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Case Report .

T-Cell Rich B-Cell Lymphoma (TCRBCL) Initially Presented with Paraplegia In a 29 Years Old Saudi Male: A Case Report

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ABSTRACT

T-cell rich B-cell lymphoma (TCRBCL) is a newly recognized, rare variant of aggressive non-Hodgkin's B cell lymphoma. The term was first described in 1988. Here we report a rare interesting case of 29 years old Saudi male who was first presented to the hospital with paraplegia, urinary and stool incontinence, Radiological studies showed global lymphadenopathy. Primary histopathology reports showed Hodgkin's lymphoma and patient started on treatment accordingly, later the TCR BCL diagnosis was made, and patients received the optimum therapy, later patient admitted to ICU and unfortunately died of respiratory failure.

Key words: T-Cell Rich B-Cell Lymphoma, Paraplegia, Oncology, Aggressive Non-Hodgkin's B Cell Lymphoma.

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INTRODUCTION

T-cell rich B-cell lymphoma (TCRBCL) is a newly recognized, rare variant of aggressive non-Hodgkin's B cell lymphoma. It is characterized by the presence of predominant reactive T cells surrounded a few neoplastic B cells.[1,2] Ramsay et.al were the first who used the term T-cell rich B-cell lymphoma (TCRBCL) for describing this lesion and the term has gained a wide acceptance.[3] Here we report an interesting rare case of A 29 Years Old Saudi Male with TCRBCL who was first presented with paraplegia.

CASE PRESENTATION

29 year old male known case of eczema was referred to KAMC hospital's oncology ward for complete loss of lower limb movement, was associated with stool and urine incontinence, MRI of spine in the refereeing hospital revealed masses and hence was referred. This episode started by back pain and lower limb paresis for the duration of 3 weeks.

The pain was gradual in onset, continuous, increasing in severity, radiating to the back of both thighs. No relieving or aggravating factors. The pain was followed by lower limb paresthesia and weakness that progressed to complete loss of the patient's ability to move his legs. Other systemic review was unremarkable, Patient did not complain of headache, photophobia neck stiffness or blurred vision.

On examination the patient was vitally stable. He had bilateral lower limb weakness (grade 1), normal reflexes, positive babinski sign. There was loss of sensation below the level of T11.

Other systems were unremarkable.

The patient was immediately started on dexamethasone 8 mg IV Q 8 hours.

Radiological Investigations

Computed tomography (CT) scan of chest and abdomen with contrast revealed Internal mammary and axillary lymphadenopathy, prevertebral soft tissue lesion suggestive for lymphomatous infiltration with spinal canal extension and high suspicion of cord compression, bone involvement in T9 and T10 vertebral bodies along with sternum, Abdominal CT showed Pelvi-abdominal lymphadenopathy with multiple lymphomatous splenic lesions, lytic bony lesions in iliac bones and L5 vertebral body.

Magnetic resonance image (MRI) of the spine was done (Fig 1)

It was highly suggestive of neoplastic infiltration.

Biopsy was taken from cervical lymph nodes, and showed the following (Fig 2)

Immunohistochemistry staining showed (Fig 3)

Large cells were positive for: CD45, CD20. EMA and Bcl-6.

Large cells were negative for: CD15 and CD 3.

CD30 is weakly staining occasional large cells.

CD3 and CD68 shows rich background of T-lymphocytes and histiocytes.

There is no follicular network found with complete negative staining for CD21.

Final decision was made as T-cell/ histiocyte rich B-cell lymphoma.

Based upon the preliminary report given by the histopathologist, as he was primarily considered to be Hodgkin's lymphoma, the patient was given D1 C1 ABVD with steroids through IJV permcath. After reaching the final diagnosis of T-cell rich B-cell Non-hodgkin's lymphoma, he was put on C1 rituximab and C1 Chop. He also received high dose methotrexate 6 mg with good tolerance.

During the course of admission the patient developed fever and received IV antibiotics and Tamiflu. Both H1N1 and MERS-CoV were negative and his condition eventually improved.

On 12-1-2017 the patient developed respiratory and distress and febrile neutropenia, he was subsequently transferred to the intensive care unit (ICU) where he was intubated and placed on mechanical ventilation and received broad spectrum antibiotics: vancomycin imipenem, septrin, colistin, caspofungin and tamiflu and he also received filgrastim for neutropenia.

Follow up CT Scans of abdomen showed Interval decrease size of abdominal and pelvic lymph nodes, Interval near complete resolution of splenic lesions, Stable iliac bones and L5 vertebral body lytic lesions.

Follow up CT of the chest showed persistent bilateral interstitial and ground-glass opacities, Persistent bilateral lower lobe consolidation with air bronchogram, Stable enlarged mediastinal; supraclavicular and left axillary lymph nodes and numerous osseous lesions consistent with lymphomatous infiltration.

During his time in ICU the patient was kept under contact isolation, he had a fluctuant level of consciousness, however GSC was 15/15. His BUN and creatinine were elevated and renal impairment ensued. Unfortunately, the patient health deteriorated afterwards until he eventually died of respiratory failure

Table 1: Laboratory investigations upon admission:

CATEGORY (CBC)	Value	Normal range	Comment
WBC	6.22 x 10^9/L	4-10 x 10^9/L	Normal
RBC	4.74 million cells/mcL	4.5 - 5.5 million cells/mcL	Normal
Hemoglobin	13.4 g/dl	13.5-17.5 g/dl (for male)	Low
Hematocrit	41.8%	40%-52% (for male)	Normal
MCV	88.2 fL	80-100 fL	High
Platelet	355 x 10^9/L	150-400 x 10^9/L	Normal
Neut. count	4.4x 10^9/L	2-8 x 10^9/L	High
Ret. count	0.1739	0.5%-1.5%	Low
CATEGORY (LIVER FUNCTION)			
Albumin	3.44 g	3.5-5.2 g	Normal
ALT	54 U/L	0 - 40 U/L	High
AST	37 U/L	0 - 40 U/L	Normal
Bilirubin (Complete)	0.07 mg/dl	0.2 - 1.3 mg/dl	Normal
Bilirubin (Direct)	0.37 mg/dl	0 - 0.4 mg/dl	Normal
Amylase	80 U/L	30 - 86 U/L	Normal
LDH	286 U/L	140 - 280 U/L	High
CATEGORY (COAGULATION PROFILE)			
PT	13.4/sec	11 - 14/sec	Normal
PTT	28.6/sec	25 - 35/sec	Normal
INR	1.01	0.8 - 1.2	Normal
CATEGORY (RENAL AND ELECTROLYTE)			
Creatinine	0.69 mg/dl	0.6 - 1.3 mg/dl	Normal
Uric acid	6.25 mg/dl	3.4 - 7.0 mg/dL	Normal
Sodium	142 mmol/L	135 - 145 mmol/L	Normal
Potassium	3.9 mmol/L	3.5 - 5.3 mmol/L	Normal
Magnesium	2.02 meq/dl	1.3 - 2.1 meq/L	Normal
Calcium	8.43 mg/dl	8.6 - 10.0 mg/L	Normal

ALT: Alanine transaminase; AST: Aspartate transaminase; LDH: Lactate dehydrogenase;

PT: Prothrombin time; PTT: partial prothrombin time; INR: international normalized ratio.

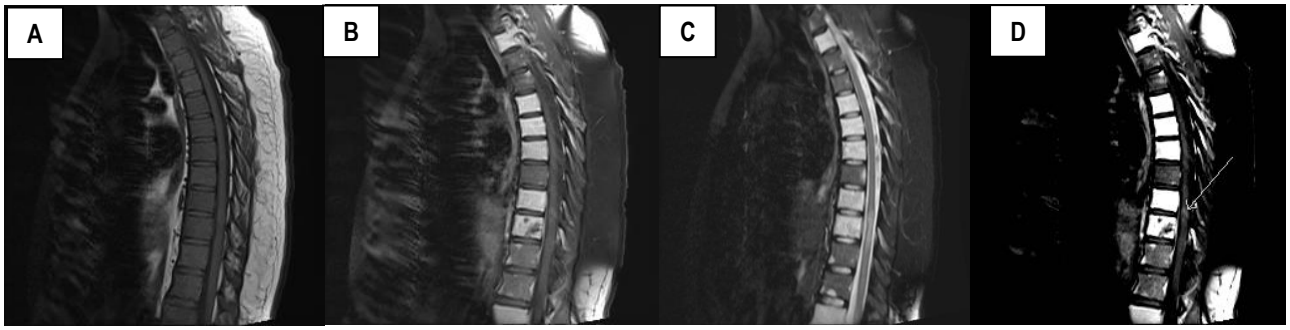


Figure 1: (A,B,C and D) Multiplanar multi sequences MRI imaging of the spine including post gadolinium sequences: Diffuse abnormal infiltrative patchy low T1/ high T2 marrow signal intensity associated with abnormal patchy enhancements involving the anterior and posterior vertebral body elements. Also, there is minimal amount of abnormal paraspinal and epidural thickening enhancement identified at thoracic and sacral region and causing moderate right T9-10 and left S2-3 neural foraminal narrowing. Neoplastic infiltration is highly suggestive. No evidence of pathological fracture or spinal canal narrowing. Non-expansile high T2 intra-medullary signal change identified extending form T9-10 to T10-11 level, with minimal parenchymal atrophy and questionable nodular enhancement. Furthermore, there is questionable tiny nodular enhancement at the cauda equina nerve roots at S1-2 level. Intrathecal infiltration would be considered.

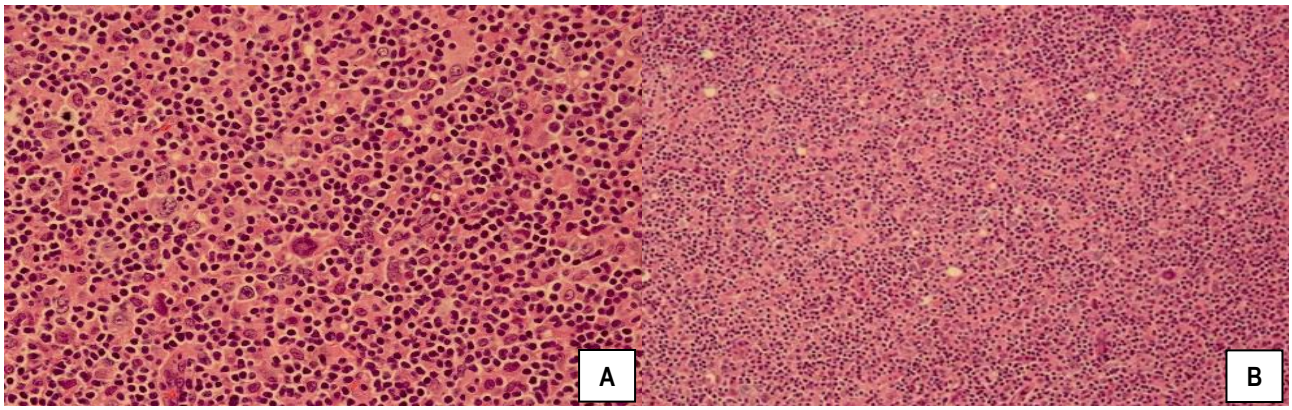
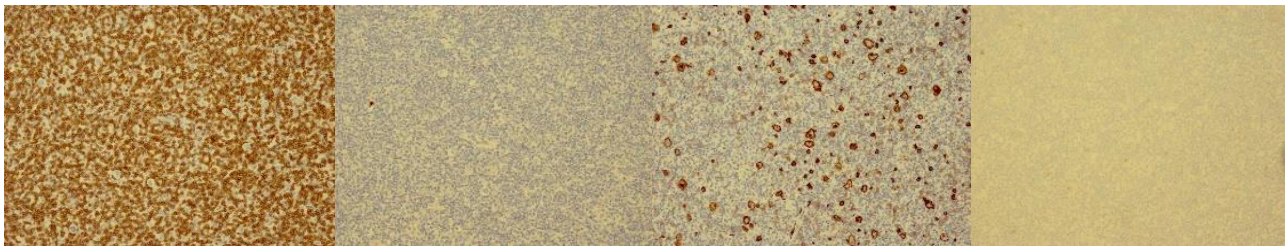


Figure 2: (H & E staining) Macroscopy: Highly atypical lymphoid proliferation consistent with lymphoma. Microscopy: Diffused infiltration of histiocytic and lymphoid background. Many scattered highly atypical large cells with nuclear atypia. No follicular or nodular pattern of proliferation.



CD3 **CD15** **CD20** **CD30**

Figure 3: Immunohistochemistry staining

DISCUSSION

Disease Definition

The most common malignancy of lymphoid tissue in adults is Diffuse large B-cell lymphoma (DLBCL), accounting for approximately 40% of Non-Hodgkin's lymphomas[4], DLCBL has several subgroups, the rarest that represents 1-3% is TCR BCL.[5] The term TCR BCL was first described in 1988 by Ramsy et al.[3] Through the years TCR BCL has gained a special attention because of the diagnosis's challenging that physicians faced; the disease is highly aggressive, but resembles higher similarities between different lymphoma entities such as Peripheral T cell lymphoma, Classical Hodgkin lymphoma, and nodular lymphocyte predominant Hodgkin's lymphoma (NLPHL).[3,6-9] Thus the disease is often misdiagnosed, subsequently patients do not receive the optimum treatment for TCR BCL, patients who are misdiagnosed are often perform poorly.[10]

In order to diagnose TCR BCL early; Several categorical criteria have been developed, recently WHO limited the diagnosis of TCR BCL for only cases with or with less than 10% of large B tumors cells[11,12] even with the criteria, the diagnosis of TCRBCL is still challenging and initial Misdiagnoses is common.[10]

TCR BCL usually occurs in the 4th decades of life, the disease showed male predominance pattern.[13] Patients are more likely to suffer from non-specific symptoms of fever, fatigue, night sweat and unintentional weight loss.[4,14] In Immunohistochemistry of malignant cells are markedly positive for CD45, CD20 and negative for CD15, CD5 and CD138, Rarely cells show weak positivity for CD30, and up to 50% of tumor cells express Bcl-2, a poor prognostic indicator in DLBCL.[15] Other immune-histochemistry markers have been reported too.[7,13,16]

Treatment of TCR BCL is based on chemotherapy drugs that is anthracycline based plus rituximab, the anti CD20 drug, similar to DLBCL therapy, also all patients should follow the same protocols according to their tumor stage that matched DLBCL.[10]

TCR BCL outcomes is similar to DLBCL patients, with 5 year overall survival rates between 45%-58%.[10]

CONCLUSION

TCRBCL is an aggressive form of Non-Hodgkin's Lymphoma, that is often initially misdiagnosed and patients are treated for other diseases, such as Classical Hodgkin's lymphoma. Higher mortality rate is associated with delay of starting the optimum therapy, highlighting the importance of precise initial diagnosis.

SOURCE OF SUPPORT

King Abdullah medical city, Makkah, Saudi Arabia.

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