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Sphincter Disturbance as the Initial Presentation of Acute Lymphoblastic Leukemia：A Case Report

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Acute leukemia is the most common malignancy involving the central nervous system (CNS) among childhood. Neurological manifestations of leukemia frequently occur during the course of acute leukemia. However, it is rarely an isolated feature as the initial presentation of leukemia. The spinal cord and spinal roots are less frequently involved than other CNS regions. It is very rare to have neurologic involvement of conus medullaris as an initial presentation of acute lymphoblastic leukemia (ALL).

This case was a 16 year-old boy who suffered from sever back pain and disturbance of sphincter function before the diagnosis of ALL. The diagnosis of acute lymphoblastic leukemia (L3, Burkitt's type) with leptomeningeal involvement was confirmed based on the bone marrow and cerebral spinal fluid examinations.

Now that management of leukemia is possible and even is successful with chemotherapy and radiotherapy, it is more important to recognize and alleviate any neurologic morbidity of these patients.

We presented this case with an attempt to cover the pathogenesis, differential diagnosis, treatment of ALL involving spinal cord and to call the attention to such case. (J Rehab Med Assoc ROC 1999; 27(1): 31 – 36)

Key words: sphincter disturbance, initial presentation, acute lymphoblastic leukemia

INTRODUCTION

Systemic malignancies of childhood that arise from structures outside the nervous system may frequently cause neurological complications. Generally, neurological involvement occurs during the course of the malignancies. It is not difficult to find the neurological disorders of patients with previously diagnosed malignancies. However, it may be difficult to have early diagnosis of malignancy when they have neurologic problems as the initial presenting symptoms. In such cases, definite diagnosis may be established after deliberate clinical and laboratory investigation [1].

In this case report, we present a 16 year-old boy of acute lymphoblastic leukemia (ALL) with initial

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symptoms of low back pain, constipation and urinary retention due to a conus medullaris lesion. By recognizing this rare situation, one should pay attention to the unusual neurological symptoms to avoid the delay in diagnosis of malignancy and to provide appropriate treatment.

CASE REPORT

This 16-year-old male patient was a healthy boy till July 1997, when he began to suffer from low back pain. At the beginning, he didn't pay much attention to it, and thought that it was a result of sports injury. Unfortunately the pain got worse progressively and it was also radiated to both thighs when bending forward. Later, the pain spread out to the perineum and scrotum. However, the muscle strength of both lower limbs was still normal at that time. Gradually, he also had difficulty in defecation and urination.

In early August of 1997, he visited our outpatient clinic for the problems of severe low back pain, constipation and urine retention. The initial neurologic examination was unremarkable. The plain films of lumbar spine showed evidence of L5 pars defect, which was considered to be non-contributory to the presenting neurologic findings. The whole spinal magnetic resonance image without enhancement only revealed mild intervertebral disc bulging from C3 to C6.

Radiological study of lower GI series was arranged to assess the severe constipation. No tumor or mechanical obstruction, but excessive feces was found. The abdomen CT revealed evidences of enlarged paraaortic lymph nodes, mild hydronephrosis, and hydroureter. The findings in EMG study indicated multiple L-S radiculopathy (L5-S2). The urodynamic study showed a dyssynergic bladder with poor uroflow and intermittent flow pattern.

The muscle strength of bilateral legs was gradually decreased since mid-August. He also had difficulty in walking. The function of voiding and stool passage also became worse gradually, and he required Foley catheter and enema for such problems in the late August. He was admitted to our hospital at that time due to the complication of urinary tract infection.

Physical examination revealed a pale, thin, quiet boy with acute ill looking. No hepatomegaly, splenomegaly or lymphadenopathy was found. The abdomen was soft, and the bowel sounds were normoactive. There were diffuse tenderness and knocking pain over the lumbosacral area. Neurologic examination revealed intact cranical nerves, normal muscle strength and sensation of bilateral upper extremities, but weakness of both lower extremities (against mild resistance) with intact sensation. Deep tendon reflexes of bilateral knee jerks were normal but bilateral ankle jerks were absent. No Babinski sign was noted and the bulbocavernous reflex was normal. The anal sphincter tone was decreased, and the rectal ampulla was full of stool.

Initial hematologic study showed evidence of only mild anemia, but blast cells (table 1) were found on the follow-up data.

Bone marrow examination was performed on November 11, 1997 and revealed hypercellularity with predominant blast cells (Burkitt's type). Spinal fluid study was also performed, and it revealed many abnormal lymphoid cells.

Based on the above data, the diagnosis of ALL with leptomenigeal involvement was established.

Intensive treatment with systemic chemotherapy and intrathecal chemotherapy plus craniospinal radiotherapy were given as soon as the diagnosis was confirmed. The neurological symptoms and signs had been much improved after treatment. All neurological symptoms and signs were almost completely disappeared within 2 weeks. The hematologic laboratory data had been improved (table 1), and the bone morrow and CSF findings also became normal after treatment.

DISCUSSION

Leukemia is the most common childhood malignancy. Because of advances in the treatment and the prolonged survival, there has been an increase in the incidence of neurological complication in leukemia [2-4]

Clinically, significant spinal cord involvement is unusual in leukemia [2,5-7], although intracranial (meningeal) involvement is common. Even among patients with meningeal leukemia, only 5-10% of cases develop clinically detectable lesion in the spinal cord. Leukemia has been estimated to account for

approximately 3% of all spinal cord tumors [5].

It is even more rare to have spinal cord involvement as the initial presentation of ALL. Petursson and Boggs (1981) reported a case of Ph+ acute myeloid leukemia (AML) with an initial presentation of conus medullaris syndrome. By reviewing literatures, they concluded that spinal cord involvement may be more frequent in AML than ALL, and it is most common in the second decade of life [5]. Pui et al (1984) reported 4 (1.4%) of 276 children with non-Hodgkin lymphoma (NHL) and 7 (0.4%) of 1721 with acute leukemia had symptoms due to epidural spinal cord compression as the initial manifestation. Most cases had involvement at the thoracic level, but no conus medullaris syndrome had been found [8]. Lewis et al. (1986) reported spinal cord compression occurred as the presenting sign of malignancy in 6 children (4 sarcomas and 2 lymphomas) [9]. Spiegelmann et al. (1988) reported a case of spinal cord involvement as the presenting syndrome of acute monocytic leukemia [7]. Asyun et al. (1994) have found that acute leukemia is the first, and neuroblastoma is the second, most common malignancy among childhood systemic malignancies presenting with neurologic involvement. However, a majority of those patients with acute leukemia (8/11) had features referable to the nervous system due to anemia or bone pain with no real nervous system involvement. None of the leukemia patients had spinal cord compression in previous reports [1]. Similar findings were reported by Huang et al. in 1995. They studied the occurrence of neurologic complications of 200 pediatric patients with systemic malignancies. ALL was also the most common malignancy with neurologic features at the initial presentation, but none of them presented with spinal cord compression [4]. In our case, the patient presented with a conus medullaris syndrome, with low back pain, fecal and urinary retention and loss of ankle DTR; it is a rather uncommon initial presentation of ALL.

Leptomeningeal metastasis is an important complication of CNS leukemia [10]. Although it is a subject of controversy, it has been generally accepted that leukemic cells may invade the meninges via the arachnoid veins, with subsequent involvement of the CSF spaces. These cells may also penetrate into the deep Virchow-Robin spaces to cause disruption of the pia-glial membrane, and to invade the adjacent parenchyma [3,11,12]. Patients with neurologic problems as the only initial symptoms of acute leukemia may perplex physicians. Complete and careful physical examination to find out

Table 1. CBC data during hospitalization

Date	8/25	8/28	9/1	9/4	9/5	9/8	*9/15	*9/26	*10/23
RBC (million/cmm)	4.24	4.27	4.07	3.98	3.81	3.34	3.04	3.12	3.15
Hb (g/dl)	11.1	11.1	10.5	10.3	9.9	8.4	8.0	8.5	8.6
Hct (%)	31.9	31.9	31.5	30.3	28.8	25.1	23.6	26.2	27.8
WBC (/cmm)	8000	12400	27400	24000	17500	12000	400	1800	6500
Seg (%)	72	65	55	51	71	42	34	63	88.7
Band (%)	0	0	16	11	6	3	0	0	0
Lymph (%)	20	19	17.5	20.5	4	24	64	32.1	3.9
Mono (%)	6	10	5	11	1	12	0	2.2	5.2
Eos (%)	2	2	1	1	2	3	2.4	1.6	1.7
Baso (%)	0	0	0	0.5	0	0	2.4	1.1	0.5
Blast (%)	0	3	0	4	10	13	0	0	0
Myelo (%)	0	1	4	1.5	2	3	0	0	0
Metamyelo (%)	0	0	1.5	0	4	0	0	0	0
Platelet (10000/cmm)	28	25.4	15.7	8.3	6.5	7.9	4.0	8.6	12.3

* after treatment

suggestive signs of leukemia, such as pallor, petechiae, ecchymosis, organomegaly or lymphadenopathy [1], and laboratory tests including CBC with differential count can provide important clues to detect acute leukemia in the early stage. In our case, initial CBC data (table 1) showed only mild anemia. Although blast cells were identified on the following CBC examination, it is an obvious initial sign of marrow infiltrative disease, especially acute leukemia. Bone marrow examination should be done and adequate treatment should be initiated as soon as possible. We emphasize that series of follow-up are important for any abnormal data, even only mild abnormality initially.

The clinical signs and symptoms of spinal cord compression of systemic malignancy depend on the level invaded. The most common presenting symptom is pain, usually confined to back but often becoming radicular, followed by weakness, autonomic dysfunction, sensory loss, etc. [8,9,13]. The initial presentation of this patient is the syndrome of conus medullaris, which is less frequently reported than proximal spinal cord or cauda equina in the cases with spinal cord compression [9].

A conus medullaris syndrome is due to the damage of the lower sacral segments of the spinal cord and is very similar to the cauda equina syndrome. These two syndromes can be distinguished clinically in that weakness and paralysis of the lower extremities precede loss of sphincter tone in the latter, but paraparesis or paraplegia follows loss of sphincter tone in the former, if it occurs at all [5].

Examination of CSF is the most important diagnostic study to identify the evidence of leptomeningeal metastases. In the definition of CNS leukemia used by the major pediatric groups, it has been traditionally specified that at least five leukocytes per microliter of CSF and the unequivocal presence of leukemic blast cell in a cytospin specimen must be present [14]. Malignant cells in the CSF can be identified in approximately 90% of patients, and multiple samples of CSF are often required [11].

Myelography may be required in some patients who have spinal root or spinal cord signs and symptoms. Contrasted MRI of the spine may uniquely visualize subarachoid disease in some patients, but unenhanced spinal MRI is less sensitive. This case received unenhanced spinal MRI study, but no special abnormal finding was found.

The standard treatment for overt CNS leukemia includes intrathecal chemotherapy, systemic chemotherapy and cranial or craniospinal irradiation [14]. Results of treatment have been generally good [8,14].

CONCLUSION

Neurologic manifestation of the spinal cord involvement as the initial presentation of ALL is uncommon. Since cure is possible, and even can be expected, it has become more important to recognize these patients for early treatment to improve the prognosis.

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以括約肌失調爲起始表現之急性淋巴球性白血病：病例報告

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根據研究，發生在兒童之系統性惡性腫瘤中，最易侵犯神經系統的是急性淋巴球白血病。但急性淋巴球白血病以侵犯腦部及軟腦膜爲主，較少影響脊髓及神經根，且中樞神經系統症狀的出現多發生在診斷出白血病的同時或以後之治療過程中，很少以此爲起始表現。而以侵犯脊髓圓錐及其附近之神經根爲起始之表現，則更少見。

本例是一 16 歲男孩，過去沒有白血病病史，於八十六年七、八月間出現嚴重背痛、生殖器處疼痛及括約肌失調的現象，最後合併雙下肢肌力漸減至步行困難。經一系列的檢查，最後由骨髓及腦脊髓液檢查証實爲急性淋巴球性白血病併腦脊髓膜侵犯，以臨床症狀、理學檢查、肌電圖及尿路動力學各項結果推論，其起始之表現應是影響到脊髓圓錐及其附近神經根的結果。這在急性淋巴球白血病是較少見的。

由於此病經化學治療及放射治療反應良好，早期發現可減少其神經系統方面的後遺症，如本例在接受治療後兩週其症狀有十分明顯的改善。現提出此病例，並討論其侵犯中樞神經系統的路徑、鑑別診斷、治療方法等，以供大家參考。（中華復健醫誌 1999; 27(1): 31 - 36）

關鍵詞：括約肌失調 (sphincter disturbance)，起始表現 (initial presentation)，
急性淋巴球性白血病 (acute lymphoblastic leukemia)

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