CASE REPORT

Garcin's syndrome secondary to rectal carcinoma in a Nigerian child

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ABSTRACT

Colorectal carcinoma in childhood is rare in spite of the rising prevalence of the condition in the African population. Even rarer is the occurrence of multiple cranial nerve paralysis simulating Garcin's syndrome in this age group. This report discusses a case of an 11-year-old boy who presented to our unit with histologically confirmed adenocarcinoma of the rectum with cerebral metastasis and multiple cranial nerve paralysis suggestive of Garcin's syndrome. Challenges in management were highlighted, and we reviewed the literature on colorectal carcinoma in children.

Key words: Children, Garcin's syndrome, Nigeria, rectal carcinoma

INTRODUCTION

Garcin's syndrome is a progressive ipsilateral involvement of the cranial nerves resulting in paralysis of all or most cranial nerves without the involvement of long tracts or the cerebellum and without signs of increased intracranial pressure.[1] Garcin's syndrome has been described in association with cancers of nasopharyngeal and paranasal carcinomas, rhabdomyosarcoma cylindromas, craniopharyngiomas, and metastases from cancers of the breast, lung, liver, or uterus as well as colorectal carcinoma in an adult.[2] We present a case of an 11-year-old boy with rectal carcinoma with cerebral metastasis who was managed in our unit with features suggestive of Garcin's syndrome, and a review of literature on colorectal carcinoma in the pediatric age group was done.

CASE REPORT

Master AF is an 11-year-old primary four pupil who presented at the Pediatric Surgical Outpatient Clinic with 5 months history of passage of bloody mucous stools associated with anal protrusion, anorexia, weight loss, and low back pain. There was a history of constipation and progressive abdominal distension for 2 days before presentation. He had no history of fever, vomiting, or abdominal pain, and no family history of similar illness.

Examination revealed a chronically ill-looking boy, wasted, pale, and dehydrated. The abdomen was grossly distended with visible peristalsis. There was no tenderness and no palpable mass. Percussion notes were tympanitic and bowel sounds were hyperactive. Rectal examination revealed a soiled perineum with lax anal sphincter. A circumferential anal mass palpated about 2 cm from the anal verge was firm and tender, and the lumen admits only tip of the finger.

A provisional diagnosis of rectal carcinoma complicated with intestinal obstruction was made.

He was resuscitated and had an emergency sigmoid colostomy done. The findings were those of distended small and large bowel with 150 ml of golden yellow ascitic fluid. Rectal biopsy confirmed infiltrative signet ring adenocarcinoma of the rectum. The colostogram showed an irregular narrowing of the distal rectum 7 cm from the anal

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verge, with shouldering and rat-tail appearance [Figure 1]. Computerized tomography (CT) could not be done due to financial constraints.

With satisfactory hemogram, chemotherapy (intravenous 5-fluorouracil and oral levamisole) was commenced and was being planned for abdominoperineal resection. However, the patient defaulted after the 1st course of chemotherapy from the clinic. He re-presented 4 months later with complaints of poor urinary stream, straining, and frequency. He also had throbbing headache but no vomiting.

Examination revealed left periorbital nodule, right eye squint (right abducens nerve palsy), tongue deviation to the right (right hypoglossal nerve palsy), and right facioparesis (right facial nerve palsy-upper motor neurone type) [Figure 2]. He had normal vital signs and fundoscopy did not show features of raised intracranial pressure. The bladder was distended up to 15 cm from the pubic symphysis.

An assessment of an advanced colorectal cancer with chronic urinary retention and cerebral metastasis was made.

Following an aborted attempt at urethral catheterization, a suprapubic cystostomy was done during which tumor infiltration to the urinary bladder neck was noted. He was placed on cytotoxic drugs, but he had worsening symptoms, severe weight loss, anemia, proptosis, and blindness in the left eye with increased growth of the periorbital nodule.

He was thereafter admitted for resuscitation. However, soon after transfusion with the second pint of blood, he was noticed to have stopped breathing and all resuscitative efforts proved abortive. He was certified dead about 11 months after his first presentation.

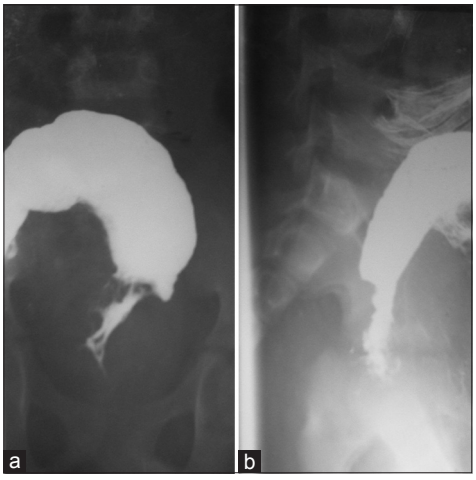

Figure 1: Distal colostogram showing irregular narrowing of the distal rectum, "shouldering" and rat-tail appearance. (a) Anteroposterior view. (b) Lateral view

DISCUSSION

Colorectal carcinoma in childhood is rare in spite of the rising prevalence of the condition in the African population.[3,4] Most cases occur in the teenage year and occurrence below the age of 13 years is very uncommon. Even rarer is the occurrence of multiple cranial nerve paralysis simulating Garcin's syndrome in this age group.

Garcin's syndrome is the progressive ipsilateral paralysis of all or most of the cranial nerves with normal intracranial pressure. The syndrome has been described in association with cancers of nasopharyngeal and paranasal carcinomas, rhabdomyosarcoma cylindromas, craniopharyngiomas, and metastases from cancers of the breast, lung, liver, or uterus as well as colorectal carcinoma in an adult,[2] however, the authors are not aware of such in children.

Paralyses of all the cranial nerves are rare, and some authors have reported patients with less than seven cranial nerves as Garcin's syndrome.[5] Our patient did not have all his cranial nerves paralyzed but did have multiple cranial nerve paralysis on the left fitting into a variant of this syndrome. Similarly, most authors who have reported patients with this syndrome agree that the prognosis is poor[5] and mortality is high. Our patient died within few months of manifesting this syndrome which supports the experience of other workers in literature.

CT scans and magnetic resonance imaging (MRI) techniques allow for early diagnosis and possible intervention. However, our center does not have MRI and the CT scans available were not performed due to financial ineptitude on the part of the parents. This challenge in resource-limited settings like ours can be surmounted if treatment of pediatric patients with oncologic diagnosis is made free. Advocacy in this regard should be pursued by all health-care givers involved in pediatric oncology.

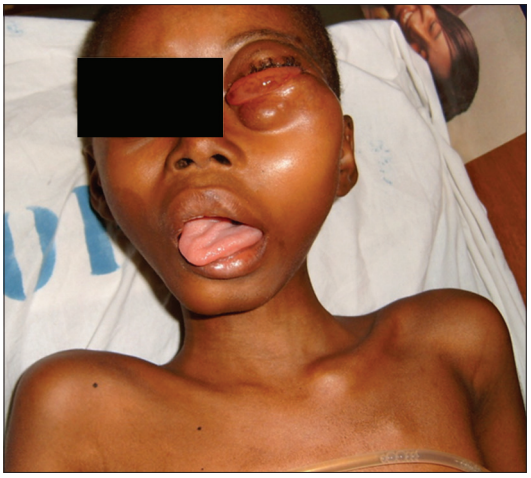

Figure 2: Deviation of the tongue to the right and angle of the mouth to the left. Note also the periorbital swelling

In a report by Ajao *et al.*, about 51 patients with colorectal cancer were reviewed, of which 15 were below 30 years but the youngest in that series was 18 years old.[6] In some reports, 50% of all patients were over 60 years.[3] Despite the clear relationship with aging, colorectal cancer is not entirely a disease of the elderly; between 2.2% and 7% of cases occur in children, adolescents, and young adults.[6] About 1–2% is between 20 and 30 years,[3] and it is a rarity (<1%) to have the disease in the teens.[4]

It had been previously thought that colorectal carcinoma is rare among African populations[7] but an increasingly younger patient generation is being seen in tropical Africa.[8] Most of the reports of the rare presentation in childhood are in teenage years.[9,10]

Many of the patients in this age group are believed to present late probably because they conceal the symptoms; however, other reports show that many patients present within a year of onset of symptoms.[9] Our patient actually presented within 5 months of the onset of the symptoms and could have benefited from surgical resection and probably this may improve his survival, as surgery is still believed to be the best form of treatment if tumor is resectable. However, it is known that occurrence below 30 years carries a poor prognosis probably because of late presentation or due to the biological behavior of the tumor.[9,10] However, other workers reviewing over 950 cases of patients under 40 years postulate that the prognosis in these patients are similar to that of adults and may even be better,[6] but it may be difficult to extrapolate this to children and adolescents as the study did not reveal the different age groups <40 years.

Our patient presented with features of intestinal obstruction as well as passage of mucous bloody stools, weight loss, change in bowel habits, and vague abdominal symptoms. This is in keeping with many reports on this subject in the young as well as in adults.[6] It is also instructive to note that the tumor was palpable with the examining finger in the rectum. This goes on to emphasize the need for this simple, inexpensive, and straightforward tool in the surgeon's armamentarium in elucidating the possibility of this condition in the young. Other authors have also emphasized this fact noting that the condition may be picked with this examination in 60–80% of cases,[5] and in a report of 8 cases from northern Nigeria,[9] all the tumors were limited to the rectosigmoid area of the large bowel.

Treatment of these patients usually includes surgical excision, chemotherapy based on 5-fluorouracil and levamisole with or without radiotherapy.[4] Surgery remains the most promising method of reducing bulk of the tumor and correlates well with survival in adults.[4] Our patient was billed to have abdominoperineal resection and chemotherapy, but financial constraints contributed to inadequate treatment in this patient coupled with incessant industrial disputes rampant in the country at the time of presentation of this patient. Resection with chemotherapy in a case report improved survival and the patient had been disease free for 5 years.[10]

Prognosis of the condition in children and adolescents and young adults has been said to be poor mainly because of the late presentation and the intrinsic nature of the tumor type – many of them being mucin-secreting tumors. Our patient had a histopathologically confirmed signet ring adenocarcinoma, and this ran a very aggressive course with clinical features of cerebral metastasis and even infiltration of the bladder.

In summary, malignant colorectal and anorectal conditions are rare in children but can occur as in this case report. Metastasis can be to the lungs, liver, bones, or brains; when the metastasis is to the brain, it may lead to multiple cranial nerve paralysis simulating Garcin's syndrome. Garcin's syndrome in its complete form is rare and this case certainly fits into one of its variants.[11] Bloody mucous stools, altered bowel habits, and vague abdominal symptoms are usual clinical features colorectal carcinoma, and digital rectal examination will probably pick these lesions in most children. Prognosis is still poor in children with this condition due to the biological behavior of the tumors and also in developing economies due to poverty and late presentation limiting the treatment options to palliative measures.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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