

Burkitt's Lymphoma in the Boy: Infiltration in the Stomach, Colon and the Retroperitoneum – Ileocecal Invagination

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ABSTRACT

A 4-year-old boy was hospitalised because showing signs of weakness, slight pain in the abdomen and while urinating. The symptoms occurred 7 days before hospitalisation. The boy did not vomit, nor did he have the urge to vomit, the defecation was regular showing no traces of blood. The physical visit a soft and painless tumefaction was confirmed ileocecally. The echography tests and the computed tomography suggested invagination, not excluding the second substrate. Barium enema showed irreducible invagination. The operative test showed that it was about the ileocolic invagination with extreme thickening of the cecum, the ascendant colon, the intestine and the retroperitoneum walls. A resection of the small intestine and a ileocolic anastomosis was performed. The pathohistological test shows the primar abdominal Burkitt's lymphoma. In spite of the subsequent therapy the boy dies three weeks after the first symptoms' manifestation. We, herewith, suggest at the importance of the echography analysis when diagnosing the Burkitt's tumor and give advantage to this analysis against the computerized tomography. We also point at the huge level of malignancy of the Burkitt's tumor in this boy.

Key words: children, Burkitt's lymphoma, invagination, diagnosis, treatment

Introduction

In 70% of the cases the non-Hodgkin tumor lymphoma groups arouse through metaplasia and hyperplasia of the cells in lymphatic nodes, and in 30% in tissue organs¹. These 30% are called extranodal or primary lymphoma. In this group of extranodal lymphoma 40% originate in the abdomen^{2,3}, and especially in the stomach and the small intestine^{1,4}.

The Burkitt's lymphoma, which is highly malignant, belongs to this non-Hodgkin group of tumors^{5,6}, and has an acute flow similar to acute leukaemia. The tumor grows fast, and the reproduction of cells occurs within 24 hours⁷. If it is diagnosed early and the patient begins with chemotherapy, the tumor immediately retrogresses and a shorter or longer remission is followed. In most of the cases the diagnosis is given when the tumor has already developed, causing a frequency of recidives and a low surviving percentage^{5,8}. The incidence of this tumor,

as reported by Brichon and assistants⁹, in juvenile population is of 0.2 on 100.000 children.

In 1958 Burkitt described jaw tumor in a boy, which we today call the Burkitt's lymphoma². There are two types of the Burkitt's lymphoma: African and non-African (American) type. The African type is usually connected to the Epstein-Barr virus that is found in Burkitt's lymphoma tumor cells, whereas the same virus is much less present in the American type with only 25% of the patients suffering from this lymphoma type⁹. Both the American and the African type belong to the B-type cells lymphoma. The histopathologic characteristic of Burkitt's lymphoma may give rise to specific ultrasonographic findings with diffuse involvement of organs such as a »starry sky« appearance¹.

The abdominal Burkitt's lymphoma first manifests general symptoms: weakness, loss of appetite and loss of

weight. The ileus symptoms are also frequent in this case due to ileocolic^{9,11,12} or ileoileal invagination¹³. The tumor tissue grows extremely fast and the symptoms also develop quickly.

In diagnosing the Burkitt's tumor of the abdomen an echography analysis gives best results in diagnosing the tumor itself and its expansion¹⁴, as well as in diagnosing the invagination provoked by the lymphoma^{9,15}. It especially gives reliable diagnostical results when showing the intensity of the colon walls thickening seized by the lymphoma, which is manifested through the echographical sign, the so called »target» or »pseudokidney sign»^{5,16}. It is also important when diagnosing other extranodally located Burkitt's lymphoma¹⁶. According to some experts the echography has an advantage over the computerized tomography¹⁷. Others claim the advantage of the computerized tomography when diagnosing the Burkitt's stomach tumor giving a more precise insight of the walls and their thickening, which can grow up to 3-5 cm in children and adolescents, delineating the stomach columnas and the lymphedema of the submucosis and the mucosis².

The Burkitt's lymphoma is sensitive to chemotherapy and it retrogresses very fast when subjected to initial citostatic therapy, but the percentage of relapse and deadly outcome is very high⁷.

Case Report

A four-year-old boy was admitted to the Mostar University Hospital feeling weak, slightly pained in the stomach, which was localized more on the right, as well as when urinating. The symptoms had occurred 7 days before the boy came to hospital. The medical physical examination showed that the abdomen was slightly distended and no pain was detected, but through an ileocecal inspection a painless soft tumefaction of the size of a child's fist was palpable. The boy does not vomit, nor is he sick, the defecation is regular with no bloody traces.

The echography examination of the abdomen shows a tumefaction in the ileocecal area. As the most probable diagnosis is ileocecal invagination, it also shows the possibility of a solid tumor (neuroblastom). The symptoma-

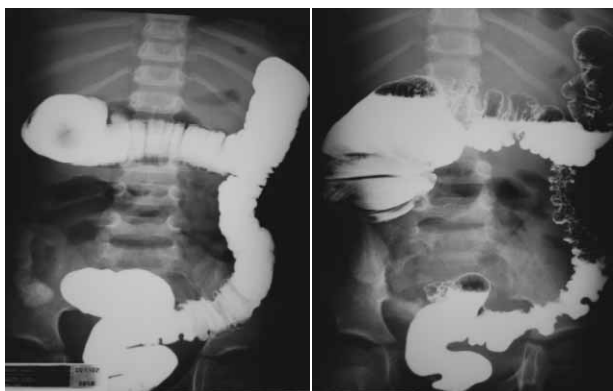


Fig. 1 Radiography with barium enema.

tology during the first day of the boy's permanence in the hospital is the same as when admitted: the boy shows no sharp abdominal colics, which, together with the blood tests, shows invagination; the abdomen is still slightly distended, soft and painless, the boy does not vomit and the defecation is regular after a Dulcolax suppository, showing no traces of blood.

On the second day in hospital the symptoms are almost the same or slightly intensified. The boy has regular defecation after a Dulcolax suppository, with no bloody traces, does not vomit, the abdomen is more distended, and also slightly tense, still painless. Due to the more swollen abdomen a nasogastric suction was ordained, which showed some gastric juice with blood traces. On the same day the echography examination was repeated and again the diagnosis was invagination, which was also confirmed by the computerized tomography. Barium enema showed that it was the irreducible invagination (Figure 1). The picture of the intravenous pyelography shows stasis (Figure 2).



Fig. 2. Intravenous pyelography.

A laparotomy was prescribed and done. By exploring the abdomen an ileocecolic invagination was found presenting a thickened ileum and extremely swollen cecum, as well as the ascendental colon (thickness 1-1.5 cm). The whole conglomerate was similar to a big soft tumefaction. The stomach is very swollen, presenting soft walls, the retroperitoneum being like a thick slab up to the pelvis, and in the Douglas area a larger ascites. A resection is performed of the invagination affected by ileocecolic conglomeration and the ileocolic anastomosis. The pathohistological results of the preparation are shown in Figure 3. A »starry sky» phenomenon is seen. The pathohistological diagnosis of the Burkitt's lymphoma is established. An early postoperational recovery started successfully and continued successfully for some time.

After a laparotomy and the resection of the intestine segment recovery was successful, but the ninth day after the operation abdominal symptoms and signs of ileus were present. A relaparotomy is performed, the appendy-

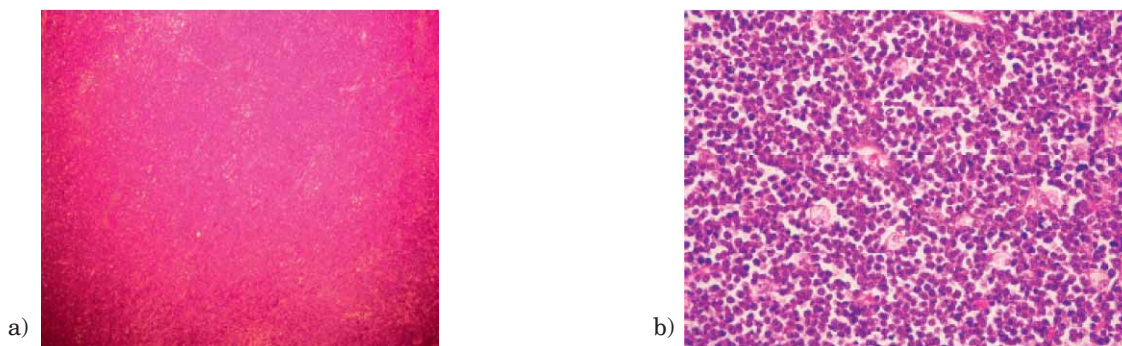


Fig. 3. The pathohistological results: a) of the resected bowels' conglomerate (»starry sky« phenomenon) and b) of the resected omentum segment (»starry sky« phenomenon)

xes were cut and a continuity of the intestine is achieved. A resection is also performed over a part of a very swollen and strangled omentum. The stomach, intestine and the retroperitoneum tests showed that these segments of the digestive apparatus were extremely swollen and hard compared to the first laparotomy, the omentum being far thicker and harder than earlier so that the enlarged mesenteric lymphatic nodes could be identified only by palpation. The pathohistological test of the resected omentum segment is shown in Figure 3, which clearly shows the »starry sky« phenomenon. In this case the Burkitt's lymphoma was also diagnosed. Reanimation and postoperative recovery are as successful as after the first laparotomy. Notwithstanding the adequately started therapy the boy deceased after the 14th postoperative day.

Discussion

Amongst the non-Hodgkin lymphomas, the Burkitt's abdominal lymphoma proved to be the most malignant one. According to the course of the illness and the tests of our patient, the solid tumor's proliferation of malignant cells exceeds in intensity all other solid malignant tumors in children. In our case, we discovered only by palpation just within 48 hours, i.e. starting with the boy's hospitalization till the decision for a laparotomy, an extreme enlargement of the ileocecal tumor mass, which suggests an extremely fast reproduction of cells⁷ and the Burkitt's lymphoma.

The symptomatological period in the boy was extremely short. Only 7 days before he was hospitalized, the boy started feeling sick and weak, slightly pained in the abdomen located more on the right, accompanied with difficulties when urinating. After being hospitalized he had regular defecation with no blood traces. The boy did not feel sick, nor did he vomit. The abdominal pains did not have a colic character that would point to the possible invagination. A physical examination showed a painless soft ileocecal tumefaction. With such symptomatology and physical test it was thought at first that the patient was suffering from a perforated chronic retrocecal appendix (perforatio chronica tecta) due to such symptoms and tests. In any of these moments the patient

did not show signs of being fatally ill, nor was he feverish.

As soon as the patient was hospitalized an echography visit of the abdomen was performed. The test was, among others, pointing at the invagination as the most likely diagnosis. Figure 1 shows not so typically marked »target sign« and the »pseudokidney sign«. The computerized tomography also pointed to invagination as the most probable diagnosis. Notwithstanding these tests that pointed to the invagination as the most probable diagnosis, the clinical picture of the patient was not matching the tests at all and therefore laparotomy was not prescribed straight away. The boy was under surveillance after the second day in hospital.

During the second day of the boy's stay in hospital it was discovered by palpation that the ileocecal tumefaction was visibly larger than the day before, and the symptomatology did not get worse: the child does not vomit, abdomen is slightly distended, the defecation is regular after a Dulcolax suppository showing no traces of blood. Everything still discretely announced a progressive course of symptoms and physical tests that were showing subileus marks, but the symptomatology did not point to invagination as its background.

On the second day of the boy's permanence in the clinical hospital an echography analysis confirmed that, in spite of the mild clinical course of the illness, it was invagination after all.

In order to confirm the diagnosis and a possible reduction, a barium enema was performed which confirmed that complete hydrostatic reduction has not been achieved.

The laparotomy found a massive ileocecal invagination with extreme thickening of the cecum, colon ascendens and a slightly smaller thickening of the terminal ileum. A similarly large thickening, but softer, of the stomach was confirmed, as well as of the parietal peritoneum and retroperitoneum that was expanding like a thick slab into all parts of the abdomen until the bottom of the pelvis filled with abundant yellowish liquid (ascites). The bibliography suggests that patients affected by the Burkitt's lymphoma have first affected the stomach, then the small intestine, and just then the colon¹⁷ by

tumor cells. This is one of the rare examples where the Burkitt's lymphoma is most evident in the ileocecal segment.

A huge discrepancy was established in this patient between the operative test regarding small intestine obstruction by ileocecal invagination and evident symptomatology of the ileus. Moreover, an extremely fast growth of the tumor mass is evidenced, which expanded so evidently in 48 hours that it was possible to feel it by touch. This shows an extremely fast proliferation of the cells and a huge malignancy of the Burkitt's lymphoma⁷. All of it also suggests a lethal outcome of the illness within three weeks of the first symptoms.

Analyzing the results of the diagnosed tests we can confirm like others had done before⁵ that an echography analysis offers the most indicative data about the small

intestine infiltration with the Burkitt's lymphoma and a possible invagination, which is confirmed by our tests »pseudokidney sign» in both of our analysis. According to our modest experience when diagnosing the Burkitt's abdominal lymphoma we would, like some other authors, give advantage to the echography analysis rather than a computed tomography, whereas in diagnosis the gastric lymphoma the advantage would be on computed tomography².

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REFERENCES

1. LIOU JM, WANG HP, KO BS, CHENG TY, WU MS, LIN JT, J Gastroenterol Hepatol, 20 (2005) 1616. — 2. REGENBOGEN VS, GHAREMANI GG, ZABROWSKI RJ, ROCHESTER D, J Computed Tomography, 8 (1984) 273. — 3. SHARMA A, RAINA V, GUJRAL S, KUMAR R, TANDON R, JAIN P, Am J Hematol, 67 (2001) 48. — 4. BRADY LW, ASBELL SO, Radiol, 137 (1980) 291. — 5. ASAI S, MIYACHI H, HARA M, FUKAGAWA S, SHIMAMURA K, ANDO Y, J Ultrasound Med, 21 (2002) 657. — 6. BETSCH B, BRADO M, WUNSCH C, RICHTER GM, KAUFMANN GW, TRÖGER J, Radiologe, 37 (1997) 62. — 7. TIEFENBACH A, ZUPANČIĆ-ŠALEK S, ZERGOLLERN Lj. In: ZERGOLLERN Lj, REINER-BANOVAČ Ž, BARIŠIĆ I, RICHTER D, VOTAVA-RAIČ A. Pedijatrija 2, (Naprijed, Zagreb, 1994). — 8. BRAZIEL RM, ARBER DA, SLOVAK ML et

al., Blood, 15 (2001) 3713. — 9. BRICHON P, BERTRAND Y, PLANTAZ D, Ann Chir, 126 (2001) 649. — 10. RAK CK, HOPPER KD, PARKER MSH, J Ultrasound Med, 7 (1988) 279. — 11. SZABO A, FARKAS P, LASZLO A, BERCZI L, SPERLAGHI M, SUGAR I, Magyar Sebeszet, 56 (2003) 116. — 12. VERNOOY JE, BOS AP, TIBBOEL D, HAZEBROEK FW, Nederl Tijdschrift Geneeskunde, 136 (1992) 1489. — 13. CAPELLE J, LECLERE J, KRAIEM A, J Radiol, 65 (1984) 471. — 14. GOERG C, SCHWERK WB, GOERG K, AJR, 155 (1990) 795. — 15. LAM AJ, FIRMAN K, Australas Radiol, 35 (1991) 343. — 16. STUCKMANN G, ZOLLIKOFER C, Schweiz Med Wochenschr, 126 (1996) 813. — 17. DANEMAN A, ALTON DJ, LOBO E, GRAVETT J, KIM P, EIN SH, Pediatr Radiol, 28 (1998) 913.

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BURKITTOV LIMFOM U DJEČAKA: INFILTRACIJA ŽELUCA, KOLONA I RETROPERITONEUMA – ILEOCEKALNA INVAGINACIJA

SAŽETAK

Četverogodišnji dečak je hospitaliziran zbog slabosti, bolova u abdomenu i pri mokrenju. Simptomi su se prvi put očitovali sedam dana prije prijema dječaka u bolnicu. Nije povraćao niti je imao nagon na povraćanje, a stolica je bila uredna i bez tragova krvi. Fizikalnim je pregledom utvrđena ileocekalna mekana i lagano bolna tumefakcija. Ultrasonografski nalaz i nalaz kompjutorizirane tomografije ukazuju na invaginaciju, a i nalaz irigografije ukazuje isto tako na ireduktibilnu ileocekokoličnu invaginaciju. Operativna je eksploracija abdomena potvrdila ileokoličnu invaginaciju s ekstremnim zadebljanjem stijenke terminalnog ileuma, cekuma i ascendentnog kolona, a u manjem opsegu i ostalih crijevnih vijuga uz jako zadebljanje pripadnog retroperitoneuma. U bloku je resecirani terminalni ileum, cekum i ascendentni kolon. Patohistološkom pretragom preparata postavljena je dijagnoza Burkittovog limfoma. Uprkos poduzete odgovarajuće terapije dječak je tri tjedna nakon pojave prvih simptoma preminuo. Ukazujemo na vrijednost ultrazvučne pretrage u dijagnostici Burkittovog limfoma. Isto tako ukazujemo i na vrlo visoki stupanj malignosti ovog tumora u dječaka.