Clinical Neurology and Neurosurgery 115 (2013) 117–120

Contents lists available at SciVerse ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Review

Turcot's syndrome associated with intestinal non-Hodgkin's lymphoma: Case report and review of literature

ShunChang Ma[a], YeShuai Hu[a], Jun Yang[a,*], XiaoNa Zhou[b]

[a] Neurosurgery, Fuxing Hospital Affiliated to Capital Medical University, Beijing, China
[b] General Surgery, Friendship Hospital Affiliated to Capital Medical University, Beijing, China

ARTICLE INFO

Article history:
Received 11 March 2012
Received in revised form 12 April 2012
Accepted 30 April 2012
Available online 5 June 2012

Keywords:
Turcot's syndrome
Colon
Astrocytoma
Glioblastoma
Non-Hodgkin's lymphoma
Polyposis

ABSTRACT

A 15-year-old boy was admitted with the diagnosis of colonic polyposis, and during a 2-year follow-up, he underwent operation for right parieto-occipital anaplastic astrocytoma, left-side colonic non-Hodgkin lymphoma (NHL) and cerebella glioblastoma which were all confirmed by histology. Although cases of Turcot's syndrome (TS) (colonic polyposis and primary brain tumour occurring in the same patient) have been previously described, association with haematological malignancy is rare. We hereby report such a case with TS.

Contents

1. Introduction

TS was first described in a case report by Crail in 1949 [1], and it became known as TS in 1959 when Turcot identified two siblings diagnosed with polyposis coli who each had a primary CNS tumour [2]. It was a rare hereditary disease and has been seen as a phenotypic variant of hereditary non-polyposis colorectal cancer (HNPCC) and familial adenomatous polyposis (FAP) [3]; however, the mode of inheritance was complex and still in controversy. Although cases of TS have been previously described, association with haematological malignancy is rare. We hereby report such a case with TS.

2. Case report

The patient is a 15-year-old boy who presented with stomachache and rectal bleeding. He is the second sibling of the two and there was no history of familial illness or of parental consanguinity.

By sigmoidoscopy and colonography (Fig. 1), a few small rectal polyps were identified and several polyps in the distal descending colon were excised endoscopically, and histology revealed lower-level tumour-like lesion and tubular adenomas, mild epithelial hyperplasia. The patient later underwent a limited sigmoidectomy, and histology revealed lower-level tumour-like lesion and tubular adenomas, mild and moderate epithelial hyperplasia (Fig. 2).

He remained well for 5 months until he was admitted for investigation of cephalagia, nausea and vomit. Subsequent CT (Fig. 3) of the head revealed a right parieto-occipital lesion. The patient underwent parieto-occipital craniotomy with resection of the mass and histology revealed anaplastic astrocytoma (Fig. 4). The patient received a course of radical radiotherapy (60 Gys in 30 fractions), accompanied with oral steroids and cytotoxic chemotherapy (oral

* Corresponding author.
E-mail addresses: shunchangma@hotmail.com (S.C. Ma), junhou2000@hotmail.com (Y.S. Hu), yangjun6778@tom.com (J. Yang), doctorma1513@hotmail.com (X.N. Zhou).

0303-8467/$ – see front matter © 2012 Elsevier B.V. All rights reserved.
http://dx.doi.org/10.1016/j.clineuro.2012.04.026

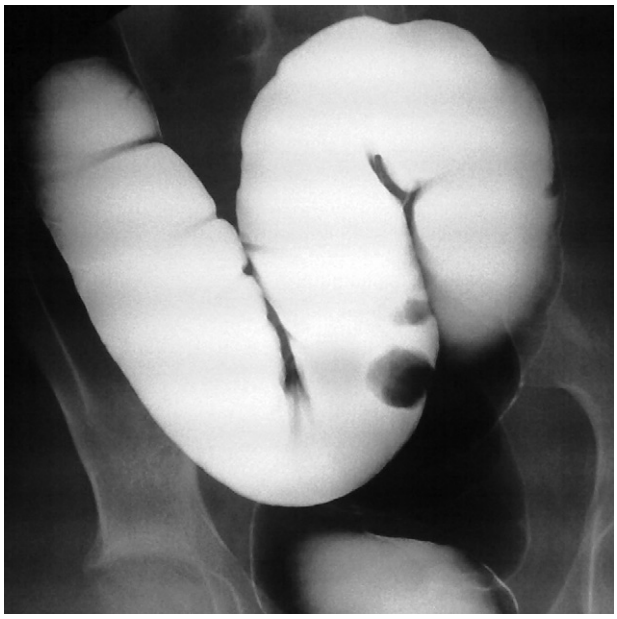

Fig. 1. Colonography showed filling defect in the colon.

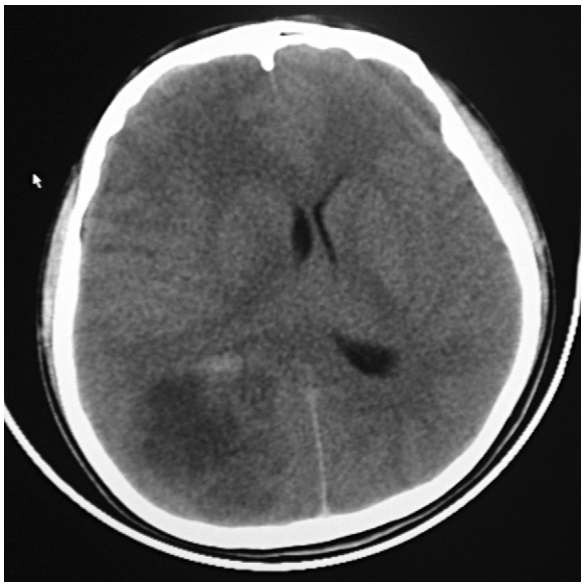

Fig. 3. Preoperative computer tomography showed a right parieto-occipital mass with low density.

Temodal 6 cycles for 42 days, 75 mg/m^2). He made a good recovery and had no further neurological sequelae. Serial MRI brain scans showed no further change.

Three months later he was admitted again due to rectal bleeding and a left hemicolectomy was performed, and histology revealed high-level tumour-like lesion and tubular adenomas, moderate to severe epithelial hyperplasia, non-Hodgkin lymphoma (NHL) (Fig. 5). The patient received a standard chemotherapy of CHOP regimen (CTX, 750 mg/m^2, ADR, 40 mg/m^2, VCR, 1.4 mg/m^2, iv. for the 1st and 8th days, prednisone 60 mg, po.1st to 14th days, 28 days a cycle for 6 cycles) for the NHL.

During the post-operative follow-up of contrast magnetic resonance imaging (MRI) (Fig. 6) of the head 2 months later, a cerebella mass was identified. The patient underwent craniotomy with resection of the mass and histology revealed glioblastoma (Fig. 7). The patient received a course of cytotoxic chemotherapy (oral temodal 6 cycles for 42 days, 75 mg/m^2). He made a recovery with no other sequelae except for the emaciation and myelosuppression.

He was referred to the regional cancer genetics service for counselling and molecular gene testing. Following initial contact with the service however, he has elected not to pursue gene testing. His siblings have been offered colonoscopic and head MRI surveillance and nothing positive was found.

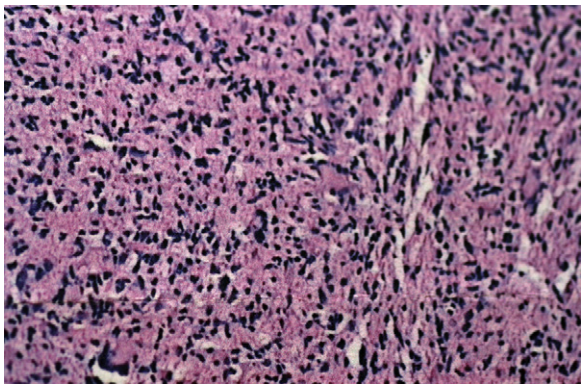

Fig. 4. Histology of the parieto-occipital lesion revealed anaplastic astrocytoma (1*100,HE).

During the years the patient has been followed up by upper and lower endoscopy and magnetic resonance imaging at regular

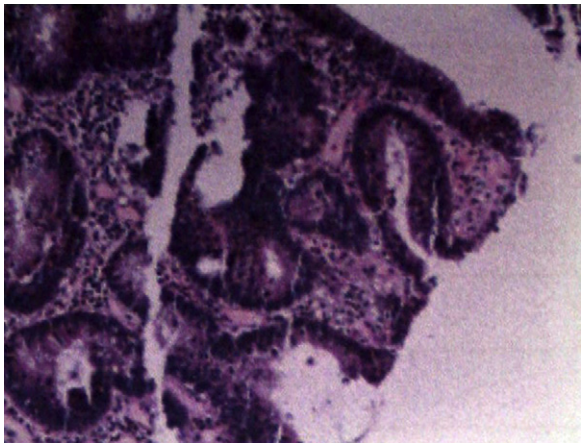

Fig. 2. Histology of the colon lesion resected revealed lower-level tumour-like lesion and tubular adenomas, mild and moderate epithelial hyperplasia (1*10,HE).

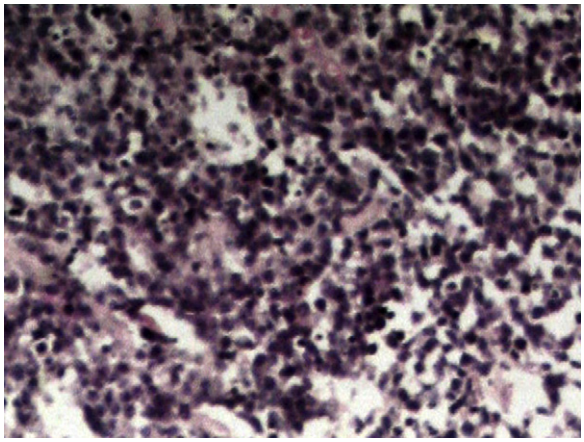

Fig. 5. Showed the histology of non-Hodgkin lymphoma (NHL) of the colon resected (1*10,HE).