found to have a single colonic polyp, which was removed; biopsy revealed adenomatous polyp with no dysplasia.

Classically the polyps in Peutz-Jegher syndrome are hamartomatous, but adenomas and adenocarcinomas are also reported. Hizawa et al described his experience with 75 GI polyps from 7 patients with PJS where 71 had hamartomatous polyps, 2 adenoma, 1 cancer in adenoma, and 1 pyogenic granuloma.

Small bowel polyps are treated with laparotomy with enteroscopy and polypectomy. Our patient had developed colonic and ovarian malignancies along with small bowel polyps; over just a two-year follow-up, all of these were succesfully treated. A hamartoma-adenoma-carcinoma sequence as been described in PJS.

References


Malakoplakia of colon in a child with celiac disease and chronic granulomatous disease

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Malakoplakia is a rare pseudotumoral inflammatory disease known to affect immunocompromised subjects. We report a 4-year-old boy with malakoplakia of colon who was diagnosed with celiac disease in late infancy; despite aggressive nutritional and medical management for celiac disease, symptoms did not resolve. His nitro-blue-tetrazolium test was positive. Duodenal biopsy showed moderate duodenitis with moderate villous atrophy.

A 4-year-old boy was hospitalized for chronic diarrhea, severe malnutrition and abdominal pain. He was a normal 3300-gram neonate at birth with normal growth and development during the first year of life. He had been hospitalized one year ago for chronic diarrhea. At that time anti-endomysial antibody test was positive twice and duodenal biopsy showed moderate duodenitis with moderate villous atrophy. He was on gluten-free diet, but because of no response over 9 months, his parents discontinued the regimen.

His brother had had chronic intractable diarrhea and recurrent skin infections with no response to gluten-free diet. He died due to an unknown infection at age 6 years.

On examination he was a cachectic child with 9.5 Kg weight, 85 cm height and 49 cm head circumference. He had pale conjunctiva, brittle hair, distended abdomen and no organomegaly. His mental status was normal.

Stool examination was negative for parasites. Immunologic assessment was near normal (35% T cells, 28% B cells, 74% CD4, 17% CD8) except for positive nitro-blue-tetrazolium test. Anti-HIV and anti-HTLV1 were also negative. On colonoscopy he had erythematous mucosa in the rectosigmoid and descending colon; biopsy showed Michaelis-Gutmann bodies.

After starting ciprofloxacin followed by co-trimoxaole, on regular diet, his diarrhea stopped. His weight increased by 2.5 Kg after one month but bloating and abdominal distension did not resolve. We reassessed him for celiac disease. Anti-tissue transglutaminase test was positive. Duodenal biopsy showed partial villous atrophy; cryptosporidium infestation was also seen.

To our knowledge, this is the first case reported of gastrointestinal malakoplakia in a child, occurring with CGD and celiac disease.

The etiology of malakoplakia is unclear. Current evidence points to a defect in macrophage killing activity. Nondigested micro-organisms are found within the lysosomes of macrophages in affected persons. Macrophages from these patients show a decrease
in cyclic guanosine monophosphate (cGMP), resulting in impaired bactericidal activity. Peripheral blood monocytes are also found to have decreased bactericidal activity.\(^3\)

Malakoplakia has been reported in patients receiving chemotherapy or immunosuppressive therapy for organ transplantation, as well as various immune deficient states.\(^4\)

CGD is also manifested by defective phagocytosis (primarily neutrophil). It is caused by impaired NADPH oxidase function, resulting in deficient production of toxic oxygen metabolites, such as hydroxyl radicals and hydrogen peroxide, which are important in bactericidal activity. Consequently, although phagocytes ingest microorganisms normally, killing is defective, and affected persons are susceptible to pyogenic and fungal infections.

In CGD, chronic granulomatous inflammation without detectable infection occurs in a variety of locations, including the gastrointestinal tract, mimicking Crohn’s disease. CGD therefore should be considered in the differential diagnosis of any young patient with atypical inflammatory bowel disease.\(^4\)

The association between malakoplakia and CGD has not been reported in literature. Malakoplakia has been associated with chronic autoimmune systemic disorders, although association with celiac disease has not been reported. The association of celiac disease and CGD has been described in two cases.\(^6,7\)

The presence of malakoplakia, CGD and celiac disease together can be explained by an immune abnormality. A dysfunction of the innate, nonspecific immune protection provided by neutrophils, monocytes and macrophages might allow bacterial penetration of the bowel mucosa, triggering a chronic T-cell driven inflammation.

References

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Acknowledgment: We thank Dr. Zahra Abbaspoor, Dr. Vahedi, and Ms Malihe Javadzade for their kind assistance

Received October 3, 2005. Received in final revised form February 10, 2006. Accepted March 18, 2006

Correction
An advertisement for the Ranbaxy Science Foundation "Ranbaxy Research Awards – 2004" was erroneously inserted in the March-April 2006 issue of the Journal. The advertisement is invalid and stands withdrawn. We regret the error.

– Editor

Addendum
The mailing address for the completed questionnaires and pro forma of the Indian Society of Gastroenterology Task Force on Inflammatory Bowel Disease was inadvertently omitted on page 110 of the March-April 2006 issue of the Journal. This should read as: Dr Govind Makharia, Coordinator, ISG Task Force on Inflammatory Bowel Disease, Department of Gastroenterology, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110 029.

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