

HETEROTOPIC PANCREAS IN OMPHALOMESENTERIC DUCT REMNANT: A RARE ENTITY

Histopathology

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ABSTRACT

The omphalomesenteric duct also known as the vitelline duct is a connection between yolk sac and primitive midgut during the early embryonal development. When the duct fails to regress, it causes a spectrum of anomalies broadly categorised under omphalomesenteric duct (OMD) remnants. Present case shows a rare finding of heterotopic pancreatic tissue in omphalomesenteric duct remnant. An 8 year old girl child presented with persistent umbilical discharge since birth. On physical examination a granulomatous lesion was found over umbilicus. Resected umbilical tissue on histopathology revealed presence of both exocrine and endocrine pancreatic tissue with some intestinal mucosa. Omphalomesenteric duct remnants can remain asymptomatic or present with common gastrointestinal symptoms such as umbilical discharge, umbilical swelling, recurrent infection in the umbilicus, constipation, bowel obstruction, gastrointestinal bleeding and rarely intestinal perforation. The clinical symptoms overlap with other non congenital conditions such as umbilical hernia, appendicitis, intussusception, intestinal torsion and ischemia. Therefore, in young children presenting with such clinical symptoms it is important to always consider the possibility of OMD remnants as a differential diagnosis.

KEYWORDS

Heterotopic pancreas, Omphalomesenteric duct remnant

INTRODUCTION

The omphalomesenteric duct also called as vitelline duct is a connection between yolk sac and primitive midgut during the early embryonal development. It normally disappears between the eighth to ninth weeks of development¹. When the duct fails to regress, it causes a spectrum of anomalies broadly categorised under omphalomesenteric duct (OMD) remnants. OMD remnants are found only in 2% of the total population.

Meckel's diverticula is the most common omphalomesenteric duct remnant which includes 67% of the total OMD remnant cases.² Other OMD remnants consist of OMD fistulas, umbilical cysts, congenital band or fibrous cord, superficially draining sinuses. Mostly, omphalomesenteric duct remnants do not present with any symptoms, especially during early years of life. They can clinically present with persistent umbilical discharge, umbilical swelling, pain abdomen, constipation. The knowledge of development, incidences and clinical presentation of OMD remnant is vital for early detection and better patient outcome.

Heterotopic pancreas is a congenital abnormality characterised abnormal location of pancreatic tissue. It most commonly occurs in upper gastrointestinal tract. It can rarely occur in Meckel's diverticula, biliary tree, spleen, omentum. Present case shows a rare finding of heterotopic pancreatic tissue in omphalomesenteric duct remnant causing persistent umbilical discharge since birth. As per our knowledge very few similar cases have been reported so far.

Case Report

An 8 year old girl child presented with complaints of persistent discharge from umbilicus since birth. The girl child was born after a full term pregnancy through normal vaginal delivery. The mother had an uneventful pregnancy. No congenital malformations were present at time of birth. There was no history of genitourinary or gastrointestinal complications. On physical examination of the girl child, a granulomatous lesion was found over umbilicus. Ultrasound revealed possibility of patent urachus. Laboratory findings were unremarkable. Surgically, an infraumbilical incision was made and umbilical exploration was done under general anesthesia. There was no patent urachus found. Remnants of Omphalomesenteric duct were excised while umbilical exploration and incision was closed by suture tie. Resected umbilical tissue measured 2 x 1.5 x 0.5 cm which was further sent for histopathological examination. Histologically, pancreatic tissue was identified along with some intestinal mucosa and connective tissue. Both exocrine and endocrine components of pancreatic tissue were found. Pancreatic acini, pancreatic ducts were identified as part of exocrine pancreas whereas Islets of Langerhans

confirmed the presence of endocrine pancreas. Pancreatic acini were separated into lobules by intervening connective tissue. Diagnosis of heterotopic pancreatic tissue in omphalomesenteric duct remnant was made. No malignancy was detected. The postoperative follow up was uneventful.

DISCUSSION

Omphalomesenteric duct remnants develop due to the persistence of duct even after birth. Most common among the OMD remnant is Meckel's diverticula. It is a true diverticula that develops on antimesenteric border and involves all the layers of intestine. It is usually incidental finding on radiological investigations or during exploratory laparotomy in cases where the symptoms mimic appendicitis. Sometimes, Meckel's diverticula can cause complications such as gastrointestinal bleeding. These symptoms are attributed to ectopic gastric tissue or pancreatic tissue which lines the diverticula and causes hemorrhage. Other complications are intestinal obstruction, diverticula torsion, ischemia, inflammation, intussusception.

Other Omphalomesenteric duct remnants are OMD fistulas, umbilical cysts, congenital band or fibrous cord, superficially draining sinuses, polyp. Fistula is a completely patent duct that presents as a persistent umbilical discharge. Cyst develops when ends of the duct are obliterated and cyst remains in the centre. Fibrous cord is a band or septa which is not patent. Sinuses and polyps develop when the duct is patent only near umbilicus.

Clinical symptoms of OMD remnants and other non congenital conditions such as umbilical hernia, appendicitis, intussusception, intestinal torsion and ischemia usually overlap. All of them manifest symptoms such as umbilical discharge, umbilical swelling, recurrent infection in the umbilicus, constipation, bowel obstruction, gastrointestinal bleeding, and rarely intestinal perforation. Patent urachus is another congenital anomaly that occurs when there is a persistent connection between umbilicus and developing bladder. Differentiation between OMD remnants and Patent urachus is essential.

It is difficult to diagnose OMD remnants preoperatively. Radiological investigations such as Ultrasonography, CT, MRI are extremely helpful. Although, OMD remnants remain asymptomatic throughout life but they mostly present in pediatric age group as umbilical discharge. Therefore, in young children presenting with such clinical symptoms it is important to always consider the possibility of OMD remnants as a differential diagnosis.

Heterotopic pancreas is the occurrence of pancreatic tissue other than

its normal anatomical position. It occurs most commonly in upper gastrointestinal tract but it is also found in stomach, duodenum, upper jejunum. It occurs less commonly in lower gastrointestinal tract and even rarely in spleen, lungs, fallopian tubes, omentum.³ It often presents as an incidental finding during autopsy, radiological examination or surgery as it remains mostly asymptomatic.

Multiple theories have tried to explain presence of pancreatic tissue at unusual site. Most widely known is Misplacement theory. It states that pancreatic tissue is accidentally placed at non anatomical sites which later develops into pancreatic tissue. The reason for upper gastrointestinal tract being most common site for heterotopic pancreas is justified by this theory. Another theory is Metaplastic theory which says that during the embryogenesis, endodermal tissue migrates to submucosa which further transforms into pancreatic tissue. According to the totipotent theory, endodermal cells lining the gut or omphalomesenteric duct are totipotent and develop into pancreatic tissue.⁴

Complications arising in heterotopic pancreas are mainly because of location and functionality of pancreatic tissue. Heterotopic pancreatic tissue presents in gastrointestinal tract with non specific symptoms such as epigastric pain, nausea, vomiting, abdominal distention and bowel obstruction. It localises in submucosa layer of GI but it can also be transmurally placed. It is difficult to differentiate it from other submucosal tumors such as Gastrointestinal stromal tumor (GIST) even on endoscopy. It causes submucosal ulceration, gastritis, duodenitis or transmural perforation. The definitive diagnosis is made after histopathological examination.

Abnormally located pancreatic tissue can function as normal pancreas or remain latent depending upon the presence or absence of exocrine and endocrine components⁵. Lesions with functional exocrine component have been observed to have a local chemical irritation in their surrounding. Some cases developed complications such as pancreatitis, pancreatic cysts, pancreatic tumors due to absent proper ductal drainage.

Heinrich in 1909 developed a system which classified heterotopic pancreas on the basis of histology. Type 1 contains acini, ducts, islets. (both exocrine and endocrine component) which were seen in the present case. Type 2 and 3 includes only exocrine component. Both ducts and acini are present in type 2 whereas type 3 contains only ducts. This system was later revised by Fuentes in 1973. He added type 4 which includes only islets (endocrine component)⁵

CONCLUSION

Heterotopic pancreas is a rare finding in omphalomesenteric duct remnants. It is difficult to diagnose preoperatively because the symptoms are not specific. Usually the lesion can persist asymptotically but there can be serious complications if symptoms are ignored for long. It highlights the importance of in-depth understanding for an early diagnosis and effective management.

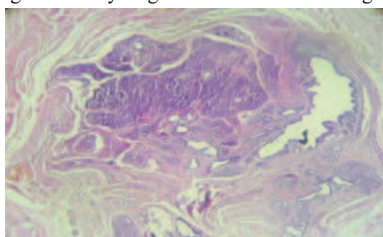


Figure 1 Omphalomesenteric duct remnants revealing pancreatic tissue, intestinal mucosal tissue and connective tissue. (Hematoxylin and Eosin stain;100X)

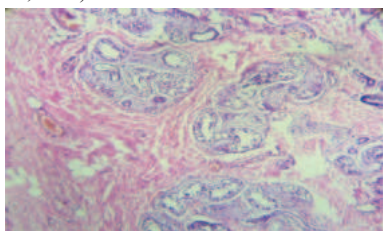


Figure 2 Omphalomesenteric duct remnants revealing intestinal mucosal glands. (Hematoxylin and Eosin stain;200X)

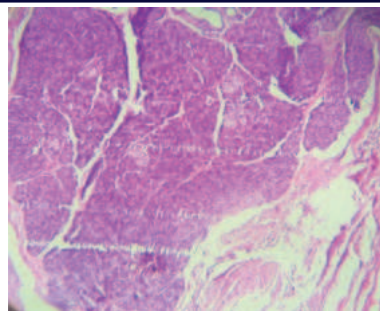


Figure 3 Omphalomesenteric duct remnants revealing Pancreatic tissue-both exocrine and endocrine components. (Hematoxylin and Eosin stain;100X)

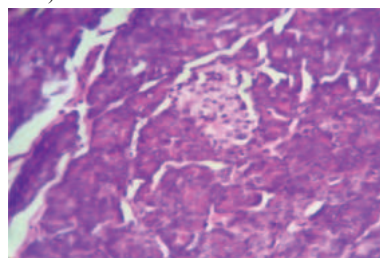


Figure 4 Pancreatic tissue-both exocrine and endocrine components of pancreas. (Hematoxylin and Eosin stain;400X)

Conflicts of Interest: None.

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