Familial Adenomatous Polyps with Synchronous Colorectal Carcinoma

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Case Report

Abstract: Familial Adenomatous Polyposis is a genetic condition. It is diagnosed when a person develops more than 100 Adenomatous colon polyps. Synchronous colorectal neoplasias, defined as 2 or more primary tumours identified in the same patient and at the same time. We hereby report a case of a 25yr female presenting with synchronous colorectal cancer and multiple adenomatous polyps.

Keywords: synchronous tumors, colorectal carcinoma, familial adenomatous polyposis.

Introduction

Familial Adenomatous Polyposis is a genetic condition. It is diagnosed when a person develops more than 100 Adenomatous colon polyps. The average age for polyps to develop in people with FAP is in the mid-teens. More than 95% of people with FAP will have multiple colon polyps by age 35. If FAP is not recognized and treated, there is almost a 100% chance that a person will develop colorectal cancer. The risk of colon cancer is 87% by age 45. Synchronous colorectal cancer, defined as two or more primary colorectal cancers identified at the same time. Each tumor must be clearly malignant as determined by histological evaluation, geographically separate and distinct.

Case Report:

A 25yr female patient was admitted with chief complaints of diarrhea and loss of appetite. On routine rectal examination patient was found to have decreased sphincter tone and growth in anorectum. On pervaginal examination posterior vaginal wall is indurated but mucosa is normal.

Sigmoidoscopy was performed and found a large villeferous growth from the anal verge (figure.1). Scope could not be negotiated across the growth. Biopsy was taken and was reported as adenocarcinoma.

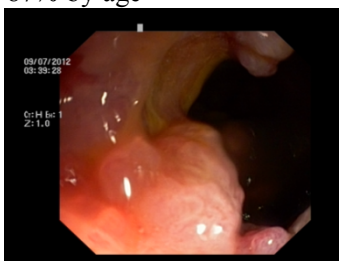


Fig.1

Ultrasound revealed Space occupying lesion in right lower pelvis with irregular margins and mixed internal echos, Left kidney hydronephrosis with hydroureter. MRI pelvis showed thickened and heterogeneous rectal and anal wall with decreased lumen and circumferential growth also associated with inflamatory abscess at left perineal soft tissues (figure.2).

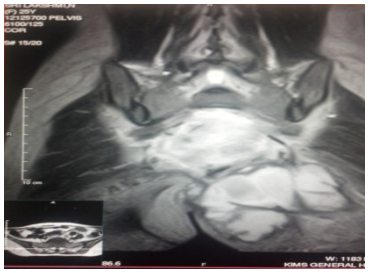


Fig.2

APR is planned and on surgery hard growth was found in rectum infiltrating lateral ligament. No distant metastasis and no lymphadenopathy found. Abdominal part of APR is completed and found to have multiple polyps upto caecum. Surgery is converted to total colectomy with end ileostomy. On examining the resected bowel another hard mass was found at splenic flexure of colon (figure.3).

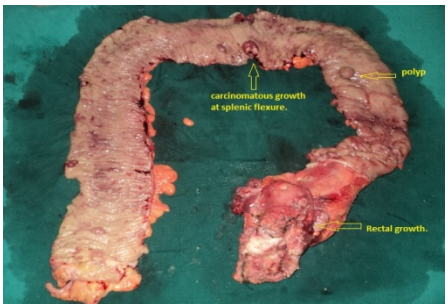
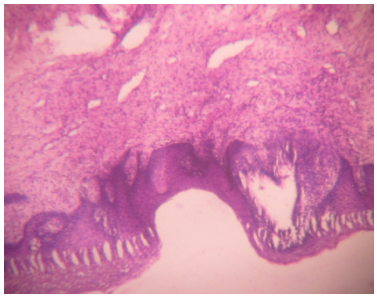


Fig.3

On histopathological examination, sections studied from rectal mass show features of well differentiated adenocarcinoma infiltrating into muscularis propria (figure.4a&4b). Margins are free from infiltration. Sections from lymph nodes show reactive hyperplasia. Sections from hard polypoidal mass at splenic flexure show features of well differentiated adenocarcinoma infiltrating into muscle coat. Sections from polyps show features of adenomatous hyperplasia and some show features of adenoma. Patient recovered well after surgery. Patients family could not be screened as patient father and mother are not alive and children are only 5yr old.



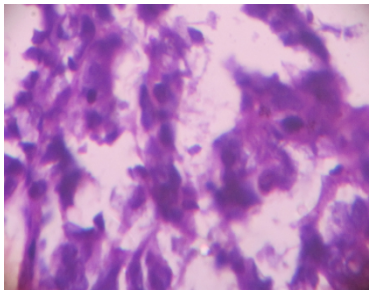


Fig 4a ***Fig 4b***

Figure.4a & 4b *showing anal mucosa and hyperchromatic nuclei.*

Discussion

A polyp is a tissue growth protruding from the mucous membrane into the lumen of GI tract.

FAMILIAL ADENOMATOUS POLYPOSIS

Inherited disorder transmitted by autosomal dominant trait. It appears in second decade of life. There are more than 100 polyps in colon or any number of polyps with positive family history. The incidence of the mutation is between 1 in 10,000 and 1 in 15,000 births. Some people have a variant of the disorder, called **Attenuated Familial Adenomatous Polyposis**, in which polyp growth is delayed. A milder type of familial adenomatous polyposis, called **Autosomal Recessive FAP** has also been identified.

SCREENING OF FAMILY MEMBERS

- All the family members should be examined at the age of 10 yrs clinically and with colonoscopy.
- Yearly colonoscopy till age of 20 yrs.
- If no polyps by 20yrs, 5yr colonoscopy should be done till age of 50.
- Tests for FAP gene and pigmented spots in retina are also helpful in screening.

SURGICAL OPTIONS FOR FAP

- Total proctocolectomy with ileostomy.
- Total Colectomy with fulguration of rectal polyp and ileorectal anastomosis.
- Proctocolectomy with ileoanal anastomosis with a ileal pouch reservoir.

The youngest patient encountered in literature was 8 years old [1].

Coffey was the first person to perform the total proctocolectomy and ileostom[1].

Ravitch et al performed ileoanal anostomosis Combined with Total proctocolectomy [1].

SYNCHRONOUS COLORECTAL CARCINOMA:

According to Cunliffe et al, synchronous adenocarcinomas can be two or more in number, detected either pre / intraoperatively, or in a 6 month period postoperatively. They should be distinctly separate by at least 4 cm distance [2].

The most voluminous synchronous cancer is called "first primitive" or "index" cancer [4].

The reported incidence of synchronous colorectal carcinoma ranged between 2.3 and 12.4%. One study has shown that intraoperative palpation can miss up to 69% of the SC while other studies have shown altered planned surgical procedure due to preoperatively diagnosed synchronous lesions in 11–44%[7].

A routine preoperative colonoscopy has been recommended for patients diagnosed with colorectal cancer in order to identify synchronous polyps and/or cancer. It has been reported that a preoperative colonoscopy was not possible in as many as 50% of cases. Preoperative barium enema examination is one of the tool for detecting synchronous tumours. If both procedures are contraindicated in these patients, colonoscopic evaluation should be performed 3 months after resection [8].

Several authors have shown the usefulness of intraoperative colonoscopy when a preoperative colonoscopy was not possible.

Preoperative or intraoperative diagnosis of the presence of synchronous colorectal carcinomas is very important because once they are overlooked; they present as early metachronous carcinomas with advanced stages.

Conclusion:

Synchronous colorectal polyps or cancer are frequent and their preoperative detection is important for optimal surgical planning and treatment. Surgeons should be alert about the possibility of presence of multiple lesions and be able to avoid second operation for missing lesion.

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