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Combination of Urachal and Omphalomesenteric Duct Remnant: A Rare Case Reports

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ABSTRACT

Umbilical problems encountered in the pediatric population are often associated with retained umbilical cord structures or with failure of the umbilical ring to close at birth. This article reported a case of umbilical anomalies in 3-years old male, with a chief complaint of a lump on the umbilical area a urinary discharge from the umbilical since birth. Incidence of patent vitelointestinal duct (omphalomesenteric duct) varies from 1 in 5000-8000 while patent urachus are still rare, ranging from 1-2 per 100000. The combination of both anomalies is very rare, only 12 reported cases since 1898 until 2016. Ultrasound can play an important diagnostic role for these types of anomalies if performed by expert. In our case, the ultrasound was not able to confirm the diagnosis due to a lot of gas in the abdomen. Based on clinical data and histopathologic examination, the patient was diagnosed with combination of urachal and omphalomesenteric duct remnant.

Keywords: histopathology, omphalomesenteric, umbilical, urachal

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INTRODUCTION

Umbilical congenital anomalies are birth defect of umbilical area that occur in children, majority comprised of omphalomesenteric duct and urachal remnant. These anomalies may be present alone or together with umbilical hernia.[1] Omphalomesenteric duct may range from patent omphalomesenteric duct to the more common Meckel's diverticulum, while the urachal remnants can be in the form of patent urachus, urachal cyst, urachal sinus, and bladder diverticulum.[2,3]

The umbilical congenital anomalies in children are rare, about 15% of cases were associated with umbilical cords. The incidence of patent vitelointestinal duct (omphalomesenteric duct) varies from 1 in 5000-8000, while patent urachus are still rare, ranging from 1-2 per 10000 deliveries.[1,2,4] The combination of both congenital anomalies is very rare, and only reported 12 cases since 1898 until 2016. All of the affected newborns were full term.[2,3] Omphalomesenteric duct remnants are often asymptomatic, but may become symptomatic at any age, including abdominal pain, intestinal bleeding, intestinal obstruction, umbilical drainage, and umbilical hernia. Infection and tumors (either benign or malignant) are two main complications of urachal abnormalities.[2]

The diagnosis of umbilical abnormalities can be done by a combination physical examination and radiological examination, such as ultrasound.[2,4] Urachal and omphalomesenteric duct remnant were difficult to diagnosed clinically. The diagnosis of these anomalies are rarely based on ultrasound. CT scan is often the preferred methode for definitive diagnosis. Winni et al affirmed that the use an ultrasound in the diagnosis of urachal remnants lack accuracy for distinguishing true negatives from false positives.[5] Nevertheless, Bertozzi et al mentioned that ultrasound can play an important diagnostic role for these types of anomalies if performed by expert physicians. Indeed, in their case, they have highlighted very well the coexistence of the two anomalies.[2]

Treatment for urachal and omphalomesenteric duct is early surgical management, to prevent complication such as small bowel prolaps, obstruction, intestinal inflammation or bleeding in the patent omphalomesenteric duct and malignant risk in patent urachus. The appearance of an enlarged or "gigantic" cord has particular association with a patent urachus, often requiring operative exploration to repair the associated urachal remnant. Although there have been rare reports of spontaneous regression of a patent omphalomesenteric duct, the vast majority of literature recommend early surgical resection as the appropriate course of management in order to prevent further complications from the malformation.[2,6,7]

We reported a case of combination urachal and omphalomesenteric duct remnant of 3 years old boy by clinicopathology and literature review perspective. This case is quite interesting to discuss because the incidence is very rare and previous literatures have reported the diagnosis can be confirmed by ultrasoud examination if performed by expert. In our case, the ultrasoud was not able to confirm the diagnosis due to a lot of gas in the abdomen. The diagnosis of this case was based on clinical and histopathological examination.

CASE DESCRIPTION

A 3 years old boy, Balinese, came to Sanglah General Hospital Denpasar with a lump in the navel since birth. On physical examination, the patient's was in good general health. Physical examination of abdominal region revealed prominent and reddish umbilicus lump (Figure 1). The clinical diagnosis was persistent urachus.

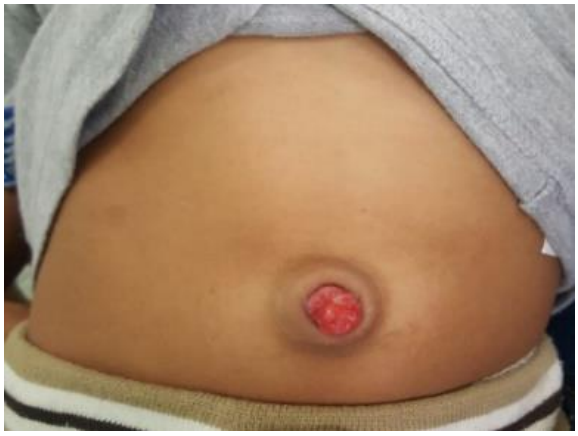

Figure 1. Prominent and reddish umbilical lump appearance.

No abnormalities were found during pregnancy, the maternal body weight was normal, and the delivery time was term pregnancy, but sectio caesarea was performed because of premature rupture of membrane at the time of delivery. The child was born in normal condition and after several days there was urinary discharge from the umbilical. There was no family history that similar with this child. It was difficult to find and evaluate the urachus by ultrasound because of the gas in the intestine.

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Other organs such as liver, gallbladder, spleen, pancreas, renal, bladder, prostat and paraaorta lymph node were within normal limit. Based on the anamnesis, clinical examination and investigation, patient was clinically diagnosed as persistent urachus.

On October 30, 2017 a surgical management with excisional biopsy was performed by removing the entire lesion, to repair the anomalies and to confirmed the diagnosis. The surgeon found the existence of canal associated to the vesica urinaria, without any complication. The excision specimen was brought to Anatomical Pathology laboratory for further examination. Macroscopically, we received one piece of tissue, measuring 3.5 cm long, 1.3 cm in diameter with elongated shape, brownish color and firm consistency (Figure 2A). Microscopic examination revealed an umbilical tissue which was layered by squamous epithelium that continues with the colonic mucosal epithelium, forming a canal structure, and also an adjacent channel structure which was lined by urothelial epithelium, surrounded by fibrocollagen and smooth muscle tissue (Figure 2B-D). We concluded this case as combination of urachal and omphalomesenteric duct remnant. After the surgery management, the postoperative course was favorable and uneventful. At the follow up, the child was fully recovered.

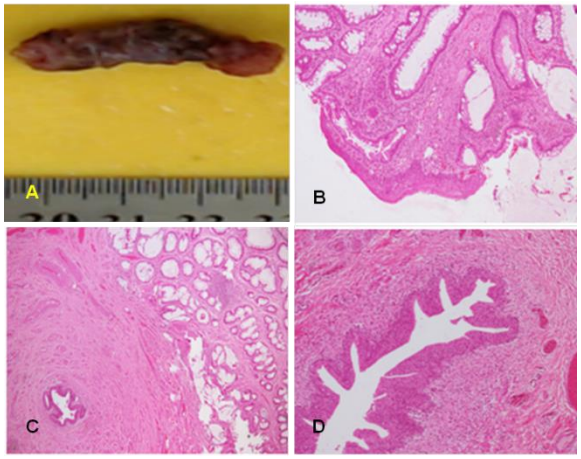

Figure 2. A. Macroscopic, tissue with elongated shape, brownish and firm consistency. B. The stratified squamus epithelial continues with the colonic mucosal epithelium, forming a canal structure (100 times magnification). C & D The canal structure is coated by urothelial epithelium, adjacent to the colonic mucosa surrounded by fibrocollagen and smooth muscle tissue (40 times and 400 times magnification).

DISCUSSION

Umbilical cord in intrauterine life contains paired umbilical arteries, umbilical vain, allantois (urachus) and omphalomesenteric duct or vitelline duct which connects the yolk sac to the midgut. The urachus obliterates by the 5-4th months of gestation to become the median umbilical ligaments.[8] The Incidence of patent vitelointestinal duct (omphalomesenteric duct) varies from 1 in 5000-8000 while patent urachus are still rare, ranging from 1-2 per 10000.[2,4] Pediatric autopsy studies have demonstrated one incidence of 7610 for urachus patents and one in 5000 for urachal cysts.[4]

Urachal remnant is the result of incomplete regression of the intraembrional relationship between allantois and cloaca. Allantois is a finger-like projection of the yolk sac, which is adjacent to the ventral cloaca at one end and the umbilicus on the other.[9,10,11] While the omphalomesenteric duct remnant is the result of persistent or failure of involution of omphalo-mesenteric duct (vitellin duct) by the 5-9th week of gestation.[8]

Umbilical congenital abnormalities are found in children and attributed to the remaining umbilical cord or umbilical ring failure to close at birth.[4] A partial or total failure of the obliteration of the urachus gives rise to various anomalies, which can be discovered in children and adults. Urachal remnants can manifest as patent urachus, urachal cysts, urachal sinus, and bladder diverticulum.[2-4] The low incidence of the urachal anomalies may be because they are not easily encountered in clinical cases as they are frequently asymptomatic.[3] Patent urachal duct remnants can show urinary leakage from a clamped umbilical stump or cyst that may persist into adulthood.[12]

Omphalomesenteric duct remnants are the most common anomalies in the gastrointestinal tract and are often asymptomatic. These anomalies may range from patent omphalomesenteric duct to the more common Meckel's diverticulum. Omphalo-mesenteric duct malformations may become symptomatic at any age, and the common symptoms include abdominal pain, intestinal bleeding, intestinal obstruction, umbilical drainage, and umbilical hernia.[2] Omphalo-mesenteric duct remnants can be symptomatic if direct communication is maintained with the fetal bowel or if ectopic gastric, pancreatic or intestinal mucosa stimulates tissue responses.[12]

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Clinical presentation of umbilical lesions depends on the age of the patient.[8] In adulthood, the onset of signs and symptoms related to urachal disease may occur rarely.[13] Delayed umbilical separation and omphalitis are common in the newborn, while in childhood and among adults, umbilical mass and umbilical discharge or wet umbilicus take precedence.[8] Physical examination of our case revealed prominent and reddish umbilicus lump and urinary discharge from the umbilical, these were consistent with other reported cases.

Infections and tumors are the two main complications of urachal anomalies.[2] Urachal remnants are associated with urachal cyst, sinus, fistula, diverticulum, and infections. In the case of infections, the drainage of the infectious fluids can occur in the bladder, umbilicus, or both. Tumors may be either benign or malignant. Urachal carcinomas are typically silent because of their extraperitoneal location, and, therefore, most of the patients present a diagnosis of local invasion and/or metastasis, which are detected by ultrasound and computed tomography.[2] The malignant tumor that arising from urachal remnants could be an adenocarcinoma of bladder, also urothelial carcinoma, and squamous cell carcinoma.[13] While malignancy can also be a complication of omphalo-mesenteric duct remnant, as an adenocarcinoma of omphalomesenteric duct remnant was reported by Zhou et. al. They believe that this may be the first report of omphalomesenteric duct adenocarcinoma with axillary lymphnode metastasis in adult.[14]

Patent urachus with omphalomesenteric duct is a rare combination to present in the same patient. Bertozzi reported one case with patent urachus associated with a patent omphalo-mesenteric duct in a newborn in 2017, and only 12 cases reported since 1898 until 2016.[2]

Usually, the diagnosis of umbilical abnormalities can be done by a combination of good physical examination and appropriate radiological examination. Any child who presents with a periumbilical mass should have an ultrasound performed, which should be diagnostic for urachal cyst.[10,11,15,16] Patients with periumbilical drainage should perform sonogram examinations to establish a diagnosis such as sinus tract, patent urachus and patent omphalomesenteric duct. Fistulography may be unreliable because of the difficulty of forecasting the channel or the description of a very short channel. Ultrasound is also recommended for early assessment of other urachal or midline abnormalities.[4] It is difficult to diagnosis clinically the urachal and omphalomesenteric duct remnant. The diagnosis of these anomalies is rarely based only on ultrasound. Computed tomography is often the preferred method for a definitive diagnosis. Winni et al affirmed that the use of a ultrasound in the diagnosis for Urachal remnants lacks accuracy for distinguishing true negatives from false positives.[5] Nevertheless, Bertozzi et al mentioned that ultrasound can play an important diagnostic role for these types of anomalies if performed by expert physicians. Indeed, in their case, they have highlighted very well the coexistence of the two anomalies.[2] Ultrasound of the urachal was difficult to evaluate in our case due to a lot of abdominal gas in the abdomen.

Pathological findings related to persistent urachal remnants usually manifest in children and adolescents. Histologically, it is composed of 3 layers; epithelium (stratified, columnar or urothelium), connective tissue and an outer smooth muscle. No goblet cells, no atypia in the epithelium. Usually persist as fragmented tubules separated by fibrous cords, but without a desmoplastic tissue response.[13] The histology of omphalomesenteric duct remnants shows remnant of enteric mucosa which is usually of small intestine or colonic type, but occasionally of gastric type in direct continuity with the adjacent epidermis.[8]

In our case, prominent and reddish umbilical lump was found on physical examination. Persistent urachus was suspected by surgeon. No abnormalities were found during pregnancy, the maternal body weight was normal, and the delivery time was term pregnancy, but sectio caesarea was performed because of premature rupture of membrane at the time of delivery. The child was born in normal condition and after several days there was urinary discharge from the umbilical. There was no family history that similar with this child. Ultrasound of the urachal was difficult to evaluate in our case due to a lot of abdominal gas in the abdomen. Other organs such as liver, gallbladder, spleen, pancreas, renal, bladder, prostat and paraorta lymph node were within normal limit. The existence of canal associated to vesica urinaria was found during surgery. Histopathological examination revealed an umbilical tissue which was layered by squamous epithelium that forming continues structure with colonic mucosal epithelium. At the adjacent tissue of those structure, we also found a channel which is lined by urothelial epithelium,

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surrounded by fibrocollagen and smooth muscle tissue. We concluded this case as a combination of urachal and omphalomesenteric duct remnant. The diagnosis of this case was based on clinical and histopathological examination.

Case with a patent urachus associated with a patent omphalomesenteric duct which diagnosed by histopathological was reported by Lexer et al in 1898. Cullen et al in 1916 diagnosed the patent urachus associated with a patent omphalomesenteric duct intraoperative accidentally. Some authors also reported a patent urachus associated with a patent omphalomesenteric duct incidentally intraoperative.[2]

Treatment for urachal and omphalomesenteric duct is surgery. Early surgical management is needed to prevent complications such as small bowel prolaps, obstruction, intestinal inflammation or bleeding in the patent omphalomesenteric duct and malignant risk in patent urachus. The appearance of an enlarged or "gigantic" cord has particular association with a patent urachus, often requiring operative exploration to repair the associated urachal remnant. Although there have been rare reports of spontaneous regression of a patent omphalomesenteric duct, the vast majority of literature recommend early surgical resection as the appropriate course of management in order to prevent further complications from the malformation.[2,6,7,17]

In our case, a surgical management with excisional biopsy was performed by removing the entire lesion, to repair the anomalies and to confirmed the diagnosis. The surgical resection was done at the age of 3 years old, and follow up was fully recovered.

CONCLUSION

Combinations of urachal and omphalomesenteric duct remnant is very rare. The diagnosis of these anomalies is rarely based only on ultrasound. Computed tomography is often the preferred method for a definitive diagnosis, but ultrasound can play an important diagnostic role for these types of anomalies if performed by expert physicians. The diagnosis of this case was based on clinical and histopathological examination. The ultrasound in this case was not able to confirm the diagnostic due to a lot of gas in the abdomen. Early surgical management is important to prevent the complication, so patient can have a good outcome.

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