

Lymphoid Polyposis Mimicking Malignant Lymphoma in Twin Girls: Case Report

J Yi Li, Z Yu, E Gillies

Citation

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Abstract

The identical twin sisters had rectal bleeding due to multiple polypoid lesions in their colon. Histologic findings were identical and showed the lesions were located in the submucosa and were made up of lymphoid tissue with follicle formation. The enlarged germinal centers were occupied by numerous large atypical lymphocytes and small population of small mature lymphocytes and scattered tingible body macrophages. The large atypical lymphocytes were positive for CD20. The small lymphocytes were positive for CD3 and CD5. Bcl-2 was positive in the mantle zone alone. Both large and small lymphocytes lacked kappa and lambda immunoglobulin light chain expression and cyclin D1. The marked cytological atypia and high proliferative index of large atypical lymphocytes mimicked malignant lymphoma. Immunoglobulin heavy chain gene rearrangement assay did not reveal a monoclonal rearrangement. Immunohistochemical profile and molecular analysis confirmed the diagnosis of lymphoid polyposis, an entity which had not been previously reported in identical twins.

INTRODUCTION

Lymphoid polyposis is a rare disorder and is characterized by multiple sessile polyps in the rectosigmoid colon and ileum formed by lymphoid nodules with reactive germinal centers.^{1,2} It can be confused with malignant lymphoma of the gastrointestinal tract. Colonic polyps are one of the more common causes of rectal bleeding in children even though polyps are not very common in kids.¹ To the best of our knowledge, the case presented here is the first report of lymphoid polyposis in identical twins. We also demonstrate the status of immunoglobulin heavy chain gene rearrangement, the expression level of kappa and lambda immunoglobulin light chain, and proliferation index in our case, which have not been studied in this entity previously.

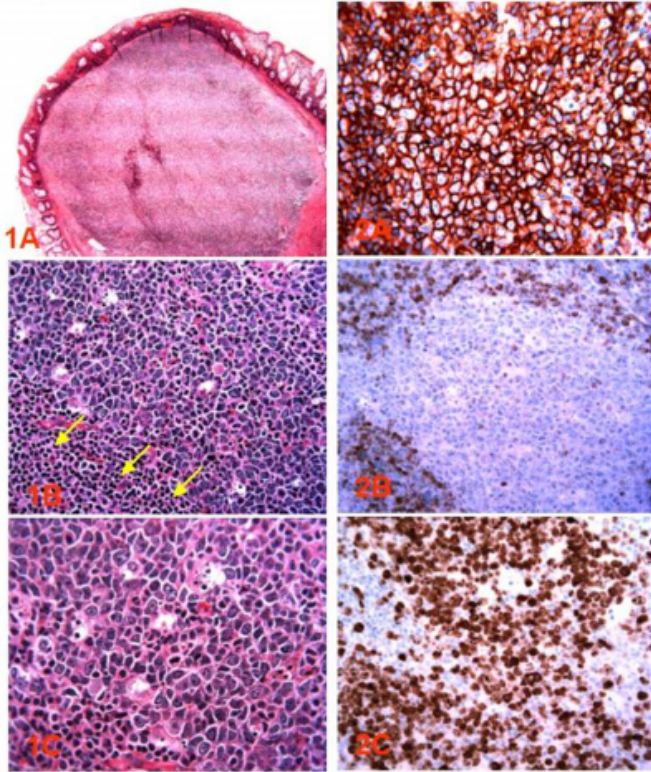
CASE PRESENTATION

The identical twin girls, at age 6, were admitted for rectal bleeding. Both underwent endoscopic examination. The endoscopy showed multiple polypoid lesions in colon. Polypectomies were performed. Grossly, the specimens were multiple pink to brown oval to cylindrical polyps ranging from 0.3 to 1 cm in the greatest dimension. Microscopically, the polypoid lesions from both twins were histologically identical and were comprised of multiple, single to confluent submucosal nodules consisting of one or more massively enlarged germinal centers with a thin rim of the mantle zone.

The germinal centers were made up of large atypical lymphocytes with clear chromatin and prominent central nucleoli, small mature lymphocytes and occasional tingible body macrophages. Most of the large atypical lymphoid cells and minor population of small lymphocytes in the nodules and the lamina propria were diffusely immunoreactive for CD20 (Figure A). Majority of small lymphocytes were positive for CD3 and CD5. BCL-2 was positive for most of small lymphocytes in the mantle zone, but not for the large atypical lymphocytes in the germinal centers (Figure B). Cyclin D1 was non-immunoreactive. A large population of large atypical lymphocytes (>80%) were labeled with Ki-67 (Figure C). Almost all of the lymphocytes lack kappa and lambda immunoglobulin heavy chain expression. Immunoglobulin heavy chain gene rearrangement assay was consistent with polyclonal lymphoid proliferation.

Figure 1

Immunohistochemical staining profile of lymphoid polyposis (400 X). 2A: CD20, large atypical lymphocyte are positive. 2B: Bcl-2, most of small lymphocytes in the mantle zone are positive. 2C: Ki-67, majority of large atypical lymphocyte are positive.



DISCUSSION AND CONCLUSIONS

Lymphoid polyposis of GI tract is a benign reactive process, which is also named as lymphoid hyperplasia, pseudolymphoma and rectal tonsil.^{1,2,3,4} Majority that occurred in children are multiple, but the lesions in adults are predominantly solitary. Colon et al. report 147 children with lymphoid polyposis with lesions in large bowel (57%) being more frequent than those in the small bowel (43%). More than half of the children have abdominal pain and one third of them presented with bright red blood per rectum. Lymphoid polyposis in children between the ages of 2 and 6 years were mainly confined to the colon. Pain and bleeding occurred equally. None of these children with isolated lymphoid polyposis have developed malignant lymphoma after long-term follow-up.³ There is also a reported association of lymphoid polyposis with familial adenomatous polyposis.¹

On histologic examination, these lesions are located in the mucosa and submucosa and are made up of lymphoid tissue with reactive germinal centers. The germinal centers are

obviously enlarged with compressed rim of small lymphocytes in the mantle zone. Tingible body macrophages are found in the germinal centers.^{1,2,4} They are strongly reactive to CD20 and demonstrate the B-cell character of the germinal centers. The BCL-2 immunostain often failed to react in the germinal center.^{2,4} The major differential diagnosis of lymphoid polyposis should be multiple lymphomatous polyposis (MLP). MLP is a distinctive and rare gastrointestinal non-Hodgkin's lymphomas in adults characterized by polypoid lymphomatous tissue in entire GI tract, which was first described by Cornes in 1961.⁵ MLP is more common in men than women and occurs in the fifth and sixth decades of life.⁶ The patients often present with abdominal pain, diarrhea, hematochezia, and a palpable mass.⁶ Vast majority of MLP are mantle cell lymphomas (MCL). The histologic feature of MCL is a monomorphic population of small lymphocytes with mantle zone formation around reactive follicles. Immunohistochemically, the MCL cells are positive for CD5, CD20 and cyclin D1, but is negative for CD23. Translocation (11;14)(q13;q32) is found in most MCL.^{6,7} MLP is an aggressive disease. Even with aggressive combination therapy, the 5-year survival rate only reaches to 60%.^{6,7} Other GI lymphomas such as follicular lymphoma (FL), angioimmunoblastic lymphoma and MALToma manifest as MLP.⁸ Follicular lymphoma presenting as lymphomatous polyposis demonstrates Bcl-2 immunostaining of follicular germinal centers which is not seen in lymphoid polyposis.

In conclusion, we present the first case of lymphoid polyposis from 6-year-old identical twins. The polyclonal profile of immunoglobulin heavy chain gene rearrangement assay and the absence of BCL-2 staining of germinal centers support the benign feature of our cases. However, the lack of light chain expression,⁹ marked cytological atypia and high proliferative index of large atypical lymphocytes raises the possibility and suspicion of transformation to malignant lymphoma even though no such sequelae for all the children reported with isolated lymphoid polyposis have so far occurred.

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Author Information

Jian Yi Li, MD, PhD

Department of Pathology, The Methodist Hospital

Zhongxin Yu, MD

Department of Pathology, The University of Oklahoma Health Sciences Center

Elizabeth M. Gillies, MD

Department of Pathology, The University of Oklahoma Health Sciences Center