

Cystic Diseases of the Organs

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Abstract: The cystic diseases in various organs were observed in the anatomy and OBG department, in that the polycystic disease of ovary was very common compared to all other organs cystic diseases.

I. Aim:

Enumerating the cystic conditions of various organs we came across in the last five years.

II. Materials And Methods:

During a period of 5 years, in the Department of Anatomy and in the Department of OBG, Polycystic conditions of the organs were observed

Observations:

Polycystic conditions of various organs that were observed in the Department of Anatomy and OBG are:

- a. Polycystic Lung-----1
- b. Polycystic kidney-----2
- c. Polycystic Ovary-----34
- d. Polycystic Liver-----1

- a. **The left lung** specimen we came across in a male cadaver showed the following findings
 - The left lung was affected with miliary tuberculosis with involvement of mediastinal lymph nodes
 - Left lung showed the presence of Emphysematus bullae and multiple cysts (Fig-1)
 - Multiple cysts were seen in the interior of the left lung.(Fig-2)

Congenital cysts represent an abnormal detachment of a fragment of primitive foregut, and most consist of **bronchogenic cysts**. Bronchogenic cyst may occur anywhere in the lungs as single or, on occasion, multiple cystic spaces from microscopic size to more than 5 cm in diameter. They are usually found adjacent to bronchi or bronchioles but may or may not have demonstrable connections with the airways. They are lined by bronchial-type epithelium and are usually filled with mucinous secretions or with air. Complications include infection of the secretions, with suppuration, lung abscess, or rupture into bronchi, causing hemorrhage and hemoptysis, or rupture into the pleural cavity, with pneumothorax or interstitial emphysema (Ramzi S. Cotran et al 2001).

- b. There are number of inherited ,Developmental and acquired cystic diseases of the kidney the gross and histologic appearance of some of these conditions may be diagnostic(Bisscegila et al 2006)

The polycystic condition of the kidney can be explained embryologically. Polycystic Kidney may be associated with similar condition of lung or ovary. The Polycystic ovary has a genetic basis Autosomal dominant polycystic kidney disease is the most prevalent, potentially lethal, monogenic disorder. It is associated with large interfamilial and intrafamilial variability (Vicente E Torres et al 2007) It is an autosomal dominant polycystic kidney disease these manifestations include cysts in other organs, such as the liver, seminal vesicles, pancreas, and arachnoid membrane, and other abnormalities, such as intracranial aneurysms and dolichoectasias, aortic root dilatation and aneurysms, mitral valve prolapse, and abdominal wall hernias. We observed bilateral polycystic kidney in a male cadaver (Fig-3) and a family of polycystic kidney.

The classification of cystic disease of kidney by Osathanondh and Potter (1964)

Type 1 – Caused by saccular enlargement of interstitial portion of collecting ducts – familial – death in new born period.

Type 2 – Collecting tubules inadequately branched and terminated in cysts – not familial.

Type 3 – Cysts anywhere along the nephron. Commonly called adult polycystic kidney (Baert (1978)).

Type 4 – Urethral obstruction caused by increased intratubular pressure, – most died at birth.

The adult type affected successive generation being inherited as a Mendelian dominant

c. Multiple cysts were observed in single or both the ovaries (Fig-4)

Polycystic ovaries can be associated with normal or abnormal ovarian function. Patients with polycystic ovarian syndrome have polycystic ovaries with menstrual abnormalities, including amenorrhea in some cases, infertility, hirsutism and obesity (Guzick D) (Franks S). Stein and Leventhal were the first to call attention to this syndrome in the 1930's. It accounts for 1-3% of cases of female infertility. The underlying pathophysiology is complex and not completely understood, but it includes variable combinations of disordered steroidogenesis, alterations in hypothalamic-pituitary-ovarian relationships and insulin resistance (Legro RS); (Balen A., Michelmore K); (Slowey MJ); (Homburg R).

- The infantile type affected only siblings and was thought to be inherited as a recessive. As per the Slowey (2001) polycystic ovary syndrome is the most common endocrinopathy in women of reproductive age - resulting from the insulin resistance and the compensatory hyperinsulinemia. This results in adverse effects on multiple organ systems and may result in alteration in serum lipids, an ovulation, abnormal uterine bleeding and infertility.
- It was associated with sedentary life. Most common age group was 18 to 21 years Metformin was the drug of choice to make ovulatory cycles

b. Polycystic liver disease is the most common extrarenal manifestation. It is associated with both PKD1 and non-PKD1 genotypes. The disease also occurs as a genetically distinct disease in the absence of renal cysts. Like autosomal dominant polycystic kidney disease, autosomal dominant polycystic liver disease is genetically heterogeneous, with two genes identified (PRKCSH and SEC63), which account for around a third of isolated cases. (Li A, Davila S, Furu L, et al.2003; Drenth JP2003; Davila S 2004) Liver cysts arise by excessive proliferation and dilatation of biliary ductules and peribiliary glands Hepatic cysts are rare in children. Their frequency increases with age and might have been under estimated by ultrasound and CT studies. Their prevalence by MRI in the CRISP study was 58% in 15–24 year olds, 85% in 25–34 year olds, and 94% in 35–46 year old participants.

Polycystic liver disease is the most common extrarenal manifestation of ADPKD. The severity of polycystic liver disease usually parallels that of polycystic kidney disease, but exceptions are common.

Hepatic cysts are rare in children. The frequency of hepatic cysts increases with age and may have been underestimated by ultrasound and CT studies. Their prevalence by MRI in the CRISP study in 58% in 15-24 year old participants, 85% in 25-34 year olds, and 94% in 35-46 year olds (Bae et al., 2006). Polycystic liver disease develops at a younger age in women than men and is more severe in women pregnancies. Women who have multiple pregnancies or who have used oral contraceptive drugs or oestrogen replacement therapy have worse disease, suggesting an oestrogen effect on hepatic-cyst growth. (Gabow P, et al, 1990; Sherstha R et al 1997) After menopause, the size of the liver cysts increases in those women who receive estrogen replacement therapy, suggesting that estrogens have an important effect on the progression of polycystic liver disease (Everson & Taylor 2005).

Liver cysts are usually asymptomatic and never cause liver failure. Symptoms, when they occur, are caused by the mass effect of the cysts, the development of complications, or rare associations. Mass effect Include: abdomoinal distension and pain, early satiety, dyspnea, and low back pain. Liver cysts can also cause extrinsic compression of the inferior venacava (IVC), hepatic veins, or bile ducts (Torres 2007).

Typically, polycystic liver disease is asymptomatic; but symptoms have become more common as the lifespan of patients with polycystic kidney disease has lengthened with dialysis and transplantation Symptomatic cyst complications include cyst haemorrhage, infection, and occasional torsion or rupture. The present case was a lady who came with dyspnoea and pain in the abdomen, Ultrasonogram revealed multiple cysts in the liver (Fig-5) and the patient never reported again.

Todani's classification of five types of choledochal cysts is summarized as follows:

- I. **Solitary fusiform extrahepatic cyst**, Single cystic dilation of the common bile duct (80-90% of cases)
- II. **Extrahepatic supraduodenal diverticulum**, Double gallbladder, with one element sessile without cystic duct. Epithelial lining is that of normal gallbladder, according to Vohman and Brown (personal communication from Vohman and Brown to J.E. Skandalakis, (1987) (3%)
- III. **Intraduodenal diverticulum/cholechocele** - Cystic biliary dilation within the duodenal wall (5%)
- IV. Any combination of multiple cysts, i.e., types I, II, III (10%).
- IV. A **Fusiform extra and intrahepatic cysts** - Combination of types I and II.
- V. B **Multiple extrahepatic cysts** - Combination of type I with multiple intrahepatic cysts.

Caroli's disease / multiple intrahepatic in Polycystic condition of the liver - the more common condition is multiple cysts due to Hydatid cyst. The present case is not due to Hydatid cyst, as we can clearly see the empty cystic spaces without water-lily appearance. The present case probably belongs to type V of Todani's

classification. Caroli's disease is treated by partial hepatic lobectomy when localised. Conservative treatment is advised when diffuse cystic disease is present

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Fig-1-Left lung showing the presence of Emphysematus bullae and multiple cysts



Fig-2- Multiple cysts were seen in the interior of the left lung



olycystic kidney disease 1 (PKD1) is the major locus of the common genetic disorder autosomal dominant polycystic kidney

Fig-3-Bilateral Polycystic Kidney



Fig-4- Multiple cysts were observed in single or both the ovaries in ultrasonogram

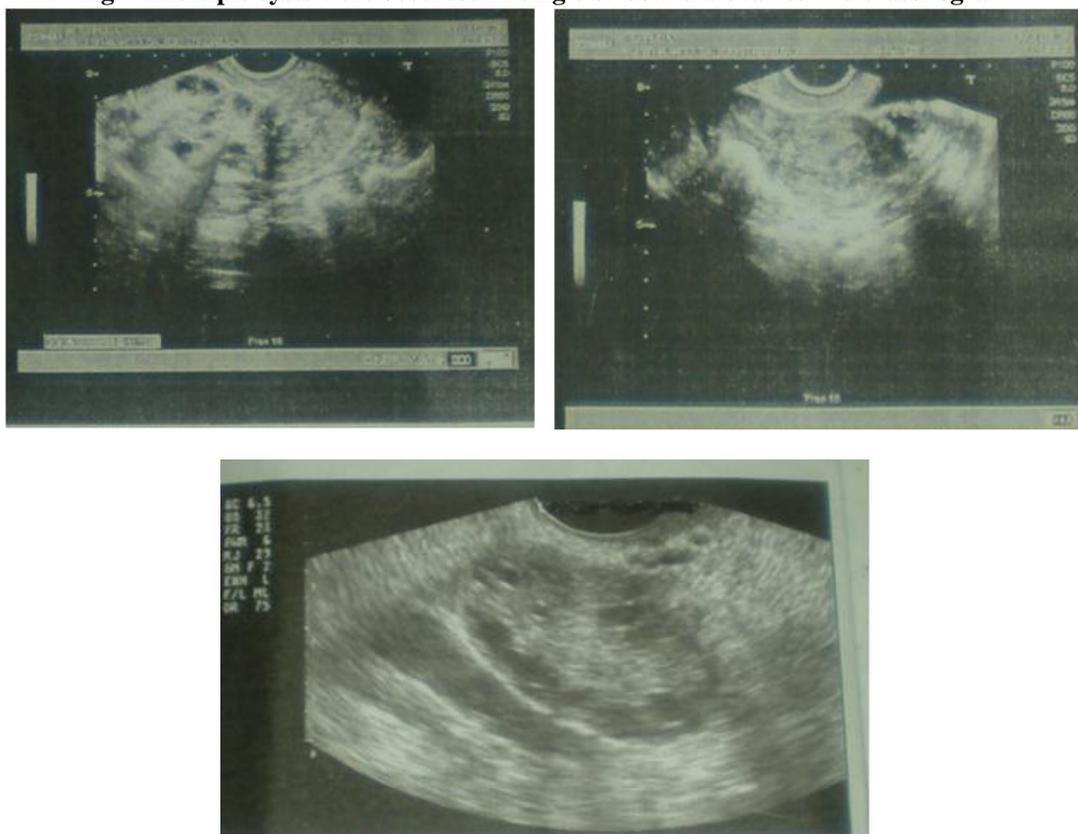


Fig-5-Ultrasonogram of a middle aged woman showing multiple cysts in the liver

