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Case Report

Plummer Vinson syndrome in children: case series

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ABSTRACT

Plummer Vinson syndrome (PVS) is a rare entity in children and classically presents with a triad of anemia, dysphagia and post cricoid web. We report four paediatric cases of PVS reported in a tertiary care institute in Chennai, India over a period of 6 years. All children were above 5 years of age with M:F of 1:3. Mean duration of symptomatology was 13.5 months. Mean age at the diagnosis was 9.3 years. Classical triad of PVS was seen in all of our children who responded well to endotherapy without any recurrence. Though rare in children case series of PVS are highlighted to stress the importance of evaluation, when children present with dysphagia and anemia and the necessity of early referral to paediatric GI centre for successful management.

Keywords: Endoscopy, Iron deficiency, Post cricoid web

INTRODUCTION

Esophageal web is a thin (2-3 mm), eccentric, smooth extension of normal esophageal tissue consisting of mucosa and submucosa which may be congenital or acquired.[1]

Acquired webs which are more common than congenital webs typically appear in the cervical area (postcricoid) in contrast to congenital webs which occur in middle and inferior third of esophagus.

Plummer Vinson Syndrome (PVS) previously called Kelly-Paterson syndrome is characterized by iron deficiency anemia and post cricoid web.

Around 20 cases have been reported in children worldwide.[2] We report 4 paediatric cases of post cricoid web associated with PVS.

CASE REPORT

Case 1

7 years old female child of non-consanguineous marriage presented with symptoms of dysphagia and easy fatigability of one-year duration. Clinical examination revealed anemia with glossitis, angular stomatitis and koilonychia with a BMI of 12.01 (<3rd percentile as per revised IAP growth charts). Investigations showed very low hemoglobin of 4.1 g/dl with microcytic hypochromic anemia in peripheral smear. Iron studies were suggestive of iron deficiency anemia with low serum iron 13 µg/dl, low serum ferritin 6.6 ng/ml and high TIBC 620 µg/dl). Stool for occult blood was negative on three different occasions. Barium swallow showed narrowing at the cricopharyngeal region as depicted in Figure 1. Upper GI endoscopy showed web at the level of post cricoid region as depicted in Figure 2 which was dilated using savary guillard dilators. Child was started on oral iron therapy

along with deworming and dietary advice. Child is currently under follow up and had required 3 endoscopic sittings.

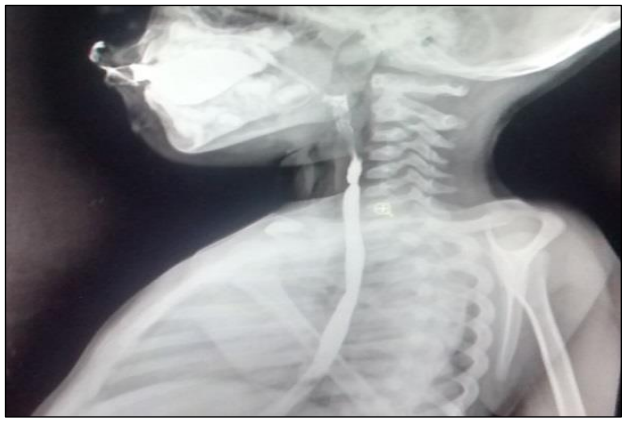

Figure 1: Lateral view barium oesophagogram showing post cricoid web.

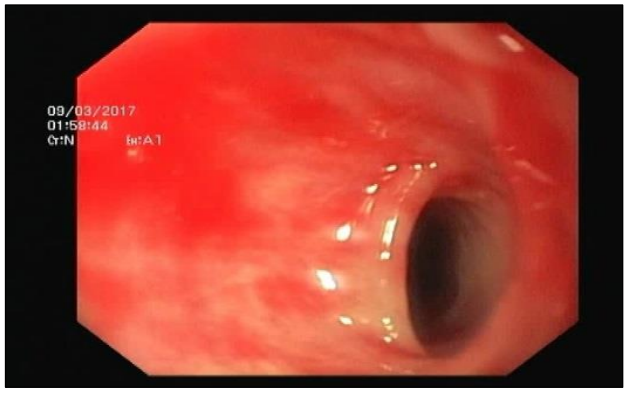

Figure 2: Upper GI endoscopy showing post cricoid web showing postcricoid web

Case 2

9-year-old female child of 2nd degree consanguineous marriage presented with complaints of dysphagia to both solids and liquids for two years duration. Clinically she had anemia, glossitis, angular stomatitis and koilonychia with a BMI of 11.06 (<3rd percentile as per revised IAP growth charts). Her haemoglobin was 8 g/dl and peripheral smear was suggestive of microcytic hypochromic anaemia. Serum iron (12 µg/dl) and serum ferritin (3.7 ng/ml) were low with elevated levels of TIBC (449 µg/dl).

Barium swallow showed an indentation at the post cricoid level suggestive of post cricoid web.

Endoscopy confirmed the web and dilatation was done in the same sitting and child was started on oral iron and albendazole with nutritional advice. She required two sittings of endoscopic dilatation at an interval of four weeks without any recurrence of symptoms on follow up.

Table 1: Clinical and anthropometric profile of patients.

	Case 1	Case 2	Case 3	Case 4
Age	7	9	10	11
Sex	F	F	F	M
Consanguinity	NCM	2nd degree	2nd degree	NCM
Weight (Kg)	12.5	17	18	20
Height (cm)	102	124	122	128
BMI	12.01	11.06	12.09	12.21
Duration of symptoms	1 year	2 years	6 months	1 year

Table 2: Biochemical, radiological and treatment details of patient.

	Case 1	Case 2	Case 3	Case 4
Haemoglobin (g/dl)	4.1	8	6.2	7.4
Serum iron (µg/dl)	13	12	17	15
Serum ferritin (ng/ml)	6.6	3.7	4	7.2
TIBC (µg/dl)	620	449	470	492
Barium swallow	Post cricoid web	Post cricoid web	Normal	Cricopharyngeal web
Number of sittings	3	2	3	1

Case 3

10-year-old female child born preterm to 2nd degree consanguineous marriage with birth weight of 1 Kg presented with complaints of dysphagia to solids, weight loss and easy fatigability of six months duration. Clinically she was thin built (BMI 12.09 <3rd percentile as per revised IAP growth charts) with anaemia, glossitis, angular stomatitis, and koilonychia with hemic murmur. Investigations showed haemoglobin 6.2 g/dl, microcytic hypochromic anaemia. Stool for occult blood was negative on three different occasions. Serum iron profile revealed low serum iron (17 µg/dl), low serum ferritin (4 ng/ml) with high TIBC (470 µg/dl) suggestive of iron deficiency anaemia. Barium swallow was normal but however endoscopy confirmed the diagnosis of post

cricoid web. Child was started on oral iron supplements with albendazole and dietary advice. Child required three endoscopic sittings at an interval of four weeks and now the child is symptom free for the past 5 years.

Case 4

11-year-old male child of non-consanguineous marriage presented with symptoms of dysphagia for 1 year. On clinical examination he was thin built (BMI 12.21 <3rd percentile as per revised IAP growth charts) with pallor, glossitis and angular stomatitis. His hemoglobin was 7.4 g/dl and peripheral smear showed microcytic hypochromic anemia. Iron profile was suggestive of iron deficiency anemia with low serum iron (15µg/dl), low serum ferritin (7.2 ng/ml) and high TIBC (492 µg/dl). Barium swallow was suggestive of cricopharyngeal web and endoscopy findings confirmed the same and was treated by single endoscopic dilatation without any recurrence. He was started on oral iron therapy, albendazole with nutritional advice.

DISCUSSION

PVS is characterized by a classic triad of dysphagia, iron-deficiency anaemia and oesophageal webs, mostly reported in middle-aged women, and less commonly reported in children.[3] M:F ratio in our series 1:3. PVS is named after Henry Stanley Plummer and Porter Paisley Vinson who reported a series of patients with long-standing iron deficiency anaemia, dysphagia and spasm of the upper oesophagus without anatomic stenosis, however Paterson DR and Brown-Kelly A were the first to describe the characteristic clinical features of the syndrome.[4-7] Exact incidence of the disease is unknown and only few case reports are reported in world literature with declining trend in incidence probably due to improvement in standard of living.[8]

Though the exact etiopathogenesis is uncertain, iron deficiency has been recognized as an important cause. Reduction of iron containing enzymes causing mucosal degenerations, atrophic changes and web formation and even leading to cancer development has been proposed as a possible mechanism.[9] Pyridoxine deficiency has also been proposed to cause oesophageal dysmotility in PVS.[10] Decreased contraction amplitude of the oesophageal muscle in iron deficiency leads to prolonged transit times of proximal and median oesophagus.[11] Some of the other factors like malnutrition, genetic predisposition and autoimmune processes has been incriminated since PVS is known to occur with autoimmune disorders like rheumatoid arthritis, thyroiditis, celiac disease, and pernicious anaemia.[12]

The classical triad include intermittent painless dysphagia to solids, features of anaemia and post cricoid web, similarly seen in all our children.[3] The development of anaemia in our children is probably nutritional as all our cases were underweight and thin built.

Diagnosis is made by constellation of clinical, biochemical, radiological, endoscopic finding. Radiological findings of barium swallow showing a postcricoid esophageal web that can be single or multiple is characteristic but however it could be normal, as seen in one of our patient.[13] Webs on endoscopy appear as smooth, thin, gray, with a centric or lateral lumen at the anterior oesophageal wall and should be performed carefully not to miss or accidentally injuring it and biopsy is essential in adults as malignancy is quite common unlike in children.

Management consists of oral iron supplementation, mechanical dilatation, nutritional advice with regular follow up. Though oral iron therapy benefits many, endoscopic stricture dilation is needed in children with persistent dysphagia as seen in our series.[14] Other treatment modalities include argon plasma coagulation and surgical disruption or excision of the web rarely.[15,16] As there is a risk of malignancy, yearly surveillance endoscopy is recommended.[17]

CONCLUSION

Though rare in children case series of PVS are highlighted to stress the importance of evaluation when children present with dysphagia and anaemia and the necessity of early referral to paediatric GI centre for successful management.

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