

Case Report

B-cell Lymphoma, Unclassifiable, with Features Intermediate between Diffuse Large B-cell Lymphoma and Burkitt Lymphoma of Abdominal Wall with Invasion to Colon

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Soft tissue lymphoma is a very rare clinical entity with varying presentation characteristics and atypical clinical and imaging features. Besides, B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma is a new category of malignancy of B-cell lymphoma according to the 4th edition of the World Health Organization Classification of Tumours of Haematopoietic and Lymphoid Tissues (2008). We present here a case of this tumor located in the abdominal wall with invasion to colon, treated by means of surgical intervention and chemotherapy.

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Key Words

Soft tissue lymphoma;
Abdominal wall mass;
Colonic invasion

The clinical presentation of lymphoma arising in soft tissue is extremely rare and variable.¹ It may present in almost any part of the human body and mimic other soft tissue malignancies leading to diagnostic misconceptions and often to suboptimal management.^{2,3}

B-cell lymphomas with features intermediate be-

tween diffuse large B-cell lymphoma (DLBCL) and Burkitt lymphoma (BL) are aggressive lymphomas that have morphological and genetic features of DLBCL and BL, but for biological and clinical reasons should not be included in these categories.⁴

The cellular morphology is variable. The morphological characteristics are intermediate between

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DLBCL and BL. In some cells that are smaller than typical DLBCL, resembling BL (a high proliferation rate—many mitotic figures, starry-sky pattern—impacted by numerous benign macrophages that have ingested apoptotic tumor cells), and some cells that are larger than typical BL, resembling DLBCL (diffuse large lymphocyte with irregular nuclei).^{4,5}

The immunophenotypic features resemble B-cell markers (e.g.: CD19, CD20, CD22 and CD79) and BL (e.g.: CD10+, BCL6+, BCL2-, Ki67 labeling index is usually high).⁶⁻⁹

Case Report

A 60-year-old woman with history of hypertension in the past suffered from intermittent left abdominal pain with duration of about thirty minutes every time. However, she had neither fever nor night sweats. At first, she visited Ping-Tung Christian Hospital where X-ray study was performed and reported the discovery of renal stone. Medication was prescribed. Abdominal pain seemed to relieve after medication. However, severe left abdominal pain occurred several days later. She was brought to Paochien Hospital and was transferred to our emergency room under the impression of perforated peptic ulcer.

At our ER, localized tenderness over the left abdomen without rebounding pain and no obviously palpable lymph nodes were noted on physical examination. The laboratory examination revealed a white cell count of $6,430/\text{mm}^3$. The contrast-enhanced computed tomography scan of abdomen was performed and revealed a mass lesion measuring about $9 \times 6 \times 6$ cm in size with homogenous enhancement, arising from left transverse abdominis muscle, abutting distal transverse colon, suspicious of desmoid tumor of left abdominal wall, with peritoneal seedings (Fig. 1).

After admission, the Division of General Surgery and the Division of Colorectal Surgery were consulted and performed en-block excision of the abdominal wall tumor, segmental resection of the distal transverse colon and end-to-end anastomosis. The operative findings are described as follows: one $9.2 \times 7.5 \times 7$ cm tumor over the left upper abdominal wall. Skin, subcutaneous fatty tissue, the fascia of Scarpa of external



Fig. 1. Computed tomography scan of abdomen revealed a mass lesion measuring about $9 \times 6 \times 6$ cm in size with homogenous enhancement (star), arising from left transverse abdominis muscle, abutting distal transverse colon (arrow).

oblique muscle, and rectus muscle were intact. The tumor arised from left internal oblique or transverse abdominal muscle. It grew into the abdominal cavity and caused desmoplastic reaction of surrounding soft tissue including greater omentum and distal transverse mesocolon. The left side transverse colon was severely adhered to the tumor itself. Serosal or possibly muscular layer of this segment of transverse colon was suspected to be invaded by the tumor (Fig. 2).

The histopathologic examination documented B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma. The sections of the specimen showed a picture of malignant lymphoma. The intraabdominal skeletal muscle and adipose tissue was diffusely infiltrated by medium to large atypical lymphocytes with irregular or round nuclei. Many individual neoplastic cells with apoptotic necrosis were seen. These atypical lymphocytes showed positive staining in the CD20 stain (B-cell marker), CD10 and BCL-6 stain, and negative staining in the CD3 stain (T-cell marker), and BCL-2 stain. The Ki-67 proliferation index was about 95%. The tumor invaded subserosa of colon, but the bilateral cut ends of colon

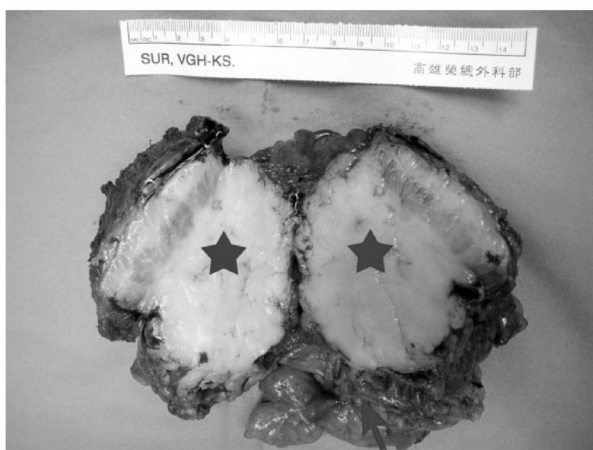


Fig. 2. The specimen consisted of one piece of soft tissue, labeled abdominal mass, measuring $16.8 \times 12 \times 8.3$ cm (star), with one segment of transverse colon (arrow), 8 cm in length and 4.5 cm in circumference.

were tumor-free (Fig. 3A, 3B).

The patient then received chemotherapy with the CEOP regimen (Vincristine, Epirubicin, Cyclophosphamide and Prednisolone) and was closely followed up at Hemato-Oncology OPD regularly.

Discussion

About 20-40% of Non-Hodgkin's Lymphomas (NHL) present as primary extra-nodal disease.^{1,12} Presenting sites include the gastrointestinal tract, the skin, the pleura, CNS and bone and more rarely soft tissue, the testes, the paranasal sinuses, the kidneys and adrenals, the urinary tract, the eye and orbit, the pancreas, the breast and the female genital tract.^{1,12} Primary soft tissue lymphoma is a rare entity accounting for 0.1-1% of new cases of NHL and 1.2-2% of all soft tissue tumors.^{1,13}

In our patient, no B-symptoms were present and pre-operative CT-guided or sono-guided biopsy was rejected by the radiologist due to a high risk of colon perforation. Under the impression of a desmoid tumor of the left abdominal wall, with peritoneal seedings, en-block excision of the abdominal wall tumor was performed. After pathologic examination, we (surgeons, pathologists and oncologists) convened a combined meeting and concluded that we held the opera-

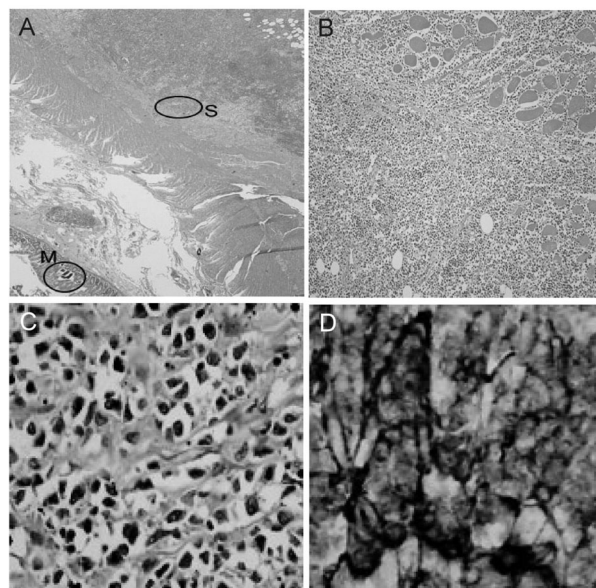


Fig. 3. B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma. Abdominal mass. (A) The atypical lymphocytes invades subserosa of colon (S: serosa of colon; M: mucosa of colon) (H&E, 20X). (B) The soft tissue and skeletal muscles are infiltrated by sheets of atypical lymphocytes (H&E, 100X). (C) Lymphoma is composed of sheets of cells with irregular nuclei and clear cytoplasm. Apoptosis and mitoses are seen (H&E, 400X). (D) CD20 is positive.

tion if the diagnosis of lymphoma was made pre-operatively.

The surgical specimen showed the morphological features that were diffusely infiltrated by medium to large atypical lymphocytes with irregular nuclei, resembling DLBCL; and the lack of starry sky macrophages with positive staining in the CD10, BCL-6 stain and high Ki-67 proliferation index resembling BL. The pathologist made the diagnosis and finally concluded that the malignancy encountered in this case is B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma.⁶⁻⁹

B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma is a new category of Burkitt lymphoma.⁴ These cases are rare and are mainly diagnosed in adults. They often invade extranodal tissue, but bone marrow and peripheral blood may also be in-

volved. The most appropriate therapeutic management has not been established because these are aggressive lymphomas.^{4,6} Our patient was treated for Burkitt lymphoma because BL is more aggressive than DLBCL. CEOP, CODOX-M and CNS prophylaxis using intrathecal methotrexate with cytarabine were prescribed and studies of CSF and bone marrow showed no malignancy after treatment. The total follow-up duration was eight months. The patient is still in healthy condition without symptom recurrence as of July, 2009.

Conclusion

Soft tissue lymphoma is a very rare clinical entity. In addition, B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma is a new category of the disease. The more we study, the more we will acquire in treating these patients in the future.

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病例報告

不可分類型的 B 細胞淋巴瘤發生於腹壁 並侵犯到橫結腸

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從軟組織長出的淋巴瘤是非常罕見的，在臨床上及影像學表現出不典型的特質。加上，這種不可分類型的 B 細胞淋巴瘤，特徵介於瀰漫性大 B 細胞淋巴瘤和伯基氏淋巴瘤之間，又是一種新的且罕見的類型，本篇文章提出一位發生於腹壁並侵犯到橫結腸的病例。

關鍵詞 軟組織淋巴瘤、腹壁腫瘤、侵犯大腸。