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Plummer-Vinson Syndrome; A Report of Two Cases and Review of Medical Literatures

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Introduction

Patients with long-standing dysphagia are seen rather frequently in Japan. Yet, the cause in most cases is not definitely diagnosed. The so-called Plummer-Vinson syndrome must be included in the differential diagnoses. The esophageal narrowing is difficult to establish unless fluoroscopic examinations are carefully performed with a high index of suspicion.

The Atomic Bomb Casualty Commission (ABCC)-Japanese National Institute of Health (JNIH) Adult Health Study is a long-term follow-up investigation of A-bomb survivors and comparison subjects in a population sample originally consisting of 20,000 individuals. Included in their biennial examinations are routine chest roentgenography and other roentgenological studies as indicated by clinical and laboratory findings. Detailed gastrointestinal examinations are performed only by trained radiologists.

In the ABCC Department of Radiology all roentgenological diagnoses are routinely coded for even-
tual data-retrieval using an electronic computer. These methods assist in establishing prevalence of diseases and abnormalities. Within this population sample only one case of Plummer-Vinson syndrome was roentgenologically proven. One additional case was detected but was not in this sample. Both are described in this report.

CASE I (MF#207530) A 54-year-old Japanese female complained of gradually increasing dysphagia of 19 years duration. This first occurred during ingestion of solids, and was accompanied by glossitis. She had an appendectomy at 24 years of age. Her estimated Hiroshima A-bomb dose was 28 rad. She complained of lumbago of 3 years duration, the cause of which was not established. Her mother died of uterine cancer at 46 years of age. Her maternal grandmother suffered from dysphagia and died of gastric cancer. Her maternal aunt also died of gastric cancer. Eleven years prior to her current illness "spooned" finger nails and anemia were noted on examination elsewhere. Repeated oral hematinics for more than a year had no effect. On her first ABC ad mission 10 years ago she complained of dysphagia and "spooning" of finger nails. There was a moderate hypochromic anemia (Hb, 9.5 g/100 ml; RBC, 531 × 10⁴/mm³), with sideropenia. Barium swallow showed a circular narrowing of the upper part of the esophagus, compatible with Plummer-Vinson syndrome (Figures 1 a, b, c).

![Fig. 1a: Case I, 14 September 1959. PA view of the esophagus just below the pharyngeal junction; with marked circular indentation but smooth margins](image1)

![Fig. 1b: Case I, 14 September 1959. PA view of the esophagus; the portion below the web visualized by air-contrast. Smooth margins are demonstrated. Jet phenomenon is present.](image2)

![Fig. 1c: Case I, 14 September 1959. A left anterior oblique view demonstrates the complete circular indentation well.](image3)

On examination 8 years ago at ABC 7 there was mild epigastric discomfort, but no dysphagia. Slight anemia was noted (Hb, 11.2 g/100 ml; RBD 456 × 10⁴/mm³). She was again examined at ABC 7, 4 and 2 years ago, and each time she had mild dysphagia and glossitis. She never experienced weight loss. Her current physical examination revealed a well developed moderately well nourished 54-year-old female with
facial and oral pallor, and minimal glossitis. There was “spooning” of the finger nails, and general excoriation due to ichthyosis vulgaris. Detailed hematologic data are shown in Table 1. Other laboratory studies were within normal limits. Repeat barium swallow examination again showed the circular narrowing in the upper part of the esophagus without internal change (Figures 2a, b).

Table 1. Hematologic Studies

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<td>400</td>
<td>473</td>
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N = within normal range

Fig. 2a: Case I, 23 June 1969. The PA projection shows the deformity to be unchanged over 10 years.

Fig. 2b: Case I, 23 June 1965. The left anterior oblique projection also shows no internal change. The upper and lower margins of the web are again smooth.
CASE II (MF $W-4284$) This 57-year-old Japanese female complained of dysphagia of 42 years duration. She had anemia at 35 years of age and undiagnosed liver disease in 1962. Her mother died of liver cancer; otherwise her family history was not contributory. At 15 years of age she visited an otolaryngologist because of choking on pickles. Regurgitation occasionally relieved the choking sensation caused by solids. At 35 years of age she was easily fatigued, and was found to have an iron deficiency anemia which responded to one month's oral hematinc therapy. During the last 5 years she experienced periodic nausea. Although she initially had difficulty swallowing solids, she eventually experienced some difficulty with thick liquids. For many years she needed long periods to finish her meals. On 14 October 1969 she first visited ABCC.

Her physical examination was essentially negative. Her hematologic studies were as follows: $\text{RBC}, 420 \times 10^4/\text{mm}; \text{Hb}, 12.8 \text{ g}/100 \text{ ml}; \text{Hematocrit}, 37.5\%$; Serum Iron, 92 $\gamma$/dl, (normal: 72–130 $\gamma$/dl); UIBC 124 $\gamma$/dl, (normal: 144–280 $\gamma$/dl); and TIBC, 226 $\gamma$/dl (normal: 216–410 $\gamma$/dl).

Barium swallow revealed marked circular narrowing of the upper portion of the esophagus, compatible with Plummer-Vinson syndrome (Figures 3a, b, c).

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**Fig. 3a:** Case II, 12 December 1969. This PA view of the esophagus shows a marked circular indentation just below the pharyngoesophageal junction. The upper and lower margins of the web are smooth.

**Fig. 3b:** Case II, 12 December 1969. This PA projection visualizes the web as a lip-like protrusion into the esophageal lumen.

**Fig. 3c:** Case II, 12 December 1969. This lateral view shows that the web is mainly on the anterior aspect of the esophagus.

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**Discussion**

In 1906, Lindvall quoted Paterson’s reported case of anemia with dysphagia. According to Lindvall, and Bockus, Plummer, Paterson, and Kelly independently described such cases in 1919. Bockus cites Vinson’s description of the characteristic features of this disease; namely, dysphagia in women.
with anemia and splenomegaly. Virson credited Plummer with the first discovery of this disease, but it was eventually called Plummer-Vinson syndrome, according to McGee et al.13)

Some British authors adopted the name Paterson-Kelly syndrome.5) In 1939 Waldenstrom et al.22) who first reported radiological findings, suggested the term "sideropenic dysphagia", since it was not always associated with anemia and achlorhydria but low serum iron level.

Endoscopic findings were first reported by Hoover8) in 1935. According to Fujimori et al.,6) the first Japanese case was described in 1941.

Clinical Picture:

Dysphagia and anemia are the prominent features, but Bockus3) described splenomegaly in addition. Epithelial atrophy, particularly of mucus membrane, such as in the tongue and oral cavity, have also been described,123)17)20)28) as well as angular stomatitis.3) Achlorhydria was reported associated with the epithelial changes.13)22) The anemia is of long duration,5)20) and of the hypochromic microcytic iron deficiency type,7)10)22) "Spooning" of the nails,22)28) has been reported. It and splenomegaly are regarded manifestations of the anemia. Bockus3) says practically all patients have low serum iron concentrations. Waldenstrom et al.22) described three cases without anemia. Administration of large doses of iron has reversed the mucosal changes and relieved the dysphagia except in very far advanced cases.5)22) It is now generally accepted that iron deficiency with or without anemia precedes the development of dysphagia, contrary to earlier suppositions that the dysphagia caused malnutrition and anemia.

Dysphagia:

Disturbance of the pharyngeal and esophageal neuromuscular mechanism was earlier regarded the cause of the dysphagia.5)14) However, a combination of functional and organic changes is also mentioned.14) That is, the constriction could be due to spasm, accentuated by the formation of one or more small fibrous webs. It is now generally accepted that the web or constriction is the main cause of the dysphagia.20)12)17)20)28)22) Shamma's18) denies the functional theory entirely. However, congenital as well as sideropenic webs can be present.14)22) According to Waldmann22) web-origins are of four types: Congenital, developmental, post-inflammatory, and post-abrasive.

The web is located in the pharyngo-esophageal area,3)14)19)22) particularly behind the cricoid cartilage on the anterior aspect of the esophagus4)15) and the anterior wall of the lower border of the hypopharynx.17)24) But webs in the middle and lower portions of the esophagus have also been reported.18)

Nearly all cases of Plummer-Vinson syndrome have webs.12)17)22) Thomas9) found webs in about half

<table>
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<tr>
<th>Reporters</th>
<th>Total Cases</th>
<th>Male</th>
<th>Female</th>
<th>Anemia with Dysphagia</th>
<th>Anemia</th>
<th>Web</th>
<th>Dysphagia</th>
<th>Cancer</th>
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<td>58</td>
<td>7</td>
<td>51</td>
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<td>58/58</td>
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<td>MacMillan17)</td>
<td>114</td>
<td>16%</td>
<td>84%</td>
<td>5/53</td>
<td>1/4</td>
<td>1/4</td>
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<td></td>
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<td>11/2</td>
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<td>4</td>
<td>3</td>
<td>1</td>
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of the 29 cases in his series. As to frequency of anemia in web cases 5 cases of anemia and dysphagia in 53 cases of webs were reported by Seaman; and 23 cases of anemia and dysphagia in 37 cases of webs, by Shamma'a.

The incidence of the Plummer-Vinson syndrome among "control" groups has been reported as 1 in 145, by Waldenstrom et al.; none in 104 cases with postcricoid dysphagia, by Pitman et al.; none in 71 cases without symptoms, also by Pitman et al.; 23 with anemia and web in 58 cases with dysphagia, by Shamma'a and 114 cases of web in 1600 cases with dysphagia, by MacMillan.

Webs may be single or multiple. Multiple ones are said to be congenital; single ones, acquired.

Three multiple types were found among 53 cases of webs.

Roentgerologic Appearance of Webs:

Webs arise from the anterior wall of the esophagus and extend posteriorly along the lateral walls. Unusual circular webs have also been described. Therefore, lateral roentgenograms usually show them in the upper portion of the esophagus, beneath the cricoid cartilage, as exceedingly thin, lip-like transverse folds with smooth contours. Except in advanced and severe cases, the frontal projection is not usually useful. A mouthful of swallowed barium is needed to distend the hypopharynx and esophagus.

Kingham advocated the use of a barium suspension of creamy consistency in the PA projection, with the patient's head turned to the left or right. Use of barium-filled capsules were described by Thomas.

Radiologically, the web sometimes cannot be easily distinguished from the indentation on the anterior wall of the esophagus in the post-cricoid region. Pitman described 104 cases among 121 patients with post-cricoid dysphagia. In a "control" group, 64 of 71 patients had this finding.

Endoscopic Appearance of Webs:

When the esophagoscopy is inserted, the lumen of the esophagus appears reduced by a thin membranous wet or band of raised mucus projecting into the lumen from the anterior esophageal wall or surrounding lumen in the post-cricoid region. The web usually presents a smooth, thin, gray appearance and contains very small arterioles and venules. It is usually friable and easily ruptured by the tip of the esophagoscope. Esophagoscopy is therefore sometimes used both for therapy as well as diagnosis.

Cancer as a Complication:

The most important consideration of the Plummer-Vinson syndrome is that the web is precancerous. Many reports of cancer secondary to this entity have been published, such as 8 cases in 53 cases of webs, 9 cases in 58 cases of webs; 2 cases in 16 of Plummer-Vinson syndrome, and 1 case in 4 cases of Plummer-Vinson syndrome.

The cancer usually occurs in the pharynx and in the upper part of the esophagus proximal to the web. Lindvall described 28 cases proximal to the web in his 34 cases of tumor secondary to Plummer-Vinson syndrome. According to Lindvall, Ahalbom described 60% of patients with cancer of the mouth, larynx and esophagus who received radiation therapy and had a history of anemia and dysphagia; as to post-cricoid and upper esophageal cancer, 90% of patients had a history of Plummer-Vinson syndrome. Shamma'a emphasized that multiple webs are precancerous, and found 6 cases of cancer among 8 patients with multiple webs.

Age and Sex of Patients:
This syndrome occurs predominantly among middle-aged women,\(^{12}^{13}^{14}^{15}^{16}^{17}\) from 30–50 years of age. According to Shama\(^{18}\), 51 of 58 cases of Plummer-Vinson syndrome were women. Seaman\(^{19}\) reported 11 women among 16 patients with webs and anemia. A minority of males have this syndrome.\(^{20}\) Among 14 cases of webs found at Massachusetts General Hospital, 84% were women.\(^{13}\)

Prognosis and Therapy:

Since this is a sideropenic dysphagia, iron therapy is generally used. Symptoms usually subside within a few days of the initiation of iron therapy, and within 1 month the patient becomes entirely free of symptoms. In cases of marked narrowing, however, only partial relief has been obtained.\(^{22}\) Even without anemia, it is usually said that administration of large quantities of iron relieve the dysphagia,\(^{6,22}\) Bingham\(^{23}\) on the contrary was of the belief that no quantity of iron will correct the dysphagia, and that the inability to eat solids such as meat is an absolute indication for dilation procedures. Esophagoscopy is therefore useful in treatment as well as diagnosis.\(^{12,18,19}\) Repeated bougienage\(^{7,18}\) Vitamin A,\(^{14}\) B\(^{13,14}\) and liver extract,\(^{8}\) are regarded beneficial.

**Conclusion**

According to Kitaura et al.,\(^{10}\) 30 cases of Plummer-Vinson syndrome were reported in Japan as of 1964. In the ABCC-JNIH Adult Health Study, only one case of this abnormality was radiologically established among those examined because of symptomatology or clinical findings. The infrequency of Plummer-Vinson syndrome among Adult Health Study participants parallels the few case reports in Japan.

Usually only clinically suspected cases of Plummer-Vinson syndrome with typical findings, such as iron-deficiency anemia, dysphagia and glossitis are radiologically examined. Radiologically, lesions may be overlooked if the web is small enough not to be demonstrated and if only PA projections are used. Lateral projections are essential. Sometimes, only the middle and lower portions of the esophagus may be well-visualized because of rapid passage of barium through the pharynx and upper portion of the esophagus and also because some examiners may not anticipate lesions as frequently in the upper portion of the esophagus. Treatment must be taken to assure that these sites are well-demonstrated.

Our first case had dysphagia for 19 years and an iron deficiency anemia. She had a marked circular type web which has not changed during 10 years' observation. Two members of her family had stomach cancer; one had uterine cancer; and her maternal grandmother had stomach cancer, preceded by long standing dysphagia. It has been pointed out that Plummer-Vinson syndrome is a precancerous disorder. The family history in this case suggests that there may be a familial occurrence, though this has not been reported in the literature.

Our second case had typical symptomatology with dysphagia of more than 40 years duration, and a long history of iron-deficiency anemia. She did not seek medical assistance because she felt the dysphagia might be of exaggerated importance. She also has a marked circular upper esophageal web.

**Summary**

Two cases of Plummer-Vinson syndrome with markedly-constricting upper esophageal webs of long duration, both on oral hematonic therapy, are reported. These cases demonstrated the unusual constricting of the circular type. They stress the importance of detailed examination of the pharynx and upper portion of the esophagus during upper gastrointestinal examinations. Only one of these two cases of Plum-
ner-Vinson syndrome was a member of the population sample originally consisting of 20,000 participants in long-term ABCF follow-up examinations. A review of the literature on this subject is included.

References