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## Journal of Pediatric Surgery CASE REPORTS

journal homepage: [www.jpascasereports.com](http://www.jpascasereports.com)The first case of pediatric bile duct adenoma<sup>☆</sup>

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## ABSTRACT

Intrahepatic bile duct adenoma (BDA) is a rare benign epithelial liver tumor derived from bile duct cells. We report the first case of pediatric bile duct adenoma in the world. Furthermore, we review the diagnosis, pathology, treatment and prognosis of bile duct adenoma.

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Bile duct adenomas are benign proliferations of intrahepatic bile ducts. These lesions are usually found under the liver capsule, often as an incidental finding during surgery or at autopsy. Pediatric bile duct adenomas in the liver are very rare. Pediatric bile duct adenoma have never thus far been reported. Here we report the first case of pediatric bile duct adenoma in the world. Furthermore, we review the pathology, diagnosis, treatment and prognosis of bile duct adenoma.

## 1. Case presentation

A three-month-old baby presented with progressive jaundice. The computed tomography (CT) scan revealed an abnormal density lesion with  $1.9 \times 2.5$  cm in the left hepatic lobe (Fig. 1A). The laboratory medicine showed alanine aminotransferase (ALT) was elevated to 87 U/L and alpha fetal protein (AFP) increased to 185 ng/ml. Physical examination revealed no palpable abdominal mass. An exploratory laparotomy was performed. A  $2.0 \times 2.5$  cm substantial

tumor with clear boundary in the left hepatic lobe was completely resected (Fig. 1B). Bile duct adenoma was confirmed by pathology diagnosis (Fig. 1C). At follow-up 1 month after resection, jaundice totally disappeared and baby completely recovered.

## 2. Discussion

Intrahepatic bile duct adenoma (BDA) is a rare benign epithelial liver tumor derived from bile duct cells. BDA represents about 1.3% of all primary liver tumors and it is mainly found in adults incidentally on the surface of the liver at laparotomy or autopsy [1]. To date, this case is the first pediatric bile duct adenoma all over the world [2].

### 2.1. Imaging diagnosis

Although various abdominal imaging approaches have been reported, most BDAs show hypervascular characteristics consisting of prolonged enhancement [3–5]. Our case showed the same findings as in these reports, and it was suggested that the delayed or prolonged enhancement on dynamic CT was due to fibrous stroma within the tumor [6].

### 2.2. Pathology

The pathology reports show that in the vast majority of the cases the lesion is benign. Macroscopically, BDA is a solitary,

Abbreviations: AFP, alpha fetal protein; ALT, alanine aminotransferase; BDA, bile duct adenoma; CT, computed tomography.

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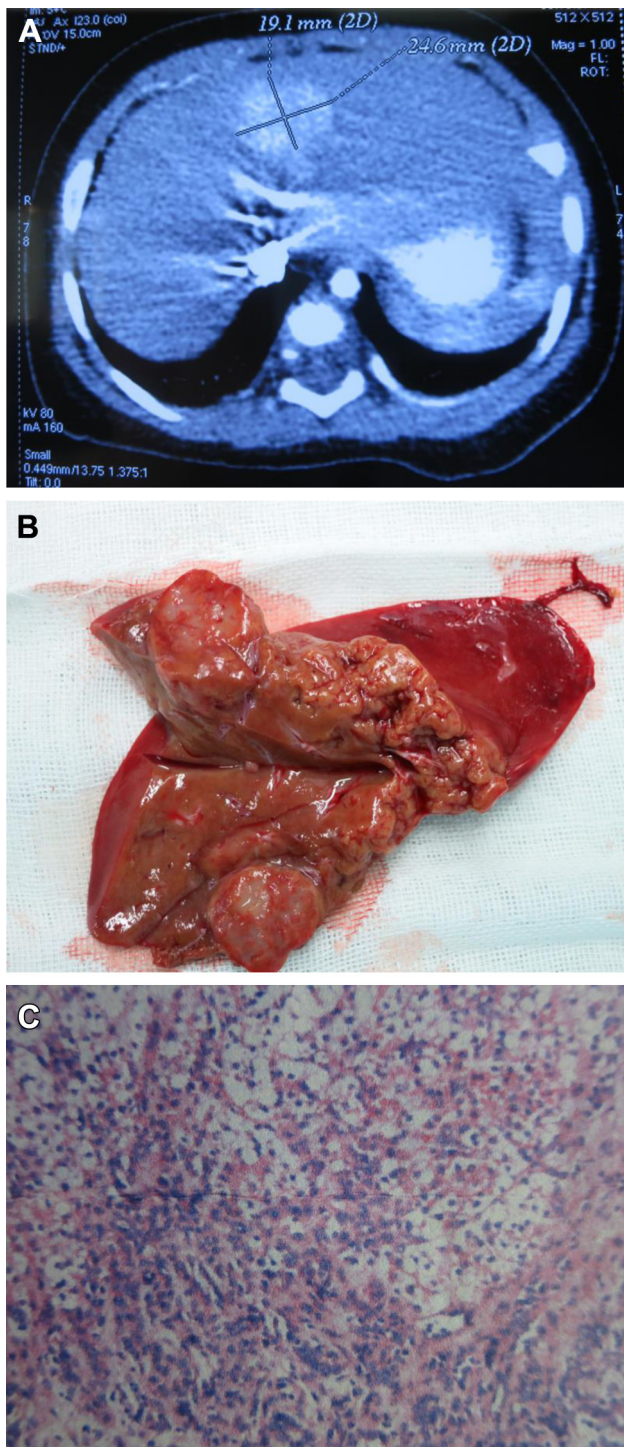


Fig. 1. Bile duct adenoma.

well-circumscribed, firm, gray–white or tan, subcapsular nodule [7]. Microscopically, BDAs are composed of the proliferation of disorganized mature peribiliary gland acini and ductules within a variable amount of connective tissue stroma showing signs of chronic inflammation and collagenization. The cells of the bile ductules have low mitotic activity [1]. Ki-67 positive indicates its potential for malignant transformation [8].

#### The pathological criteria of BDA.

##### Gross description

- Well-circumscribed but unencapsulated, firm, gray–white, tan or yellow, subcapsular round to oval nodules; 85% solitary
- May have central depression
- Usually 5 mm or less but 7% are larger than 1 cm (0.5 cm–1.5 cm)
- Larger than von Meyenburg complex and smaller than cholangiocarcinoma

##### Micro description

- Compact network of simple noncystic tubular ducts or more complex tortuous arrangement, with small or indistinct lumina
- Epithelium has abundant cytoplasm and pale nuclei compared to interlobular bile ducts in adjacent liver
- Rarely clear and oncocytic cells
- Variable fibrous stroma
- Center of larger lesions is paucicellular, densely collagenised; nodular lymphoid aggregate at periphery
- Non caseating granulomas, microcalcification, inflammatory cells; normal portal tracts with bile ducts may be present
- Usually no cystic change, no cytoplasmic or intraluminal bile, no atypia, no mitotic figures, no angiolymphatic invasion

##### Positive stains

- Mucin (intracytoplasmic), CEA, EMA, keratin, PAS highlights basement membrane

##### Negative stains

- CK20, vimentin, HepPar1, chromogranin, Ki-67 (<10% positive)

##### Differential diagnosis

- von Meyenburg complex: 0.5 cm or smaller, less cellular, curvilinear angulated inspissated bile containing ducts, more abundant stroma
- Adenocarcinoma: atypical cytologic and architectural features
- Cholangiocarcinoma: larger, infiltrative

### 2.3. Treatment

Although BDA is defined as a benign tumor, it still has the possibility of carcinogenesis. Hasebe et al. reported a case in which BDA developed into cholangiocarcinoma [9]. It is very difficult to definitely distinguish BDA from hepatocellular carcinoma. Even conducting a frozen section examination, it is still difficult to identify it from cholangiocarcinoma or liver metastasis. Therefore, surgical local resection is required for treatment [10]. Following surgical resection, most of patients have good prognosis.

### 2.4. Prognosis

Given the benign findings on pathology, most of bile duct adenomas have good prognosis.

## 3. Literature review

Intrahepatic BDA is a rare benign epithelial tumor of the liver that is usually incidentally found at surgery or autopsy [1,3,11,12]. Its incidence was reported to make up 1.3% of primary liver tumors [11]. Craig et al. reported only five cases of intrahepatic BDA in 50,000 autopsies, and until now [12]. Allaire et al. have reported the 152 cases between 1943 and 1986, which is the largest series of BDA [1]. Of the 152 cases, most BDAs were asymptomatic nodules discovered incidentally on the surface of the liver during intra-abdominal surgery in 103 cases or at autopsy in 49 cases. They were usually subcapsular, ranging in size from 1 to 20 mm (mean, 5.8 mm), and most of them were less than 1 cm in diameter. The majority of BDAs occurred in individuals between the ages of 20

and 70 years with a mean age of 55 years and no significant difference in sex distribution [1].

#### 4. Summary

To our knowledge, this is thus far the first report of Pediatric Bile Duct Adenoma and we review the diagnosis, pathology, treatment and prognosis of Bile Duct Adenoma (BDA).

#### Conflict of interest

All authors have no conflicts of interest to disclose.

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