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Hereditary Nonpolyposis Colorectal Cancer and its Management

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Opinion Article

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DESCRIPTION

Hereditary Nonpolyposis Colorectal Cancer (HNPCC), also known as Lynch syndrome, is an autosomal dominant genetic condition that is second most commonly associated with endometrial cancer and associated with stomach, small intestine, hepatobiliary tract, upper urinary tract, brain, and skin cancers. These cancers are more common because of inherited mutations that hinder DNA mismatch repair. It is a particular kind of cancer syndrome. The term HNPCC has lost favour because patients with Lynch syndrome can develop polyps. Blood in the stool, diarrhoea or constipation, and unintended weight loss are typical signs and symptoms of colon cancer, which affects the proximal colon in two-thirds of cases. The average age at which a member of a family with Amsterdam criteria meets colorectal cancer diagnosis is 44. Endometrial cancer is typically diagnosed when a woman is 46 years old. Endometrial cancer is the most frequent sentinel cancer in Lynch syndrome, occurring in about half of women with HNPCC who have both colon and endometrial cancer. Abnormal vaginal bleeding is the sign of endometrial cancer that occurs most frequently.

The most frequently reported pathology for gastric cancer at HNPCC is intestinal-type adenocarcinoma, with a mean age of diagnosis of 56 years. The median age at diagnosis for ovarian cancers linked to HNPCC is 42.5 years, and about 30% of cases are identified before the age of 40.

The most prevalent type of hereditary colorectal cancer is Hereditary Nonpolyposis Colorectal Cancer (HNPCC), also known as Lynch syndrome (CRC). Since HNPCC lacks distinct clinical stigmata of its cancer genetic risk, unlike its Familial Adenomatous Polyposis (FAP) hereditary cancer counterpart, a well-organized family history of cancer is

crucial for diagnosis. The most prevalent germ-line mutations, hMSH2 and hMLH1, were identified in the 1990s, giving the diagnosis of Lynch syndrome a significant boost. Its natural history determines how best to treat it medically. For instance, about 70% of CRCs develop close to the splenic flexure, with the cecum accounting for about one-third of the cancers, necessitating a complete colonoscopy.

The management of initial CRC should at least include a subtotal colectomy due to the high rate of metachronous CRCs. Prior to DNA testing and when the results are disclosed, genetic counselling is crucial. It is crucial to educate both patients and doctors about every aspect of this disorder. Patients must be aware of the significance of their genetic risk status and the natural history of it in order to demonstrate compliance with germ-line testing, screening, and management options.

An autosomal dominant disorder known as Hereditary Nonpolyposis Colorectal Cancer (HNPCC) is characterised by the occurrence of multiple cases of colorectal cancer in a family without gastrointestinal polyposis. Although the prevalence of this syndrome is not yet known, it may be responsible for 1%–5% of all colorectal cancers. Before the genetic cause of this syndrome was discovered, it was known that there was a familial aggregation of colorectal cancers with early onset, an excess of nearby, frequently multiple primary tumours, and an excess of cancers in specific other organs.

A series of findings that connected this type of genomic instability to a defect in the DNA Mismatch Repair (MMR) system were made in response to the recent description of an abnormality known as "microsatellite instability," which is present in nearly all cancers from HNPCC patients and in approximately 12 percent to 15 percent of sporadic cases. Four HNPCC genes have been discovered by unaffiliated researchers: hMSH2 (a homologue of the prokaryotic DNA MMR gene MutS), hMLHl, hPMSl, and hPMS2 (all homologues of the prokaryotic DNA MMR gene MutL). In the germline cells of HNPCC families, mutations in each of the four genes have been discovered.

Because microsatellite DNA sequences are unstable, deficiencies in the DNA mismatch repair process significantly raise the risk of developing a particular type of cancer. Hereditary nonpolyposis colorectal cancer (HNPCC), also known as the Lynch syndrome, is caused by a germline mutation in either the hMSH2 or hMLH1 mismatch repair gene. Since affected people have an 80% lifetime risk of developing colon cancer, aggressive cancer surveillance programmes are crucial for both them and their at-risk relatives.

By meeting the Amsterdam clinical criteria or by genetic testing for germline mutations in hMSH2 or hMLH1, HNPCC can be diagnosed. Within established HNPCC kindred, genetic testing is particularly helpful for assessing cancer risk in families with atypical clinical features. One distinguishing characteristic of tumours linked to HNPCC is DNA microsatellite instability (MSI), which is also present in up to 15% of cases of sporadic colorectal cancer. The most intriguing aspect of colorectal tumours with MSI is their favourable natural history, which is distinct in their biological behaviour. A compelling example of how molecular genetics and clinical care interact is the study of HNPCC.