Congenital Duodenal Obstruction with Preduodenal Portal Vein and Situs Inversus Totalis: Report of Two Cases and Literature Review

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Abstract Preduodenal portal vein is a rare congenital anomaly that causes high intestinal obstruction. The authors report two interesting cases of preduodenal portal vein that were diagnosed as having congenital duodenal obstruction. As there is a high potential for surgical risk, this anomaly is of special interest to the surgeon and should be kept in mind. Here, we discuss the clinical presentation, diagnosis and management along with a review of the literature.

Keywords Preduodenal portal vein · Duodenal obstruction · Neonate · Situs inversus totalis · Duodenal membrane · Malrotation

Introduction

PREDUODENAL portal vein (PDPV) is one of the rare causes of duodenal obstruction found in only 4% cases [1]. PDPV

was not exactly the cause of duodenal obstruction in most of these; the obstruction was caused by associated malrotation or duodenal atresia as in one of our cases. Duodenal obstruction caused primarily by PDPV, as encountered by us, is extremely rare in the literature [2]. Here, we report the cases of two neonates, along with their management; one neonate presented with duodenal obstruction caused primarily by PDPV and the other with duodenal web with non-obstructive PDPV.

Case 1

A 16-day-old female neonate, weighing 2.2 kg, presented with bilious vomiting, dehydration, lethargy and poor sucking. On examination, we found pneumonitis of the upper zone in both lungs and dextrocardia. On putting in a nasogastric tube, about 30 ml of bile was aspirated. The routine blood investigations were within normal range. A Babygram showed liver shadow on the left and a large gastric bubble on the right with dextrocardia (Fig. 1). Ultrasonography (USG) abdomen showed situs inversus totalis. Echocardiography findings were of dextrocardia with small patent foramen ovale. On exploratory laparotomy, we found an obstructing PDPV with malrotation and situs inversus totalis (Fig. 2). Correction of malrotation with side-to-side duodenojejunostomy was done. The patient had an uneventful recovery.

Case 2

A 12-day-old premature male baby weighing 1.2 kg presented with persistent bilious vomiting, upper abdominal distension and failure to pass meconium. At admission, the patient was dehydrated with tachycardia, feeble peripheral

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Fig. 1 Babygram showing duodenal obstruction, dextrocardia and situs inversus

pulses and hypothermia. There was minimal upper abdominal distension. Apex beat was shifted towards the right, but the trachea was placed centrally and air entry was equal bilaterally. Clinical diagnosis of duodenal obstruction (duodenal web or malrotation) with associated dextrocardia was made. Nasogastric aspirates were bilious. Blood investigations were within normal range. USG abdomen showed situs inversus totalis. Echocardiography findings were of dextrocardia in situs inversus without any other abnormality. On exploratory laparotomy, we found a non-obstructing PDPV with dilated first and second parts of the duodenum, malrotation and situs inversus totalis. There was no associated malformation of biliary system or polysplenia. Correction of malrotation was done, but we could not pass a number 6 Fr feeding tube distal to the second part of duodenum, suggesting obstruction. On enterotomy, we found a duodenal membrane

Fig. 2 Figure showing the preduodenal portal vein causing duodenal obstruction

distal to the second part of the duodenum which was excised and duodenoplasty was done. The postoperative period was uneventful.

Discussion

As many as three-fourths of cases of PDPV have some features of faulty situs determination such as polysplenia syndrome or heterotaxy syndrome [2], as in both of our cases. Review of the literature revealed the presence of ‘isolated’ symptomatic or asymptomatic PDPV in only one fourth cases [3].

The development of this complex anomaly takes place between the 5th and 10th embryonal weeks. It is a result of incomplete reduction of the periduodenal collateral duct between the two vitelline veins [4]. PDPV may lead either to complete or partial duodenal obstruction, but it may even go unrecognized in childhood, manifesting later in adult life. It may occasionally be an incidental finding. Only in about ten cases duodenal compression could be confidently described as being due to a PDPV [2], as seen in one of our cases as well.

The characteristic presentation is onset of vomiting within a few hours of birth and intolerance of attempted feedings in case of complete obstruction [1].

Partial obstruction, however, usually results in recurrent episodes of vomiting, failure to thrive and aspiration over a period of months to years, but both of our cases presented early with features of bilious vomiting and failure to thrive. Some cases are not recognized early in childhood and may present with gastric outlet obstruction later in adult life [5].

PDPV may be an incidental finding in an adult at the time of cholecystectomy [6], and therefore in such cases, we should be cautious to prevent any inadvertent division or ligation of this abnormal vein. Waldschmidt [7] proposed that a coincidentally found PDPV should not be corrected. Waldschmidt also suggested that during operative correction of all rotational anomalies, the possibility of a PDPV should be considered [7]. Surgical importance lies in the fact that it can predispose to intra-operative complications including iatrogenic haemorrhage from the abnormal vein, or damage to the biliary tract or the distended duodenum.

Because of the high incidence of associated anomalies, a prompt system review should be carried out to detect other defects including evaluation for cardiac anomalies [1, 8].

A definitive treatment in cases where PDPV is causing obstruction is duodenoduodenostomy anterior to portal vein [1] or side-to-side duodenojejunostomy [9]. In cases where PDPV is not causing obstruction and the obstruction is caused by an intraluminal web (as in one of our cases), excision of web and duodenoplasty is the treatment of choice. When the duodenal membrane is in the second part

of the duodenum, there is a potential danger of excising a duodenal membrane, which may have the common bile duct entering into it; therefore, the option of doing a duodenal bypass should be kept in mind.

It thus seems important to be aware of the possible presence of a PDPV and its potential complications. Also, when there appears to be an external cause of a duodenal obstruction (PDPV, annular pancreas, Ladd's bands) there is often an intrinsic duodenal obstruction.

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