Original Research Article

Incidence of malignancy in intestinal polyp of Peutz-Jeghers Syndrome: Case Series

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Abstract

Introduction: Peutz-Jeghers syndrome is also known as "Hereditary Intestinal Polyposis Syndrome". It is an autosomal dominant disease, characterized by the development of benign hamartomatous and adenomatous polyps in the gastrointestinal tract, and hyperpigmented macules on the lips, nostrils, buccal mucosa, palmer surfaces of the hands, genitalia and perianal region. Its incidence is 1 in 200000 births. 25 cases of Peutz-Jeghers Syndrome were studied in tertiary health centre. Diagnosis of Peutz-Jeghers Syndrome was made on the basis of clinical details, radiology reports and the histopathological study. The patients were from both urban and rural areas of Ujjain. The entire study was retrospective study.

Aims and Objectives: To study the incidence of malignancy in intestinal polyp of Peutz-Jeghers Syndrome.

Materials and Methods: Polyps from patients with Peutz-Jeghers Syndrome were studied in the histopathology department. The corresponding medical records were reviewed to confirm that the diagnosis of Peutz-Jeghers Syndrome. Hematoxylin and Eosin (H&E) stain was used for the histopathological diagnosis. Particular attention was paid to the presence and pattern of distribution of network of connective tissue, smooth muscle, lamina propria and glands lined by intestinal epithelium.

Results and Conclusion: We evaluated colonic polyps from 25 patients of Peutz-Jeghers Syndrome. Out of all patients, none of the intestinal polyps turned out to be malignant**.** The incidence of malignancy in intestinal polyps in Peutz-Jeghers Syndrome as per this study is insignificant.

Key words

Peutz-Jeghers, Hereditary, Intestinal, Polyposis, Hamartomatous.

Introduction

Peutz-Jeghers syndrome is an Autosomal Dominant condition characterized by benign Hamartomatous polyps in the gastrointestinal tract [1]. Hyperpigmented macules are present on the lips, nostrils, buccal mucosa, palmer surfaces of the hands, genitalia and perianal region. Grossly, the polyps are large pedunculated with a lobulated contour [2]. Peutz-Jeghers syndrome has an incidence of approximately 1 in 2,00,000 births [5]. These patients may also develop extra-intestinal malignancies, which include breast, ovary, lung, pancreas, cervix and testis [2]. One-third of patients are symptomatic by age 10 and half of the patients have experienced intussusception, obstruction and bleeding by age 20 [4]. Histopathologically, Polyps in Peutz-Jeghers syndrome have characteristic arborizing network of connective tissue, smooth muscle, lamina propria and glands lined by normal appearing intestinal epithelium [2].

Aim and objective

- To study the incidence of malignancy in intestinal polyp of Peutz-Jeghers Syndrome.

Materials and methods

25 cases of Peutz-Jeghers Syndrome were studied in tertiary health center. Diagnosis of Peutz-Jeghers Syndrome was made on the basis of clinical details, radiological reports and the histopathological study [3].

Polyps from patients with Peutz-Jeghers Syndrome were studied in the histopathological department of Pathology. Hematoxylin and Eosin (H&E) stain were used for the histopathological diagnosis. Particular attention was paid to the presence and pattern of distribution of muscularis mucosa. It was a retrospective study (**Figure – 1 to 8**).

Results and Discussion

25 patients of Peutz-Jeghers Syndrome were studied. Out of all patients of Peutz-Jeghers syndrome, none of the intestinal polyps turned out to be malignant. The incidence of malignancy in intestinal polyps in Peutz-Jeghers Syndrome as per this study was insignificant (**Table – 1**).

Figure - 1: Dark blue to brown macules on the lips.

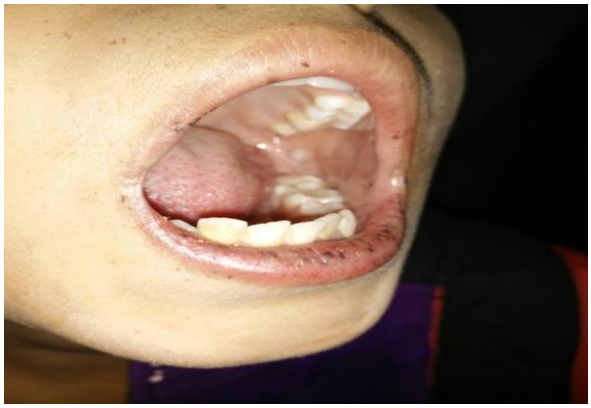


Figure - 2: Dark blue to brown macules on the lips and nostrils.

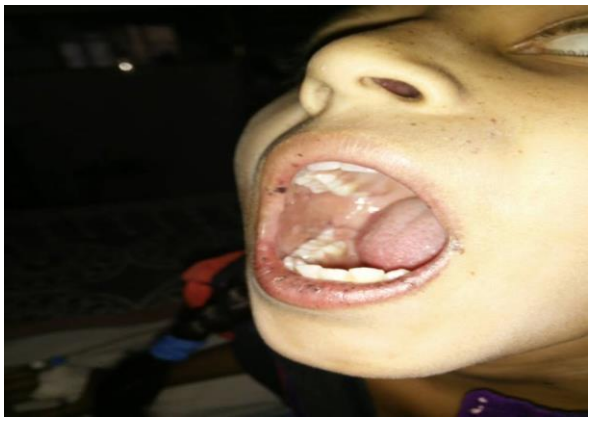
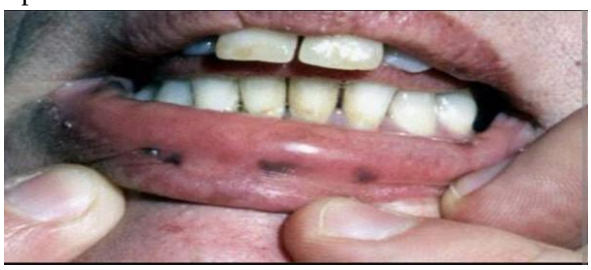


Figure - 3: Dark blue to brown macules on the lips.



For diagnosis of Peutz-Jeghers Syndrome family history plays a significant role, mucocutaneous lesions causing patches of hyperpigmentation in the mouth and on the hands and feet. The oral pigmentations are the first to appear [6]. Intra

orally it is most frequently seen on the gingiva, hard palate and inside of the cheek. The mucosa of the lower lip is almost invariably involved as well [7]. Hamartomatous polyps are benign polyps with an extraordinarily low potential for malignancy. 90-100% of patients with a clinical diagnosis of Peutz-Jeghers syndrome have a mutation in the STK11/LKB1 gene. Having 2 of the 3 listed clinical criteria indicates a positive diagnosis.

- The Histological Hallmark of Peutz-Jeghers polyp is the presence of arborizing pattern of muscularis mucosa.
- Although the frequency of cancer in this syndrome has not been studied extensively but Francis M, et al. shows cancer in 15 out of 31 cases [8].
- Our result is similar to the study done by Tamanna Choudhary, Suraiya Enam, Bangabandhu Medical University, with no malignancy reported [9].

Table - 1: Incidence of malignant polyp.

S. No.	Age	Sex	Location	Family History	Histopathology	Malignancy Detected
1	14	Female	Ileum	Yes	Benign Hamartomatous Polyp	No
2	17	Male	Colon	Yes	Benign Hamartomatous Polyp	No
3	15	Female	Jejunum	Unknown	Benign Hamartomatous Polyp	No
4	21	Male	Ileum	Unknown	Benign Hamartomatous Polyp	No
5	18	Male	Ileum	Yes	Benign Hamartomatous Polyp	No
6	16	Female	Jejunum	Unknown	Benign Hamartomatous Polyp	No
7	20	Female	Ileum	Yes	Benign Hamartomatous Polyp	No
8	14	Female	Ileum	Yes	Benign Hamartomatous Polyp	No
9	16	Male	Colon	Unknown	Benign Hamartomatous Polyp	No
10	22	Male	Ileum	Yes	Benign Hamartomatous Polyp	No
11	18	Male	Jejunum	Yes	Benign Hamartomatous Polyp	No
12	13	Female	Jejunum	Yes	Benign Hamartomatous Polyp	No
13	19	Female	Jejunum	Yes	Benign Hamartomatous Polyp	No
14	21	Female	Ileum	Unknown	Benign Hamartomatous Polyp	No
15	17	Male	Ileum	Yes	Benign Hamartomatous Polyp	No
16	21	Female	Jejunum	Unknown	Benign Hamartomatous Polyp	No
17	18	Male	Jejunum	Unknown	Benign Hamartomatous Polyp	No
18	19	Female	Jejunum	Yes	Benign Hamartomatous Polyp	No
19	17	Male	Ileum	Yes	Benign Hamartomatous Polyp	No
20	20	Male	Jejunum	Yes	Benign Hamartomatous Polyp	No
21	22	Male	Ileum	Yes	Benign Hamartomatous Polyp	No
22	21	Female	Ileum	Yes	Benign Hamartomatous Polyp	No
23	12	Female	Jejunum	Unknown	Benign Hamartomatous Polyp	No
24	18	Male	Jejunum	Unknown	Benign Hamartomatous Polyp	No
25	17	Male	Ileum	Unknown	Benign Hamartomatous Polyp	No

Conclusion

We evaluated colonic polyps from 25 patients of Peutz-Jeghers Syndrome. Out of all patients, none of the intestinal polyps turned out to be malignant. The incidence of malignancy in intestinal polyps in Peutz-Jeghers Syndrome as per this study is insignificant.

Figure - 4: Colonoscopy showing Polyp.

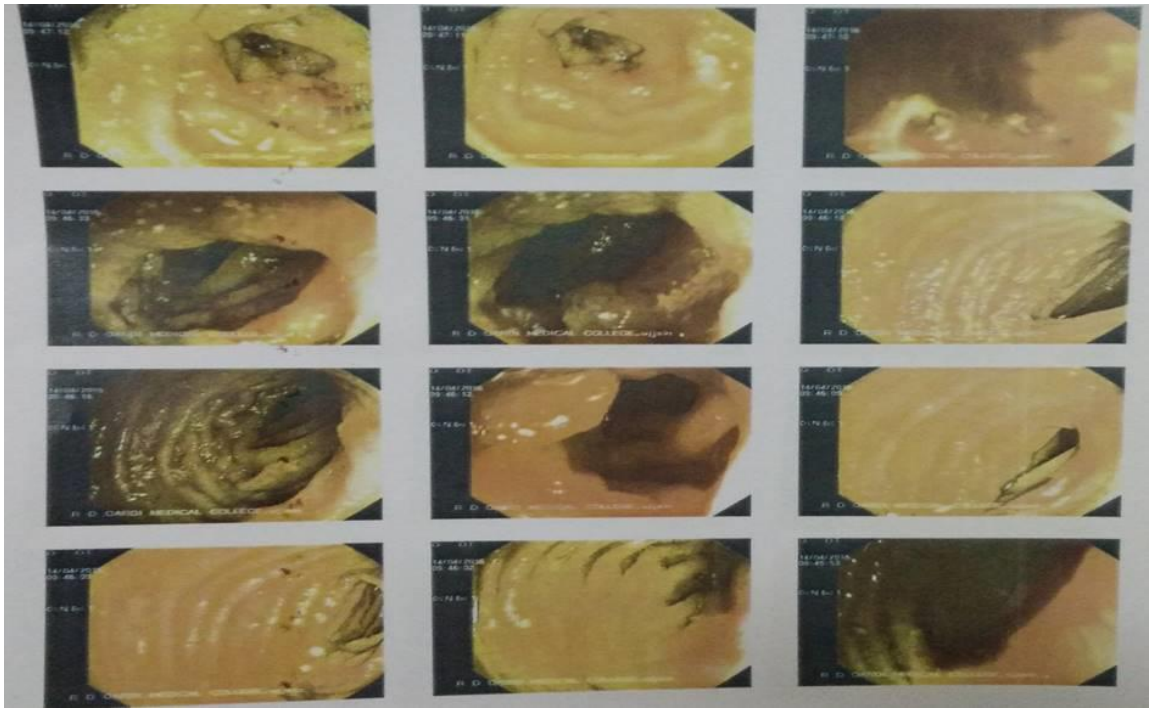


Figure - 5: Arborizing pattern of connective tissue, smooth muscle, lamina propria and glands lined by normal appearing intestinal epithelium (10X; Low power field).

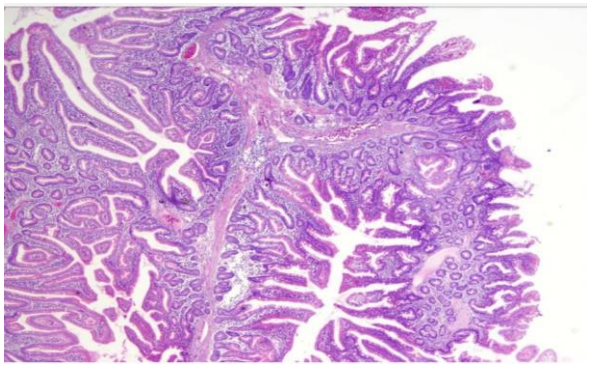


Figure - 6: Arborizing pattern of connective tissue, smooth muscle, lamina propria and glands lined by normal appearing intestinal epithelium (10X; Low power field).

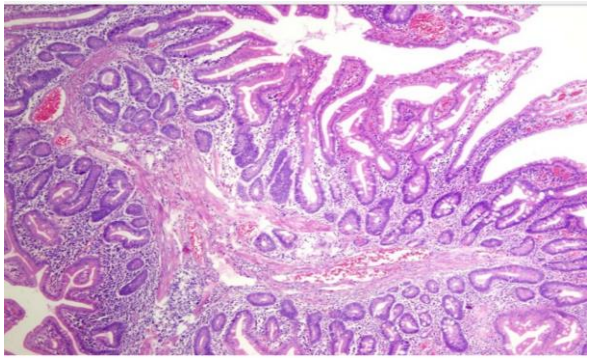


Figure - 7: Arborizing pattern of connective tissue, smooth muscle, lamina propria and glands lined by normal appearing intestinal epithelium (40X; High power field).

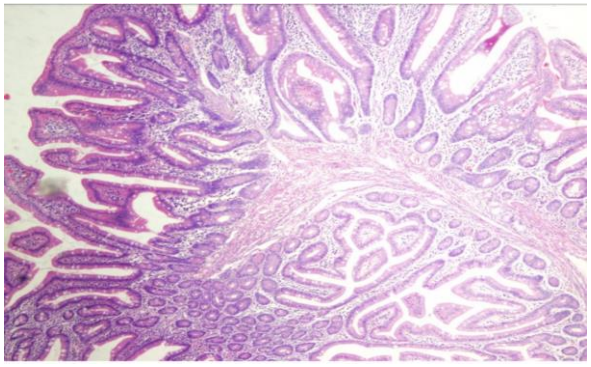
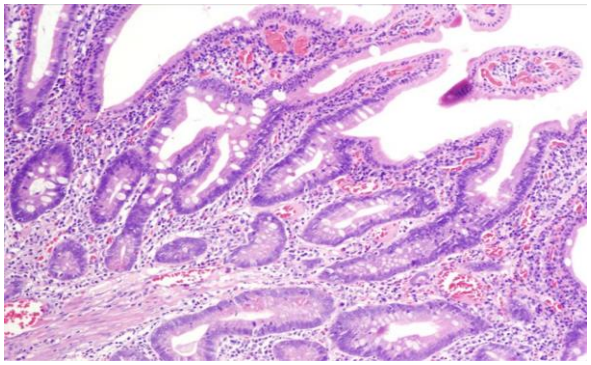


Figure - 8: Arborizing pattern of connective tissue, smooth muscle, lamina propria and glands lined by normal appearing intestinal epithelium (40X; High power field).



References

1. Rosai and Akerman's Surgical Pathology, 10th edition, Mosby, 2011, Volume I, p. 687-688.
2. Robbins and Cotran, Pathologic Basis of Disease, South Asia Edition, Elsevier, 2014, Volume II, p. 806-807.
3. Sabiston textbook of surgery, 19th edition, Saunders, 2012, Volume 2, p. 1345.
4. Swartz's principles of surgery, 10th edition, McGraw-Hill, 2014, p. 1206.
5. Thrilok C Bingi, V Pavan, B Ramulu, B Manjula, R Vemula. Peutz-Jeghers syndrome presenting as multiple intussusceptions: A case Report. Indian Journal of Applied Research, 2015; 5(3): 462-4.
6. James William, Berger Timothy, Elston Dirk. Andrews' Diseases of the Skin: Clinical Dermatology, 10th edition, Saunders, 2005; p. 857.
7. Bouquot Jerry E., Neville Brad W., Damm Douglas D., Allen Carl P. Oral and Maxillofacial Pathology, Philadelphia: Saunders, 2008; p. 16.11.
8. Francis M. Giardiello, Jill D. Brensinger. Very high risk of cancer in familial Peutz-Jeghers syndrome. Gastroentrology, 2000; 119: 1447-1453.
9. Tamana Choudhary, Suraiya Enam Dhaka. Peutz-Jeghers polyp: a retrospective study. BSMMU J, 2012; 5(1): 12-17.