upper airway obstruction during labour [11,12]. An ultrasonography can be adopted for detecting foregut duplication cysts confined to the neck although procedural sensitivity is inadequate in discerning intrathoracic lesions [11,12]. Foregut duplication cyst with clinical features as an intrathoracic mass, polyhydramnios, mediastinal shift or hydrops can be adequately demonstrated upon prenatal ultrasonography [11,12]. Transoesophageal ultrasound (TEE) can be appropriately utilized to detect intrathoracic duplication cysts situated within the posterior mediastinum [11,12].

Contrast-enhanced computerized tomography (CECT) can be adopted to localize duplication cysts which appear as a hypo-attenuating mass with an enhancing perimeter or for detection and localization of deep seated rectal duplication cysts confined to pelvic cavity [11,12].

Magnetic resonance imaging (MRI) can provide superior cross sectional anatomical exemplification and is optimal for diagnosing duplication cysts of the hindgut. Prenatal MRI is advantageous in evaluating possible occurrence of airway obstruction [12,13].

Preoperative magnetic resonance imaging (MRI) can be beneficially adopted to adequately localize heterotopic gastrointestinal cyst and cystic expansion towards adjacent anatomical structures [12,13]. Technetium-99m pertechnetate ($^{99m}TcO_4^-$) scan can be employed to detect the presence of ectopic gastric mucosa. Also, multiple, synchronous duplication cysts may be discerned. Besides, bronchogenic and rectal duplication cysts may exceptionally be enunciated by technetium-99m pertechnetate scan [12,13].

Therapeutic Options

Preoperative aspiration or marsupialization of the cyst may be performed. Although cyst aspiration is efficacious for resolving initial airway obstruction and feeding difficulties, frequent aspiration of the cyst necessitates circumvention on account of possible emergence of cyst infection and damage to adjacent soft tissue [13,14]. Enlarging duplication cyst may be unamenable to repetitive aspiration and cogent surgical excision of the cyst may be optimal [14,15]. Cystectomy or surgical extermination of the cyst is a definitive therapeutic manoeuver for incriminated children [14,15]. Comprehensive surgical eradication of the cyst is associated with an excellent extended prognostic outcomes. Cogent surgical approach for cyst eradication may be an open procedure (laparotomy or thoracotomy) or a minimally-invasive (laparoscopic or thoracoscopic) manoeuver [14,15].

Antenatal or incidentally discovered, asymptomatic duplication cysts enunciated upon an ultrasound can be subjected to preliminary surgery. However, the optimal period of surgical extermination of oral, congenital duplication cysts or heterotopic gastrointestinal cysts lacks consensus. Nevertheless, timing of surgical intervention is contingent to degree of respiratory or feeding impairment and achievement of child's growth milestones and development [14,15]. Generally, prenatal detection of heterotopic gastrointestinal cyst is followed by cyst excision within one year and exhibits a favourable prognosis [14,15].

A significant proportion of aforementioned cysts may undergo intussusception, volvulus or haemorrhage. Malignant transformation may ensue [14,15]. Optimal therapy for heterotopic gastrointestinal cyst is comprehensive surgical resection. Cyst aspiration can be optimally adopted to circumvent respiratory distress or difficulty in feeding. Cyst reoccurrence is infrequent [14,15]. Symptomatic instances can be adequately treated with preliminary surgery [14,15]. Intrathoracic oesophageal duplication cysts can be treated with a posterolateral thoracotomy and simple excision of the cyst. Also, decompression of enlarged cysts is recommended [14,15]. Gastric and intestinal duplication cysts are optimally subjected to laparotomy with simple excision of the cyst. Tubular duplication cyst or lesions abutting the gastrointestinal wall can be treated with intestinal resection with end-to-end anastomosis [14,15]. Rectal duplication cyst represents with a pre-sacral mass or imperforate anus with rectovaginal fistula. Thus, a posterior sagittal or abdominoperineal access is recommended for simple excision of the cyst [14,15]. Synchronous duplication cysts within thorax and abdomen occur in nearly ~15% instances. Preliminary resection of symptomatic cysts is optimal although dual cysts can be concurrently excised [14,15].

Delay in appropriate disease discernment and surgical intervention may engender complications such as volvulus, intussusception, perforation of enlarged duplication cysts, recurrent haemorrhage or possible malignant degeneration. Surgical complications are comprised of localized wound infection, anastomotic gastrointestinal leaks and gastrointestinal obstruction due to adhesion [14,15]. Extended prognostic outcomes are excellent following optimal surgical procedures [14,15].

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