A Case of Plummer-Vinson Syndrome

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A case of Plummer-Vinson syndrome is reported. The patient was a 43-year-old woman, with the chief complaint of dysphagia, manifesting glossitis and other oral symptoms, and had hypochromic anemia. Administration of chalybeate was followed by early disappearance of dysphagia and glossitis, as well as the improvement of anemia. It was emphasized that this syndrome develops frequently in women in middle age, that is, the age in which the incidence of upper esophageal cancer is high. In view of this, it is important in the diagnosis of this syndrome to eliminate the possibility of malignancy, which makes mandatory the scrupulous examination of the subpharyngo-esophageal region by means of fluoroscopy and endoscopy. The routine roentgen and endoscopic examinations having been performed in this country are not reliable in detecting the pathology. The effectiveness of chalybeate suggested an intimate relation between iron deficiency and the development of dysphagia, angular stomatitis and glossitis.

Key Words: Dysphagia, Glossitis, Hypochromic anemia, Esophageal web.

Plummer-Vinson syndrome (PVS) is a disease characterized by dysphagia, anemia and glossitis. Patients with this disease frequently visit oto-rhino-laryngologist with chief complaint of dysphagia, and it is rather infrequent to be seen by internists. As to the cause of dysphagia, much remains in obscurity, and it is highly controversial. We have recently encountered a patient with the chief complaint of dysphagia, accompanied by anemia, both of which were improved by the administration of a chalybeate.

CASE REPORT

A 43-year-old female visited the Out-

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patient Clinic, the 2nd Department of Medicine, Toho University Hospital, on November 16, 1971, because of pharyngalgia, sensation of pharyngeal constriction and dysphagia of about 2 months' duration. She also had anorexia and weightloss. She was treated for chronic gastritis about two years prior to this visit. Family history was irrelevant.

The patient was moderately developed and well nourished middle-aged woman, with rather pale face but normal vital signs. Her blood pressure was 120/80. Palpebral conjunctivae were anemic and angular stomatitis were present on both sides of her lips. The surface of her tongue was smooth with papillary atrophy, and the pharynx was diffusely reddened. No tonsillar hypertrophy was noted. Spoon nails were present on the fingers of both hands. The area of cardiac dullness extended 2 cm late-

rally from the left mid-clavicular line. Functional apical murmur was audible. There was no abnormal finding in the lung fields, the liver and spleen were not palpable. No pitting edema in the legs. Tendon reflex was normal and there was no other neurological abnormality.

Laboratory findings are as shown on Table 1. Erythrocyte sedimentation rate was normal, and no abnormality in urine and feces was seen. Peripheral blood showed hypochromic microcytic anemia. Plasma iron level was as low as 52 $\mu\text{g}/\text{dl}$. Gastric juice was hypochloric but response to histalog was normal. Routine roentgen examination of the esophagus and stomach showed nothing abnormal. Gastric endoscopy showed no abnormality in the stomach, although biopsy disclosed chronic gastritis accompanied by a mild intestinal metaplasia. Oral cholecystography revealed a gallstone.

Table 1. Laboratory data.

B.S.R. 1 hr. 6mm	Stool	B(-) G(-)		
2 hr. 15mm		eggs(-)		
Peripheral blood				
Hb 6.4 g/dl	Hemogram			
Ht 24 %	St	0%		
RBC 345 x 10 ⁴	Seg	52		
WBC 4,600	E	0		
Serum Fe 52 ug/dl	B	0		
	M	3		
	Ly	45		
Gastric analysis (histalog)				
	min.	HCl	T-acid	Vol.
basal	15	-29.5	39.5	15
	30	-19.0	59.0	3
	45	-13.0	70.0	7
	60			12
after histalog	15	54.5	110.0	4
	30	71.5	151.0	10
	45	68.5	199.5	7
	60	62.5	162.5	6

Suspecting PVS from these symptoms, the esophagus was reexamined fluoroscopically and as shown in Fig. 1, a typical septum projecting from the anterior wall of the upper part of the esophagus was visualized. It was then confirmed by an esophagoscopy, and the mucosa around the septum was friable and bled easily (Fig. 2). Biopsy revealed a normal squamous cell epithelium (Fig. 3), without any inflammation and

Fig. 1. Roentgenographic demonstration of the upper esophageal web.

Fig. 2. The esophageal web seen by an esophagoscope. The web is seen to project from the ventral (below) wall of the upper esophagus.

malignant changes.

On the basis of these findings, the diagnosis of PVS was made. The patient was treated by oral administration of ferrous fumarate in a daily dose of 300 mg (Table

Fig. 3. The mucous membrane of the esophageal wall around the web appeared intact.

Table 2. The clinical course.

	XI/79	XII/3	I/12	III/8	III/22	IV/19
Hb g/dl	6.4	6.2	6.3	11.0	11.2	12.0
Ht %	24	23	24	32	35	35
RBC $\times 10^4$	345	350	361	412	421	432
M.C.H. μ	18.6	17.7	17.5	26.7	26.6	27.8
M.C.V. μ^3	69.6	65.7	66.5	77.7	83.1	81.0
M.C.H.C. %	26.7	27.0	26.3	34.4	32.0	34.3
Serum Fe μ g/dl	52		14	46		55
Sore throat	+	+	+	-	-	-
Dysphagia	+	+	+	-	-	-
Ang. Stomatitis	+	+	-	-	-	-
Therapy	Ferrous fumar. 300mg/day					

2). Anemia was improved 3 weeks after the treatment was started. In association with the improvement of anemia, pharyngalgia and dysphagia also disappeared. After 4 weeks of the treatment, smooth tongue was also improved. However, serum iron level still remained at 55 μ g/dl on the 5th week. The patient was put on a further observation, but she never returned to the clinic thereafter.

DISCUSSION

In Japan, a case of PVS was first reported by Toyota¹⁾ and his co-workers in 1941. Since then, no more than 30 cases have been described. According to Komaki²⁾, only one out of about 20,000 cases in ABCC in Hiroshima was diagnosed as having this disease, and it is therefore considered to be a relatively rare disease in this country.

The characteristic esophageal lesion can be found only by a careful fluoroscopy or endoscopy. Routine roentgen study or endoscopy very often fail to demonstrate

the septum which is an important clue for the diagnosis of the disease. When no abnormality is found by fluoroscopy, PVS is often mistaken for esophageal or laryngeal neurosis, and this may be one of the reasons why there are only a few reports on this syndrome.

As for the etiology, Plummer and Vinson ascribed it to a spasm of functional disorder at the beginning part of the esophagus. Thereafter, divergent theories have been published, one considering the anemia as the primary cause (Ono³⁾, Muta et al⁴⁾, a second advancing the racial theory (Muta⁴⁾) and a third the precarcinoma theory (Suzman⁵⁾, Johnson⁶⁾, and Ahlbon⁷⁾). McGee et al⁸⁾ observed in autopsy, a mucous swelling and ablation to a muscular thickening in the upper half of the esophagus, and Suzman et al⁵⁾ recognized epithelial ablation and carnification, and regressive degeneration of muscular layer in the tongue and subpharyngo-esophageal region, all these organic changes being attributable to the etiology of the dysphagia. Besides these lesions, achalasia was considered by Hurst et al⁹⁾, who stated that inflammation and regressive degeneration of the Auerbach's plexus, accompanying the glossopharyngitis, might be the cause of the dysphagia. According to them, disorder in stimulus transmission, resulting from atrophy of the Auerbach's plexus, may induce insufficient relaxation of the sphincter at this site.

In the early stage, spasm may be evoked by abnormal excitation of the neuromuscular system. Müller et al¹⁰⁾ attributed the dysphagia to drying of mucosa and spasm of the cricopharyngeal muscle, both of which would result from the lowered tension of the autonomic nerve. Hiroto et al¹¹⁾, however, thought it inappropriate to regard PVS as a single monolithic entity. It should rather be considered, according to them, to consist of two entities, that is, functional dysphagia which was designated by them as dysphagia hysterica, and organic dysphagia. Davidson¹²⁾, McGibbon¹³⁾, and Hoover¹⁴⁾ observed the presence of web-shaped or band-shaped damages in the

esophagus, while Ono³⁾ found membrane-like elevation in the beginning part of the esophagus. Kernan¹⁵⁾ estimated that the fibrous web-shaped or band-formed changes in the esophagus might have been formed secondarily in the healing process of ulcer in that site.

It is known moreover that the incidence of PVS is high among middleaged females (about 30 to 50 years of age), that is the age in which the upper esophageal cancer develops at high frequency. It is therefore necessary to eliminate the possibility of malignant tumor. Johnson⁶⁾ regarded PVS as a precarcinomatous condition, because it corresponds well with regard to its distribution, to postcricoid cancer, which develops at the rate of 90% in woman. Ahlbom⁷⁾ described that 70% of 150 cancer patients were preceded by this disease. Watanabe et al¹⁶⁾ and Tobita et al¹⁷⁾ reported cases of PVS which were complicated by candidiasis and Recklinghausen's disease, respectively.

From these facts, it is considered that the etiology of dysphagia of this syndrome may have many stages, beginning from the functional disorder, especially spasm at the upper part of the esophagus and advancing to organic lesions.

The relation between anemia and dysphasia of this syndrome is still controversial. McGibbon¹³⁾ considers that the esophageal changes is a primary lesion, which would impair food intake, thus secondarily inducing anemia. Suzman⁵⁾ assumes that the dysphagia may have merely incidentally been complicated by essential hypochromic anemia. Against these, Witts¹⁸⁾ says that essential hypochromic anemia is followed or accompanied by PVS and gastrointestinal diseases and that the dysphasia is due to glossopharyngitis, resulting from the anemia. Hiroto¹¹⁾ et al do not consider the dysphagia to be attributable to anemia, whereas Muta⁴⁾ and Ono et al³⁾, regard the esophageal constriction, which is generally observed, as anemic constriction, that is, a part of the manifestation of the essential hypochromic anemia. In this way, there

is again divergence of opinions concerning the relation between anemia and dysphagia of this syndrome.

In the present case, it is difficult to decide solely by her complaint, which of the two symptoms preceded the other. It is, however, likely that there may at least be intimate relation between the anemia and dysphagia, because the dysphagia disappeared early after the administration of chalybeate, as had been experienced by other workers, and because in parallel with this, the anemia was likewise improved.

The fact that, as demonstrated in the present case, the dysphagia, glossitis and other symptoms were improved by the treatment for anemia, indicates that anemia may be responsible in one way or another for the development of the above mentioned symptoms and findings.

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