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Burkitt lymphoma of rectal localization to child: Role of surgery in the treatment

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Abstract
Rectal tumors are rare in children, Burkitt's lymphoma is exceptional. Only a few cases have been reported in the past 30 years, there are only 10 cases found in the litterature.

Observation: This is a 15-year-old child with a constipation table, rectal bleeding and rectal syndrome, complicated by acute intestinal obstruction. The rectal examination noted the presence of a tumoral process at 1 cm from the anal margin. A radiological and anatomopathological assessment confirmed the diagnosis of Burkitt's lymphoma of rectal localization with medullar extension. A sigmoidostomy is performed urgently in order to remove the obstacle, the patient has benefited from chemotherapy, resulting in a 70% reduction of the tumor and the disappearance of Burkitt cells from the bone marrow.

On the other hand, the patient presented a recto-urethral fistula leading to a vesicostomy, this complication has never been reported.

Conclusion: Rectal tumors in children should be suspected in front of any rectal bleeding with the interest of rectal examination.

Although treatment is based on chemotherapy, surgery occupies a significant place in the management of Burkitt's lymphoma of rectal location.

Keywords: burkitt lymphoma, Rectum, child, surgery

1. Introduction

The tumoral pathology of the rectum is rare In the child, it gathers various isolated or multiple lesions whose histological nature conditions the prognosis.

It is usually benign tumors, cancers are exceptional [1].

For malignant tumors, adenocarcinoma is the most common. Burkit's lymphoma is exceptional, only a few cases have been reported in the last 30 years, there are only 10 cases found in the literature.

We present the case of a 15-year-old child who presented Burkitt lymphoma of rectal localization, complicated by a recto-vesical fistula. This is an exceptional complication of this rectal location of Burkitt's lymphoma.

2. Observation

A 15-year-old boy with constipation and rectorrhagia associated with rectal syndrome as initial symptoms. This symptomatology became complicated 7 days before its consultation of an occlusive syndrome makes diffuse abdominal pains, stopping of materials and gases and abdominal meteorism. In addition, the patient reported acute retention of urine and bone pain in the left thigh and lumbar spine, all in a context of slimming and deterioration of the general condition.

The concept of rectal bleeding and the rectal syndrome led to the realization of a touch rectal, which revealed a voluminous tumoral process 1 cm from the sessile anal margin, budding and bleeding on contact.

An initial assessment with an untreated Abdomen (Figure 1) confirmed intestinal obstruction, a blood count, an ionogram, and an abdominal pelvic ultrasound noted the presence of irregular hypoechoic rectal thickening measuring 18 mm extended to 52 mm.

Computed tomography (CT) showed a large parietal tumoral process extending from the recto-sigmoidal hinge to the anal margin, with heterogeneous tissue density containing hypodense areas of necrosis and calcification, reaching forward to the posterior wall of the bladder without border of separation (figures 2,3 and 4).

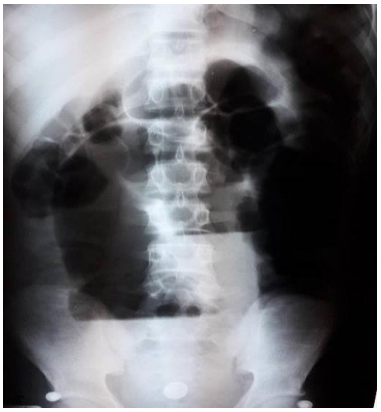

Fig 1: X-ray abdomen without preparation showing colonic hydro-aeric levels

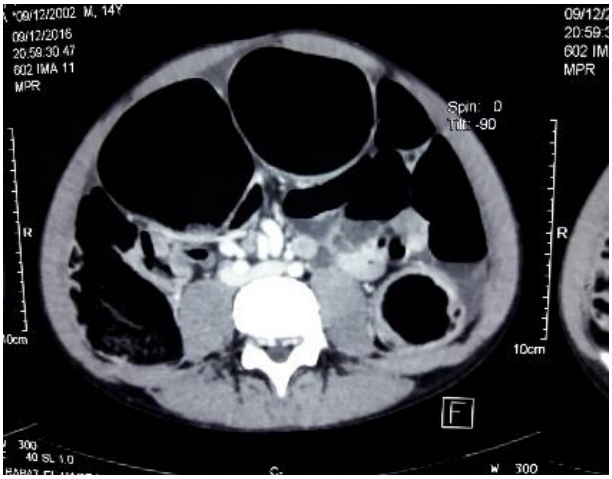

Fig 2: CT scan showing colonic and pelvic distention.

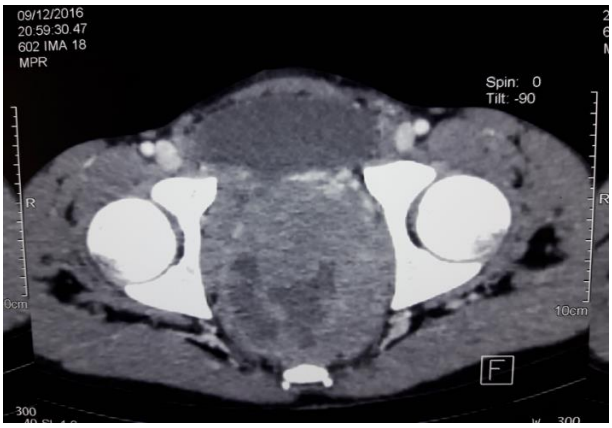

Fig 3: Bulky parietal tumoral process of the rectum, circumferential, II stenosis rectal lumen, heterogeneous tissue density enclosing hypodense areas of necrosis and calcifications to abdominopelvic computed tomography.

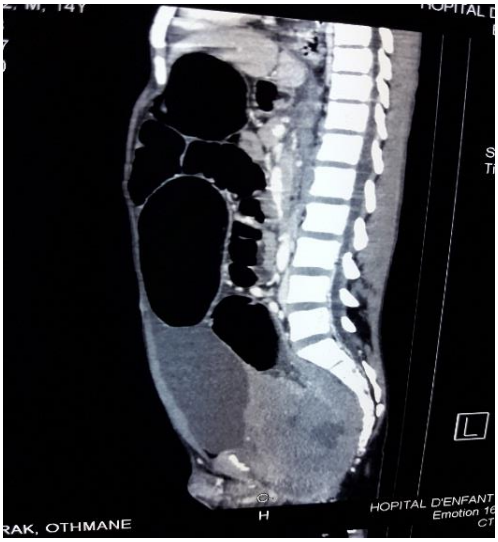

Fig 4: CT appearance showing a large tumoral process of the extended rectum of the rectosigmoid hinge to the margin leading forward to the posterior wall of the bladder without a border of separation.

In front of the occlusive rectal tumor, the indication of a digestive stoma is raised urgently, the patient has benefited from a sigmoidostomy with transanal mass biopsy.

Pathological examination confirmed the diagnosis of Burkitt's lymphoma; with immunohistochemistry, a positive positivity of CD20, CD10 and KI67 (nuclear labeling of 100% of tumor cells), CD3 negative and cytokeratin (Figure 5).

The extension assessment did not show any invasion of the cerebrospinal fluid (CSF), but on the other hand a medullary extension with presence of 66% of Burkitt type blasts on the smear of the two crests. The diagnosis of Burkitt stage 4 lymphoma of Murphy was retained.

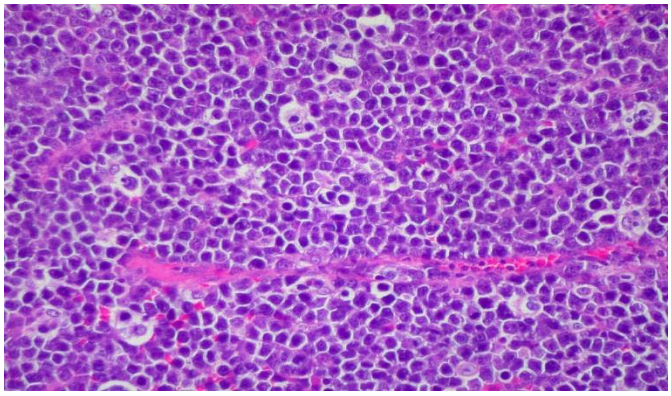

Fig 5: Typical cytologic appearance of Burkitt's lymphoma with vacuolated appearance of the cytoplasm on histopathological examination of the biopsy specimen.

Chemotherapy was started according to the protocol of GFAOP (French-African group of pediatric oncology).

After two courses of chemotherapy (COP and COPADM1), the evolution is marked by the disappearance of Burkitt cells on the control medullogram, and the decrease of the tumor volume by 70% after treatment with COPADEM 2 and CYVE 1 (Figure 6).

The patient reported afterwards, the passage of urine through the stoma: a recto-vesical fistula was confirmed by colonography (Figure 7), which led to the completion of a vesicostomy in order to stop the feeding of the fistula time to manage its tumor problem.

At the follow-up of four months, the patient is still on chemotherapy and the control CT (Figure 7) has shown a significant decrease in rectal wall thickness, which is more marked at the right lateral rectal level with persistence of the vesical fistula. rectal 11 mm in diameter. We plan to close this fistula after the end of the ongoing curative chemotherapy protocol.

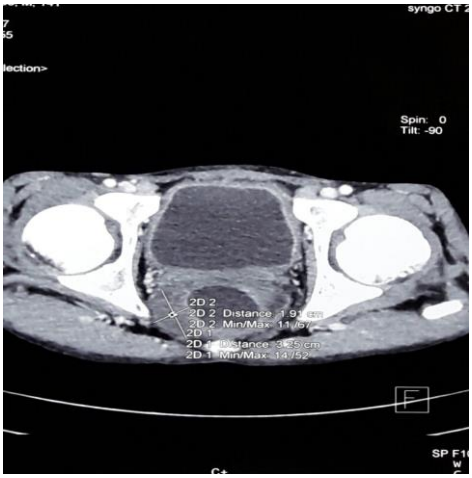

Fig 6: CT scan showing a significant decrease in the rectal tumor process, more marked at the right parietal level.

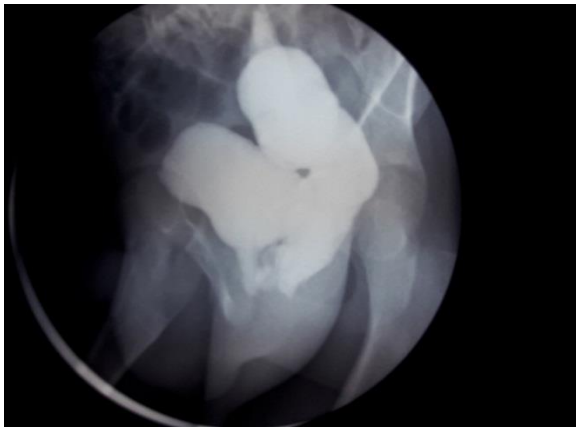

Fig 7: Colonography showing the presence of recto-vesical fistula, exceptional complication encountered in our patient

3. Discussion

Among rare rectal tumors in children, malignant tumors are mainly represented by adenocarcinoma. Primitive Burkitt lymphomas are exceptional. There are only 10 cases found in the literature.

These are subjects from 2 to 15 years old. Clinical signs are variable to type of abdominal pain, gastrointestinal bleeding or vomiting [1].

Sometimes the tumor is revealed by weight loss with an alteration of the general condition. The clinical history is generally rapid from a few days to a few weeks and a revelation in a more acute mode can be seen: intestinal obstruction table by tumor compression, hemorrhage or more rarely digestive perforation [2].

The child we followed consulted with pediatric surgical emergencies for an acute bowel obstruction, with diffuse abdominal pain, stopping of material and gas, and abdominal meteorism. Other clinical signs are constipation, acute retention of urine, weight loss and general deterioration of the condition.

The main element of the clinical diagnosis is the rectorragies, which must lead to the realization of digital rectal examination. This has an informative role on the characteristics of the tumor: the aspect (polypoid), the localization with respect to the anal margin and the infiltration of the neighboring organs, evocative of the clinical diagnosis of the tumoral mass [1].

The rectal examination in our patient revealed a bulky tumor 1 cm from the anal margin sessile, budding and bleeding on contact.

Ultrasound can guide the diagnosis; Tumoral infiltration of the digestive wall results in very hypoechoic transmural thickening that can be multi-nodular asymmetric or not [2].

The abdominopelvic computed tomography (CT) provides the same information as the ultrasound, the tumor mass has a tissue density with a heterogeneous enhancement. It also informs about the relations of the tumoral process with ganglionic attacks [2].

In our patient, the CT noted the presence of a bulky parietal tumoral process of the rectum, circumferential, extended from the rectosigmoidal hinge to the anal margin, II stenosis of the rectal lumen and leads to an important colonic and grelic distention. upstream with laminated aspect of the digestive wall (thickened) which enhances itself. This process is of heterogeneous tissue density containing hypodense zones (necrosis) and micro calcifications, heterogeneously enhanced after contrast, measuring 16.5 * 10 * 9.4 cm, extending to mesorectal fat, and surrounded by a collateral circulation.

The confirmation of the diagnosis is based on the biopsy of the tumor mass with histopathological and immunohistochemical study [3].

Burkitt lymphoma cells have a high nucleo-cytoplasmic ratio. The nucleus is round or oval with an open chromatin appearance. This nucleus contains several nucleoli (2 to 5). The mitotic index is high. The cytoplasm is basophilic with many vacuoles; Macrophages containing cellular debris are interposed between these monomorphic cells producing the appearance of "starry sky" (Figure 4) [4]. The Burkitt lymphoma cells express on their surface immunoglobulins which are IgM in 90% of cases. They also express CD19, CD20, CD22, CD79a, CD77 [3].

For our patient, there is a clear positivity of CD20, CD10 and ki 67 (nuclear staining of 100% of tumor cells), CD3 negative as well as cytokeratin.

The high aggressiveness of lymphomas requires rapid diagnostic and extension assessment [4].

The systematically sought locations that guide the treatment are neuroméningées by the lumbar puncture and / or MRI in case of suspicion of epiduritis and medullaries by the myelogram and the medullary biopsy [4].

Thorax radiography is systematic and can be used to diagnose mediastinal and pleuropulmonary disorders by visualizing a tracheal mass, mediastinal adenopathies, pleural effusion or pericardial effusion [2].

Ultrasound and computed tomography allow to inform on the secondary localizations, and the report of the tumoral process with ganglionic attacks [2].

The treatment of Burkitt's lymphoma is based on intensive polychemotherapeutic combinations and not on monotherapy [4].

Several classical associations are described and developed by different groups and seem to give comparable sustainable response and remission rates.

Although the therapeutic management of Burkitt's lymphoma is essentially based on chemotherapy, surgery has a significant role; it is indicated for surgical emergencies such as occlusion, perforation and hemorrhage. This is the case of our patient who presented an intestinal obstruction chart requiring emergency digestive stoma [5].

Surgery can also be used for diagnostic biopsy and to manage the complications of chemotherapy [6].

Our patient presented, after 2 courses of chemotherapy, a rectovesical fistula leading to the realization of a vesicostomy in order to stop the feeding of the fistula.

To our knowledge, our case is the first reported association of a fistula to Burkitt lymphoma of rectal localization, probably this fistula may be due to invasion of the posterior wall of the bladder by the tumor process. We expect the spontaneous closure of this fistula. However, surgical time is expected if persistence of the fistula after completion of the chemotherapy protocol.

Surgery is also discussed if persistence of the tumor after treatment of chemotherapy, may be indicated in the rare cases where complete resection is possible with limited morbidity provided that it is performed without mutilation and similarly in case of tumor persistence before consolidation by high-dose chemotherapy 'prognostic impact phase'.

4. Conclusion

Rectal tumors are rare in children, they must be suspected before any rectal bleeding with interest of digital rectal examination [2].

Although the treatment is mainly based on chemotherapy; we note through this observation the primordial diagnostic and therapeutic role of surgery in Burkitt's lymphoma of rectal localization.

5. Declaration of interest

The authors declare that they have no conflicts of interest in relation to this article.

7. Contributions of the authors

All authors contributed to the writing of this manuscript, all read and approved the final version.

8. References

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