DEXTROCARDIA

Relationship between Characteristics of Genetic Study and Kartagener Syndrome of Elderly People

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ABSTRACT

Objective: Total situs inversus is called Kartagener syndrome.

Design: A clinical study.

Materials and Methods: We experienced an 83-year-old female and a 95-year-old male with total situs inversus. Chest x-ray revealed dextrocardia. Abdominal ultrasonography showed situs inversus totalis. Abdominal CT showed left-sided liver, right-sided stomach.

Results: Situs inversus is a rare congenital disease.

Conclusion: Situs inversus is a rare congenital disease and often has several complicated anomalies. Although our cases had no complicated anomalies, and were alive for long term with no major events due to dextrocardia.

KEY WORDS

dextrocardia, elderly people, Kartagener syndrome, situs inversus totalis

INTRODUCTION

Situs inversus (Kartagener syndrome) is a rare congenital disease and often has several complicated anomalies. Complicated anomalies are right-sided heart, two-lobe lung, multiple spleens, IVC defect, VSD, CBA, and intestinal malrotation. More than several hundred cases of Kartagener syndrome (situs inversus, bronchiectasis and sinusitis) have been reported since Kartagener's original report. Although congenital genetical factors were emphasized in the development of the disease, no clear explanation on its pathogenesis has been made. The incidence of both diseases may involve a common mechanism. Here, we experienced a case of dextrocardia and situs inversus totalis. And she was said that she should be cut the other side when she became appendicitis. At about 50 year-old, she was pointed anomaly by examination of upper gastrointestinal tract. In our hospital she was pointed dextrocardia and situs inversus totalis by chest x-ray, abdominal ultrasonography and abdominal CT.

Physical examination: Her face color was good. She has lumber pain due to aged change of spinal bone deformity. Her consciousness was clear and we could communicate well with her. Her body temperature was 36.2°C, pulse rate 92/min, and blood pressure 158/76 mmHg. Laboratory data: White blood cell count (WBC) 8300 /μl, red blood cell count (RBC) 36.2 × 10⁵ /μl, and hemoglobin 12.2 g/dl.

RESULTS

Study population: We experienced an 83-year-old female and a 95-year-old male with total situs inversus. Chest x-ray revealed dextrocardia. Abdominal ultrasonography showed situs inversus totalis. Abdominal CT showed left-sided liver, right-sided stomach.

The present study conformed to the provisions of the declaration of Helsinki in 1995 (as revised in Edinburgh in 2000).

MATERIALS AND METHODS

Case1: Age and sex: 83-year-old female. Chief complaint: Lumber pain, Vertigo and Constipation. Family history: Four children, (one son, three daughters). Past history: Nothing in particular. Present history: In 1991, she consulted our hospital for lumbago. She had hypertension, constipation and vertigo. She used right hand for writing and cutting. At about 40 year-old, she consulted at Hiroshima university hospital and was pointed situs inversus totalis. And she was said that she should be cut the other side when she became appendicitis. At about 50 year-old, she was pointed anomaly by examination of upper gastrointestinal tract. In our hospital she was pointed dextrocardia and situs inversus totalis by chest x-ray, abdominal ultrasonography and abdominal CT.

Laboratory data: White blood cell count (WBC) 8300 /μl, red blood cell count (RBC) 36.2 × 10⁵ /μl, and hemoglobin 12.2 g/dl.

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count (RBC) 382X104 /μl, hemoglobin (Hb) 11.6 g/dl, Ht 36.9%, Plt 27.7 X104 /μl, Alkaline Phosphatase 175 IU/l, total cholesterol 219 mg/dl, glucose 104 mg/dl and BUN 16.8 mg/dl. Uric acid 6.0 mg/dl and creatinine 1.0 mg/dl were noted. Chest X-ray: Chest X-ray film showed dextrocardia and gastric gas in the right upper abdomen. Abdominal CT: Left-sided liver and right-sided stomach are shown. Abdominal CT film showed situs inversus totalis. Abdominal echo: Abdominal ultrasonography showed situs inversus totalis. Cardiac echo: Left ventricle hypertrophy was observed.

Course up to now: In 2003, she constantly consults our hospital for treating her lumbago, up to now she has no big event due to situs inversus totalis.


Course up to now: He was walking by a cane. In 2016, he fell down and broke the bone. He entered a hospital. Nevertheless, he died. Up to then he has no big event due to situs inversus totalis.

DISCUSSION

In Japan, the incidence of dextrocardia was reported one case among 2000-10000 people in Japan12). Gastric cancer associated with Kartagener syndrome was reported the most malignancy associated with Kartagener syndrome in Japan13). 6 cases of common bile duct carcinoma associated with situs inversus were reported in Japanese literatures1,3-7). 6 cases of hepatocellular carcinoma with situs inversus totalis was reported in Japan1-4). One case of the sigmoid colon carcinoma with total situs inversus and intestinal malrotation was reported15). Some cases of esophageal cancer and rectal cancer with Kartagener syndrome were reported1,2,15-18). To our knowledge, there was no relationship between situs inversus anomaly and malignancy1,2,10-14). In some cases there were some difficulties because of associated malformations at operation and diagnosis15). Usually no severe problems occurred during surgery of malignancy with Kartagener syndrome if preoperative examinations were done carefully. A case involved sigmoid colon carcinoma with some complicated anomalies11). No severe problems occurred during surgery25).

There were also reported some benign diseases as gallstone, Richter hernia and appendicitis associated with Kartagener syndrome14). If preoperative examination revealed the associated malformation of anatomy, there was no difficulty for laparoscopic cholecystectomy for gallstone and appendectomy associated with Kartagener syndrome. In 5 cases of laparoscopic cholecystectomy for gallstone, all cases were already pointed out Kartagener syndrome long before surgery. In our case, she still did not expect appendectomy for appendicitis. In our case, the doctor who diagnosed her as Kartagener syndrome advised her to cut the other side compared with normal person when she become appendicitis. As appendicitis, in emergency case, we should pay special attention to this anomaly.

Although dextrocardia itself has no meaning of lesion, situs inversus (Kartagener syndrome) often has several complicated anomalies. Complicated anomalies are right-sided heart, two-lobe lung, abnormal liver form, non-spleen, multiple spleens, IVC defect, VSD, CBA, and intestinal malrotation. Some of these anomalies may affect the outcome of Kartagener syndrome. Anomalies associated with situs inversus totalis were reported less than those associated with situs inversus partialis. Situs inversus totalis was reported more than situs inversus partialis.

Male were reported more than female. Iwamoto et al were able to examine two female siblings with this triad13-14). Their parents were half-cousins and some family members suffer from sinusitis or chronic obstructive lung disease. Genetic and biochemical studies were performed on the patients, their parents and seven other relatives. They should rule out complication of cystic fibrosis in their cases, and both of them had no evidence of pancreatic disorders. However there are many cases of cystic fibrosis showing no pancreatic insufficiency, and on the other hand, chronic sinusitis and nasal polyps are often seen in patient with cystic fibrosis. In addition, several cases were reported suffering from both Kartagener's syndrome and cystic fibrosis. Their results suggested the possibility that the pathogenesis of both diseases may involve a common mechanism. In our case they had no evidence of pancreatic disorders. Immotile cilia syndrome is sometimes a cause of non-pregnancy associate with female side.

The causes of Kartagener syndrome is researched. The cause of Kartagener syndrom is thought partially genetic. Original Kartagener syndrome was reported in 1933. Kartagener triad are bronchiectasis, dextrocardia and sinusits. In Japan more than 180 cases were reported. Genetic pattern was thought as autosomal recessive. In our cases, quality of life of them was good. They had no major complicated anomalies associated with Kartagener syndrome. We followed her for 12 years, in this year she had no big events due to Kartagener syndrome. As far as our cases, Kartagener syndrome had the good quality of life.

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REFERENCES