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Determining the frequency of Plummer Vinson syndrome among patients having dysphagia presenting at district Bahawalpur

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ABSTRACT

BACKGROUND & OBJECTIVE: Plummer Vinson syndrome (PVS) is a rare triad of dysphagia, iron deficiency, and esophageal webs. This study aims to assess the frequency of PVS in patients presenting with dysphagia.
METHODOLOGY: A cross-sectional study was conducted at the Department of Otolaryngology from 1st April 2020 to 31st March 2021. A total of 164 patients having dysphagia for a minimum of two weeks duration were enrolled. The patient's data was recorded using a pre-designed questionnaire for PVS assessments, complete blood count, serum ferritin levels and barium swallow test results were obtained, and a radiologist was consulted for the upper esophageal web.
RESULTS: The observed frequency of PVS among patients with dysphagia was 11(6.7%). Female gender turned out to be significantly associated with PVS occurrence (p=0.001), while age (p=0.748) and duration of dysphagia (p=1.00) showed no significant association with it. Among those with PVS, 72.7% of patients were >40 years of age, 90.9% were females, and 81.8% had a duration of dysphagia >4 weeks.
CONCLUSION: It is concluded that PVS is not very frequent among patients with dysphagia. But since it increases the risk of squamous cell carcinoma of the pharynx and the esophagus, therefore, such patients should be followed closely for the presence of PVS and treated promptly to avoid morbidity and mortality.
KEYWORDS: Plummer Vinson Syndrome, Dysphagia, Iron Deficiencies, Anemia.

INTRODUCTION

Dysphagia is usually intermittent, progressive and remains painless for a long duration [1,2]. Initially, hysterical dysphagia due to cardiospasm or upper esophageal spasm without anatomic stenosis was described as Plummer Vinson Syndrome (PVS)[3-6]. PVS is characterized by the triad, including cervical dysphagia, iron-deficiency anemia (IDA), and post cricoid esophageal web. The occurrence is rare and presents as difficult swallowing with decrease hemoglobin levels and/or narrowing of the upper esophagus [7,8]. The PVS incidence is more frequently observed among females and adults aged 40 to 70 years [9,10].

Although the exact pathogenesis of PVS is still unknown, among the major investigation are alterations in the complete blood cell (CBC) counts, peripheral blood smears, iron deficiency indicated by serum iron, total iron-binding capacity (TIBC), ferritin, and saturation percentage, repetition in the inflammation and healing process, disturbed motility of the pharynx or esophagus and one or more than one esophageal webs (revealed on video fluoroscopy) [2,4]. The patients with PVS commonly complain regarding weakness, paleness, fatigue, losing weight and tachycardia. Clinical findings reveal smooth tongue, cracks of the oral commissure as well as Koilonychia [11]. The affected individuals show greater susceptibility towards upper esophageal malignancy [12].

Esophagoscopy and medical management have found to be helpful for the treatment of PVS [13,14]. Furthermore, iron supplementation can help resolving sidropenic dysphagia (iron deficiency) among PVS patients [15,16]. As PVS can reoccur, follow-up is essential; investigations like complete

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blood count and serum ferritin levels need to be monitored at 6 monthly intervals. The PVS patients having dysphagia may need repeated dilatations for management [17,18].

This study was designed to evaluate the frequency of PVS among patients with dysphagia in District Bahawalpur. Moreover, this study will help in early diagnosis and management of the disease in order to minimize the chances of malignant progression.

METHODOLOGY

This cross-sectional study was conducted at the Department of Otolaryngology, Bahawal Victoria Hospital, Bahawalpur, from 1st April 2020 to 31st March 2021. Adopting a non-probability consecutive sampling method, a total of 164 male and female patients with dysphagia (for a minimum of 2 weeks duration) and more than 15 years of age were enrolled after obtaining written informed consent. While, patients with neurogenic dysphagia, malignancy of the oropharynx, hypopharynx, and esophagus were excluded from the study sample. The study was ethically approved by the ethical review committee of Quaid-e-Azam Medical College (Reference no. 280/DME/QAMC Bahawalpur; Date 2nd March 2020).

A predesigned proforma was used to record the study data, including patient age, gender and duration of dysphagia. All the enrolled patients were admitted to ENT Unit for PVS assessments, and clinical examination was carried out. Complete blood count, serum ferritin levels and barium swallow test results were obtained, and a radiologist was consulted for the upper esophageal web. In PVS-diagnosed cases, iron therapy was started immediately.

Data was statistically analyzed on SPSS version 20.0, the quantitative variables like age and duration of dysphagia were represented as mean and standard deviation. While frequency and percentages were used to present categorical variables like age group, gender and frequency of PVS. Post-stratification Fisher's exact test was used to compare study variables, where p-value<0.05 was considered statistically significant.

RESULTS

The mean age of the enrolled patients was noted to be 43.83 ±3.30 years. Out of the total 164 patients, the majority were males (57.3%). Furthermore, the mean duration of dysphagia was 4.62 ± 1.18 weeks; most patients had a prolonged duration of dysphagia, i.e. 125(76.2%) of patients had a duration of more than 4 weeks (Table-I). Only 6.7% patients with dysphagia had Plummer Vinson Syndrome while 93.3% did not.

There was no significant difference in the age (p=0.748) and duration of dysphagia (p=1.00) among the patients with or without PVS. The female gender turned out to be significantly associated with PVS frequency (p=0.001).

Table-I: Participant characteristics (n=164).

Variables	Categories	n(%)
Age (Years)	15-30 years	8(4.9)
	31-40 years	52(31.7)
	>40 years	104(63.4)
Duration of Dysphagia	2–4 weeks	39(23.8)
	>4 weeks	125(76.2)
Gender	Male	94(57.3)
	Female	70(42.7)
Frequency of PVS	Present	11(6.7)
	Absent	153(93.3)

DISCUSSION

Plummer Vinson Syndrome is less frequently reported due to better nutritional outcomes, but it still remains a challenge among developing countries like Pakistan due to nutritional deficiencies and restricted healthcare facilities. Therefore, the present study aimed to assess the frequency of PVS among patients presenting with dysphagia for developing screening and treatment guidelines to reduce disease-associated morbidity and mortality.

The incidence rate and frequency of PVS among patients with dysphagia cannot be particularly described due to scarcity of literature. An old population-based study including 2346 females and 1994 males with dysphagia showed that the overall prevalence of PVS was 0.3 % to 1.1% and 8.4% to 22.4%, respectively [19]. Currently obtained findings suggest that PVS is not very frequent among the patients with dysphagia i.e. 6.7%. Similarly, a local study aimed to identify the etiology of dysphagia among adults reported that 3.7% of the enrolled cases were found to have PVS[20]. In contrast a more recent study displayed high frequency of PVS among women with early grade dysphagia [4].

IDA and dysphagia are the key investigating features in PVS patients [5,9]. We observed that dysphagia was more frequent among adults with a mean age of 43.83 ± 3.30 years, majority of the patients presented at the study site were > 40 years of age (Table-I). Our findings were very similar to a local study from Peshawar, where 42% of the enrolled patients with dysphagia were in between 36 to 45 years of age, and their reported frequency of PVS was 5% [21]. Patil et al., also confirmed the presence of high frequency of PVS middle aged women with dysphagia [4], which is also corroborated by other published studies [5,14].

Furthermore, the association of PVS with age, gender and duration of dysphagia was also assessed. We found no significant association of age and duration of dysphagia with the presence of PVS among the enrolled patients (p>0.05). While only gender had significant association (p<0.05). Among those having PVS, 90.9% were females (Table 2), which is similar to a local study in which 95.2% of the patients having PVS were females [21]. Hence, now it is considered a fact that the female gender with dysphagia is comparatively more prone to PVS than males [5,21-23]. Most patients with PVS (81.8%) had dysphagia for more than 4 weeks (Table-II). Hussain et al. reported that out of 21 PVS positive cases, 76.1% of patients had dysphagia from 4-5 weeks [21].

Table-II: Association of Plummer Vinson Syndrome with age, gender and duration of dysphagia.

Variables	Categories	Pvs (n=11%)	No PVS (n=153%)	p-value
Age	15 - 30 years	1(27.3)	57(37.3)	0.748
	>40 years	8(72.7)	96(62.7)	
Gender	Male	1(9.1)	93(60.8)	0.001
	Female	10(90.9)	60(39.2)	
Duration of Dysphagia	2 – 4 weeks	2(18.2)	37(24.2)	1.00
	> 4 weeks	9(81.8)	116(75.8)	

***p<0.05 is considered significant**

Very little is known relating to PVS locally and the available data is limited to case reports and small cohorts. This cross-sectional study was carried out to provide an insight into the frequency of this rare condition among patients with dysphagia in Bahawalpur. Studies with prospective designs involving multiple sites and larger sample sizes are required further to enhance knowledge about PVS and its risk factors, pattern of presentation, diagnosis and management.

CONCLUSION

The low frequency of Plummer Vinson Syndrome among the enrolled patients with dysphagia suggest the rare disease instance. Furthermore, a significantly high frequency of PVS was observed among the female patients having dysphagia. These patients with dysphagia should be followed closely for the presence of PVS, as they are predisposed to squamous cell carcinoma of the pharynx and the esophagus.

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Author's Contribution:

Nasir Wakeel: Substantial contributions to the conception and design of the work.
Masood Akhtar: Drafting the work and evising it critically for important intellectual content.
Aasma Tariq: Designed & contributed in manuscript writing.
Iqra Gull: Acquisition, analysis, and interpretation of data for the work.
Hamna Hafeez: Data analysis and manuscript writing.
Nayab Anjum: Collected the data and performed the analysis.

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