[原著]Paterson-Kelly Syndrome associated with Gastric Cancer : A Case Report

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Paterson-Kelly Syndrome associated with Gastric Cancer
------A Case Report ------

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ABSTRACT: A case of Paterson-Kelly syndrome was presented. The 39 year old Japanese female had esoghageal weds, stomatitis and mild hypochromic anemia. She had also gastric carcinoma in the cardia. Total gastrectomy was performed for gastric cancer and the esophageal weds were destroyed by bougienage.

Introduction

Dysphagia localized to the post-cricoid region occurring mainly in middle aged women was first described by Paterson and Kelly in 1919.[1] Cardinal symptoms of the syndrome are dysphagia, anemia and stomatitis, that is also known as Plummer-Vinson syndrome.[2][8][9] But this syndrome is rare in the East. Only 46 cases have been reported in Japan during past forty years. This paper describes a case of Paterson-Kelly syndrome which is associated with gastric carcinoma.

Case Report

A 39 year old female was first seen in our out patient clinic on February 2 in 1977 with chief complaint of discomfort in the epigastrium for two years. She had back pain after a meal for twelve months which had increased gradually. She had lost four kilograms in body weight during past twelve months. She also stated that it had taken her relatively long time to take a meal and sometimes she had felt that something had struck in her throat since childhood. She had barium swallow examination in the other clinic and was referred to us with diagnosis of gastric carcinoma.

Physical examination revealed a well developed and well nourished female patient. Her conjunctiva were neither anemic nor icteric. The tongue was smooth and angular fissures of the mouth were noted. Virchow's node was not palpable. The chest and heart were normal. The abdomen was flat and soft. The liver, spleen and kidneys were not felt. No tumor was palpable in the abdomen.

Barium swallow examination which was done in our hospital disclosed two webs in the esophagus just below the pharyngoesophageal junction and the contrast medium showed jet stream through the weds. In the fundus of the stomach, there was a crater at the posterior wall and its border was not clear. Borrmann Ⅲ type carcinoma was highly suspected. Endoscopy was not successful because of mucosal bleeding of the upper esophagus.

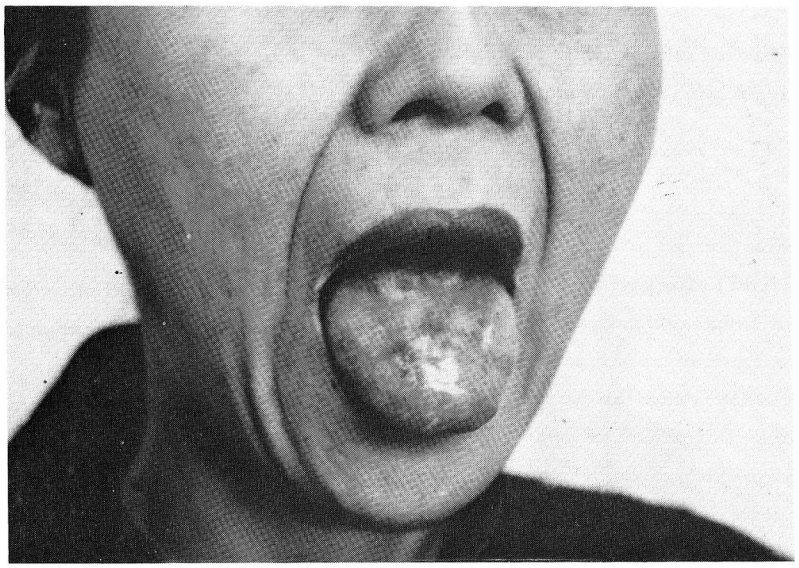

Fig. 1 Angular stomatitis and smooth tongue.

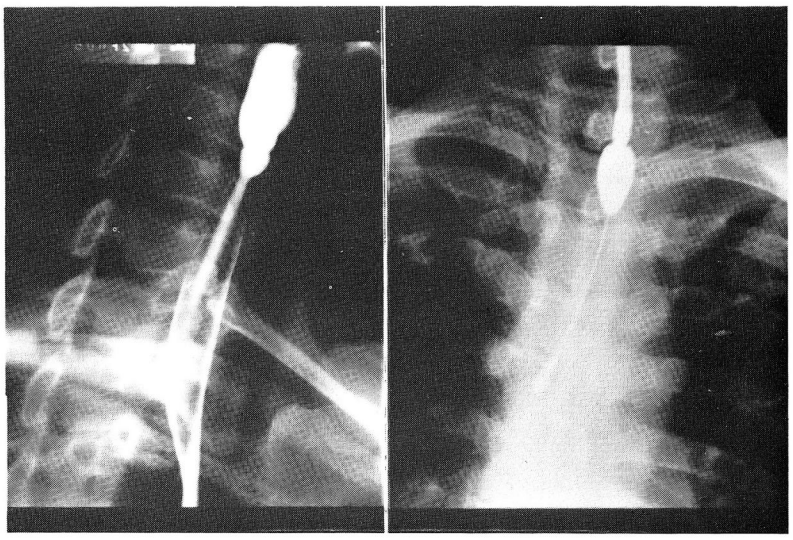

Fig. 2 Left: two webs and jet stream of cotrast medium at the pharyngo-esophageal junction.
Right: bougienage of the webs.

Hematological data were as follows; the red-blood-cell count was 384×10^4. The hematocrit was 35.5 per cent. The hemoglobin concentration was 11.5 g/dl. The white-blood-cell count was 7900 and the platelet count was 213,000. The mean corpuscular hemoglobin concentration (M.C.H.C.) was 33.6 per cent. Serum iron concentration was measured twice that was 104 μg/dl and 118 μg/dl and the serum Vitamin B12 concentration was 554.3 pg/dl (normal range : 300-960 pg/dl). The liver functions and urinalysis were all within normal limits.

On February 18 in 1977, proximal gastrectomy with lymphnode dissection was performed. According to the classification of Japanese Research Society for Gastric Cancer, it was SIN2POHO; Stage Ⅲ. The tumor was 4×5 cm in size. Pathologically it was adenocarcinoma tubulare scirrhosum. In the mucosa of distal surgical margin of the specimen, cancer cells were found microscopically. On April 13 of the same year, total gastrectomy was carried out through left thoracoabdominal approach and reconstruction was made with ρ shaped esophagojejunostomy.

A month after the second operation, bougienage of the esophageal webs was attempted. Under fluoroscopy, a guide wire was inserted into the esophagus and dilators were passed. The dilators No.26, No.29, No.31, No.34 were passed with force respectively. After the bougienage she felt better than before although narrowing of the esophagus was still observed.

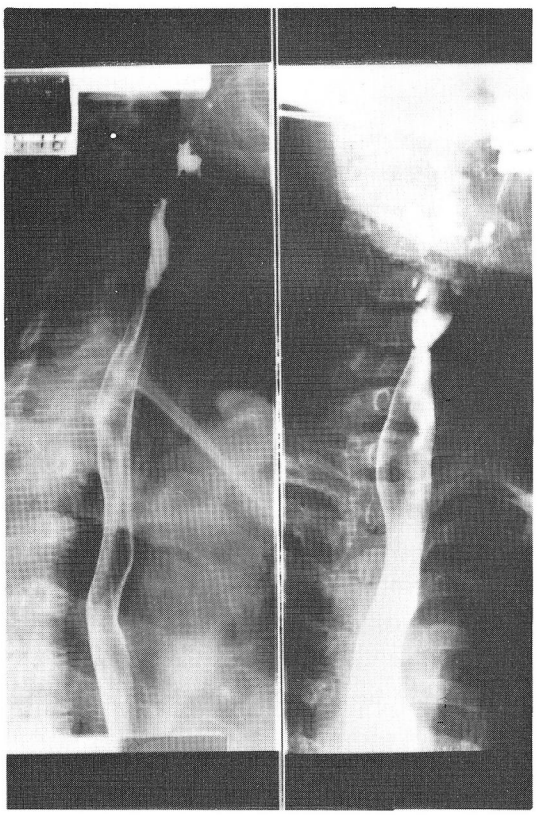

Fig. 3 Left: before treatment. Right: after treatment.

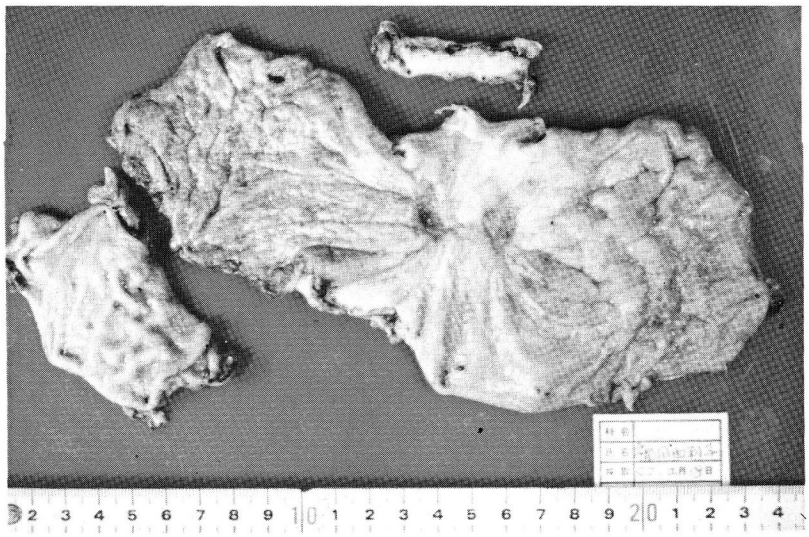

Fig. 4 Specimen of the stomach showing Borrmann Ⅲ type of gastric carcinoma.

Three years have passed since gastrectomy was performed but there is no evident recurrence of gastric carcinoma.

Discussion

The syndrome of dysphagia, hypochromic anemia and stomatitis is rather known as Plummer-Vinson syndrome. They emphasized the pallor and atrophy of the buccal and esophageal mucosa, spooning of the nails, fissures at the corners of the mouth and smooth tongue.[2][3] But the original description was made by Paterson[4] and Kelly[5] in 1919. Kelly[5] noted an association with anemia, superficial glossitis and angular stomatitis while Paterson[4] remarked on 'the not infrequent supervention of malignant disease at the mouth of the gullet.' 'Sideropenic dysphagia' was also named to this syndrome by Waldenström and Hallen[6] in 1938. They showed that the syndrome could occur with iron deficiency unaccompanied by anemia.

The syndrome is not uncommon among Northern Europeans but it is rare in the East and Central Africa. Only 46 cases have been reported during past forty years in our country.[8][9] It occurs mainly in middle aged women. In Wynder's[7] series, 134 out of 150 cases were female. Jacobs reported 53 cases of female and two cases of male.

Our case was incidentally found when she had barium swallow examination for gastric evaluation. Two webs were found just below the pharyngoesophageal junction. Her hematological data showed mild hypochromic anemia with hemoglobin concentration of 11.5 g/dl. The serum iron and Vitamin B12 concentration were within normal range. In Jacobs' series, women with a hemoglobin concentration of 12 g/dl or less were regarded as anemic. The 29 cases out of 53 were anemic by this criterion. Serum iron concen-

tration was abnormally low (less than 80 μg/dl) in 36 cases out of 54. Serum Vitamin B12 concentration was normal in 42 cases except in three. From these facts Jacobs[2] and others[2] objected to the view that iron deficiency is the sole etiological factor in this condition. And he also stated that the syndrome appeared to be especially common among Northern Europeans and it seemed probable that environmental or constitutional factors played a part in determining the effects of iron deficiency.

Wynder[7] noted high incidence of cancer of the upper alimentary tract, hypopharynx and esophagus among Swedish women. He stated that there was a close relationship between Plummer-Vinson(Paterson-Kelly) disease and cancer of these sites. Malignant change in the epithelium may eventually be found in 10 to 30 per cent of cases.[10]

Among 55 cases of Jacobs' series, there were tow cases of gastric carcinoma and a case of carcinoma at the lower end of the esophagus. Shamm'a[2] reported also a case of gastric carcinoma with esophageal webs. Some causal relations might be present between them. Because Jacobs noted achlorhydria in the 37 cases out of 48 and atrophic gastritis or gastric atrophy in these patients has been pointed out by several investigators.

The treatment consists of esophagoscopy with divulsion of weds, repeated bougienage, iron therapy or a combination of these three.[2][8] Our case was treated by bougienage only and the result of which was fairly good.

This case was presented at the 13th Congress of the Japanese Society of Gastroenterological Surgery (Kumamoto, Feb. 1979)

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