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Editorial

Serrated Polyposis Syndrome[☆]



Síndrome de poliposis serrada

For many years, the development of colorectal cancer (CRC) has been explained by means of the adenoma-carcinoma sequence based on the suppressor or chromosomal instability pathway and the microsatellite instability or mutator pathway. In the last 20 years, a third CRC carcinogenesis pathway has been identified. It is characterized by aberrant hypermethylation of the promoter region of certain suppressor genes (CpG island methylator phenotype), silencing their function. This pathway is thought to be responsible for 15%–20% of sporadic CRC.¹ Its precursor lesions are serrated polyps or lesions.² Currently, the World Health Organization (WHO) classifies serrated polyps into 3 types: hyperplastic polyps (very frequent, preferably distal location and, probably, no potential for malignization); sessile serrated polyps, with or without dysplasia (uncommon, usually proximal, with malignant potential, especially if they present dysplasia); and traditional serrated adenomas (very uncommon, distal and at risk for neoplastic transformation). The clinical criteria that have been related with higher risk for malignant progression of serrated lesions include multiplicity, size (larger than 10 mm), proximal location and presence of dysplasia.

Serrated polyposis syndrome (SPS) is a special situation that was first described in 1980.³ It is a syndrome characterized by the presence of numerous serrated polyps, which may or may not be large,^{4–6} with a family history and an exceptionally high risk for CRC.^{7,8} The genetic base continues to be unknown so that, currently, the definition continues to be phenotypic and includes a very heterogeneous group of patients. The WHO has reached a consensus on clinical criteria for the clinical diagnosis of SPS.⁹ These criteria have recently been updated,¹⁰ and at least one of the following criteria should be met: 5 or more serrated polyps proximal to the sigmoid colon, 2 of which >10 mm; any number of serrated polyps proximal to the sigmoid colon in a first-degree relative with SPS; or more than 20 serrated polyps throughout the entire colon. It is a rare disease, with an incidence of one in every 100 000 inhabitants,¹¹ although it is probably an underdiagnosed condition. The

risk for CRC has not been established, but it has been estimated at 70%.¹² Although it is thought that there would not be a greater risk for extracolonic tumors,¹³ some authors have reported a higher frequency of non-colonic neoplasms, without being able to establish whether there is a true correlation with SPS.¹⁴ Reports of a family history of CRC ranged between 0% and 59% of cases.¹² Likewise, there are reports of families with autosomal dominant inheritance patterns, recessive autosomal profiles, or with no family history at all. All these suggest that SPS encompasses a heterogeneous group of patients who probably represent diseases with different genetic bases.

As for the management of patients with identified SPS, it is recommended that they be included in a strict endoscopic follow-up program. The current recommendation is an annual total colonoscopy, which should be high-quality and preferably use techniques such as chromoendoscopy with color enhancement (e.g. indigo carmine) or virtual chromoendoscopy (narrow band imaging) in order to increase the detection rate of serrated lesions. If the disease is not considered controllable using endoscopy, subtotal colectomy with ileorectal anastomosis should be proposed (similar to attenuated adenomatous polyposis), although this aspect is controversial.

Nevertheless, the information we have to date about SPS is limited; therefore, most recommendations are based on the opinion of experts, but they are not supported by scientific evidence.

This current issue of our journal presents a retrospective cohort of 23 patients from a single center who meet the definition of SPS based on endoscopies done between 2005 and 2012, with an analysis of their clinical and phenotypic characteristics. It should be emphasized that the inclusion of patients was done by reevaluating endoscopy reports and reviewing pathology studies. In this case, 0.08% of the colonoscopies met criteria for SPS. In recent prospective cohorts in our setting, up to 0.3% of colonoscopies performed due to a fecal occult blood test were diagnostic for SPS.¹⁵ Due

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to their peculiar characteristics (flat, covered by mucus, location preferably in the right colon), serrated polyps are especially difficult to detect. Thus, the introduction of improvements in endoscopy quality (high definition, chromoendoscopy) is expected to improve future detection rates. Likewise, the higher percentage of distal and mixed SPS phenotypes observed may be overestimated as the right colon is more difficult to evaluate, and it is precisely where most undetected polyps and interval cancer are concentrated.¹⁶

Second of all, the authors identified an elevated association with smoking, which was previously reported¹⁴ and requires smoking abstinence to be recommended within the management of patients with SPS. As for surgical treatment, subtotal colectomy is currently recommended with ileo-rectal anastomosis. Nonetheless, it is a decision that should be agreed upon with the patient, taking into account the pros and cons. It is possible that more conservative surgery may be able to be adapted according to the predominant phenotype of the disease, as suggested by the authors (e.g. right hemicolectomy in the case of a proximal phenotype).

As for the endoscopic follow-up of diagnosed patients, an annual colonoscopy is recommended, although longitudinal follow-up studies are required in order to evaluate the usefulness of the proposed strategies and adapt them to the risk of metachronous lesions of each patient phenotype.

Therefore, it is necessary to study in detail the clinical characteristics of patients with SPS in order to classify them into subgroups that would aid in a future molecular diagnosis, identify the actual risk for CRC, identify epidemiologic factors that increase risk for neoplastic transformation and establish an adequate endoscopic follow-up strategy based on longitudinal follow-up studies. To further our knowledge about this disease, contributions from different groups are fundamental, as are series with large patient cohorts in multicenter studies.

Conflict of Interests

The authors have no conflict of interests to declare.

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