Duplication cyst in a 20 weeks fetus
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ABSTRACT
Intestinal duplication is a rare congenital anomaly. Duplications found in proximity of small intestine are the most common enteric duplications encountered and majority of these occur in ileum. They may be either cystic or tubular and most of them are located in the mesentery of intestine. We are presenting a case of 20 weeks embryo with a cyst detected during ultrasonography for routine check-up.

Key words: Duplication Cyst, Antenatal Screening.

INTRODUCTION
Duplications are rare congenital anomalies which can occur anywhere in gastrointestinal tract from mouth to anus. The reported incidence is 1:4500 births. (1) Duplications in proximity of small intestine are the most common enteric duplications and majority of these occur in ileum. They may be cystic or tubular. They may result in significant morbidity and mortality if left untreated. Approximately, two thirds of all intestinal duplications are discovered within the first two years of life with one third identified in the newborn period. However, the exact incidence is not known.

CASE REPORT
A primigravida woman with 20 weeks of gestation went for her sonography. The sonography of gravid uterus revealed a well defined, single, live, foetus in cephalic presentation and longitudinal lie. There was a large (147.7x120.1x92.1mm) thin walled, anechoic, subdiaphragmatic intraabdominal lesion. This lesion was thin walled, devoid of any intralesional contents. The mother was prepared for surgery and the baby was delivered. Regrettably, the baby did not survive.

GROSS SPECIMEN
We received a soft tissue mass measuring 11.5x10.5x0.5 cms. E/S Cystic with prominent vasculature. C/S showed a cystic structure containing clear fluid. The wall was soft white and thickened at places.

Figure 1: Cystic structure measuring 11.5x10.5x0.5 cms with smooth internal surface.

MICROSCOPIC DESCRIPTION
H&E stained sections revealed a cystic structure lined by flattened cells and a wall composed of normal layers of intestine i.e.
submucosa and muscularis. The mucosal layer was moderately atrophic and was lined by low columnar to cuboidal cells. Masson trichrome stain confirmed the presence of smooth muscle cells.

Figure 2: Photomicrograph of the cyst lined by low columnar cells and wall consisting of smooth muscle cells (10x).

Figure 3: Photomicrograph showing Masson trichrome stained section of the cyst wall demonstrating the smooth muscle cells.

DISCUSSION

Duplication cysts are defined as spherical or tubular structures that are firmly attached to at least one point in the alimentary tract, possess a well-developed coat of smooth muscle and have an epithelial lining resembling some part of alimentary tract. (1-3). Commonly duplication cysts are communicating with the intestinal lumen, involve the mesenteric border of the associated alimentary tract and share a blood supply with the native bowel. The symptoms depend on the size, location and mucosal lining of the cyst. Patients may present with pain in the abdomen, vomiting, palpable mass or acute gastrointestinal haemorrhage. Intestinal duplication in the thorax may present with respiratory distress. There are four major theories regarding origin of duplication cysts.

i. Partial twinning theory
ii. Split notocord theory
iii. Canalisation defects theory
iv. Environmental factors theory

Partial twinning theory states that organs can be double as a result of abnormal twinning. Multiple organs can be formed in such cases with multiple skeletal abnormalities.

The split notocord theory states that during third to fourth week of gestation, gaps may appear in the notocord through which endodermal cells can herniate and form diverticula. This theory may explain long duplication cysts as well as intrathoracic and foregut duplications. It proposes explanations for 15% of enteric duplications with vertebral defects.

The canalisation defects theory is based on the process by which all the GI organs begin as solid organs in the embryologic state and gradually vacuolate to form the lumen. During this process, many diverticula form in the foetal stage which regress with development. Most of these foetal diverticulae are found in the ileum, the most common site for duplication cysts. This is said to happen at 5-8 weeks of gestation.

Environmental factors theory postulates that stress, hypoxia and trauma may induce duplications.

CONCLUSION

Duplication cysts are rare anomalies with no clear cut cause known. The fate of a patient depends on their size and site. Earlier detection in utero is essential for the survival of a baby by sonography and in the hands of an experienced paediatric surgeon. In this case, there was no antenatal diagnosis due to lack of any type of regular sonography for the foetus. The baby was too premature to survive with this anomaly. Hence, the importance of
antenatal screening for anomalies cannot be stressed enough.

REFERENCES


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