VOLUME-6 | ISSUE-3 | MARCH - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

A Histopathological Study of Malignant Round Blue Cell Tumors

Pathology

KEYWORDS: Malignant Round Blue Cell Tumors, Non Hodgkin lymphoma

Dr.Viswanathan K — Assistant Professor, Government Mohan Kumaramangalam Medical College, Salem, Tamilnadu.

Dr. Premalatha V — Assistant Professor, Government Mohan Kumaramangalam Medical College, Salem, Tamilnadu

ABSTRACT

Malignant Round Blue Cell Tumors (MRBCT) comprise a diverse group of diagnostically challenging primitive (or) undifferentiated neoplasms.Aim : To find out the incidence, age and sex distribution and correlate the clinical and histopathological features of Malignant Round Blue Cell Tumors. Materials and methods: Surgical specimens were fixed in 10% neutral buffered formalin. Section of 5 micron thickness were cut and stained with Hematoxyllin and eosin stain. Conclusion: Non Hodgkin's Lymphoma (NHL) is the most common type of tumor in this study which is about 51.5% (36 cases) and MRBCT other than NHL constitute 48.5% (34 cases).

Introduction

MRBCT comprise a diverse group of diagnostically challenging primitive (or) undifferentiated neoplasms which composed of small cells with uniform round nuclei and scanty cytoplasm have been referred as small-cell, round-cell, or blue cell tumors[1]. The tumors included in this category of neoplasms are Non Hodgkin's Lymphoma, Ewing's sarcoma /Primitive neuroectodermal tumor (PNET), Embryonal and Alveolar Rhabdomyosarcoma, Wilm's tumor, Neuroblastoma, Desmoplastic Small Round Cell Tumor (DSRCT), Mesenchymal Chondrosarcoma, Small cell Osteosarcoma, Small cell carcinoma, Malignant Rhabdoid tumor, Myxoid Chondrosarcoma, Poorly differentiated synovial sarcoma, Round cell Liposarcoma, Epithelioid sarcoma with rhabdoid phenotype Retinoblastoma and Medulloblastoma[2,3]. Among this NHL is the most common type. MRBCT other than NHL is common in children and adolescence whereas NHL is common in the older age group with male preponderance.

Materials and Methods

The study had been carried out in the Department of Pathology, Government Mohan Kumaramangalam Medical College, Salem for a period of 2 years from June 2012 to May 2014. The specimens were received from GMKMC Hospital.

The clinical details of the patients were recorded with the informed consent. The histopathology specimens were fixed in 10% formalin. The tissues were processed, paraffin blocked, 5 micron thin sections were cut and stained with Hematoxylin and Eosin (H &E). Special stains such as PAS (Periodic Acid Schiff's) and Reticulin were used as and when required.

Results

70 cases had been reported as MRBCT with the incidence of 1.38 %.

Table-1

Tumor type	No. of cases	Percentage (%)
NHL	36	51.5
Ewing's sarcoma / PNET	10	14.5
Rhabdomyosarcoma	7	10
Wilm's tumor	7	10
Neuroblastoma	5	7
DSRCT	1	1.4
Small cell Osteosarcoma	1	1.4
Extraskeletal Mesenchymal Chondrosarcoma	1	1.4
Undifferentiated MRBCT	2	2.8
Total	**70**	**100**

The age incidence of MRBCT ranges from 3 months to 92 years in this study. It is observed from the study that MRBCT are commonly seen in pediatric age group except NHL which is common in adults. Out of the 70 cases of MRBCT, 45 cases occurred in males and 25 cases in females with the male to female ratio of (1.8:1).

Table-2

HPE Diagnosis	Age incidence	
	Mean	Standard Deviation
NHL (36)	44.9	23
Ewing's Sarcoma (10)	21.3	12.9
Rhabdomyosarcoma (7)	16.9	12.3
Wilm's Tumour (7)	4.5	4.4
Neuroblastoma (5)	13.9	21.3
Malignant round blue cell tumour (2) (Undifferentiated)	30.3	39.2
Others(3)	12.8	12.9

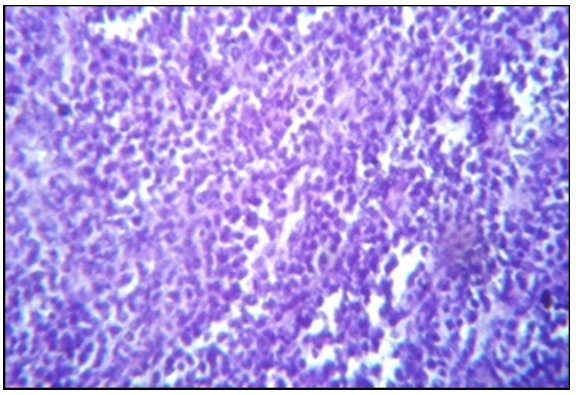

Fig 1: NHL Lymph node: Shows monotonous proliferation of lymphocytes with scanty cytoplasm and regular round nuclei

NHL predominantly occurred in males with the male to female ratio of 2.3:1. The three tumors, Ewing's sarcoma / PNET, Wilm's tumor and Rhabdomyosarcoma had a male predominance with the male to female ratio of 2.3:1, 1.3:1 and 1.3:1 respectively, whereas Neuroblastoma had a female preponderance with the male to female ratio of 1:4.

NHL was reported in the Nodal as well as the extra nodal sites. 26 cases of nodal NHL presented with generalised lymphadenopathy and one case had leukemic dissemination.10 cases of NHL occurred in different extra nodal sites with varied clinical presentation depending upon the site of involvement.

Out of 10 cases of Ewing's sarcoma / PNET , 7 were observed in lower

segment of the skeleton and the rest 3 in upper segment.

All the 7 cases of Wilms tumor presented with abdominal mass and were unilateral. Rhabdomyosarcoma occurred mainly in Head and Neck region and also in other sites such as genitourinary tract and extremities. Neuroblastoma most commonly presented as retroperitoneal mass rarely as presacral and nasal mass.

The microscopic study showed 34 cases as NHL and 36 cases as MRBCT other than NHL. Out of the 34 cases of NHL, 26 occurred in the nodal and the rest 8 in extranodal sites

Among the 36 cases of MRBCT other than NHL constituted 8 cases of Rhabdomyosarcoma, 7 cases of Wilm's tumor ,6 cases of Ewing's sarcoma / PNET and 5 cases of Neuroblastoma. The study also showed one case in each of small cell Osteosarcoma , Extra skeletal Mesenchymal Chondrosarcoma and Desmoplastic Small Round Cell Tumor (DSRCT). Rest of the cases didn't fit into any of the specific type and were categorized as Undifferentiated MRBCT.

Special stains

Special stains such as Reticulin and Periodic Acid Schiff's (PAS) were done in selective cases. Reticulin stain was done for all the cases of nodal NHL which showed fine branching reticulin network with pericellular fibrils characteristic of Lymphoma. PAS stain was done for 6 cases of Ewing's sarcoma/PNET, which demonstrated abundantcytoplasmicglycogenpositivity[4,5].

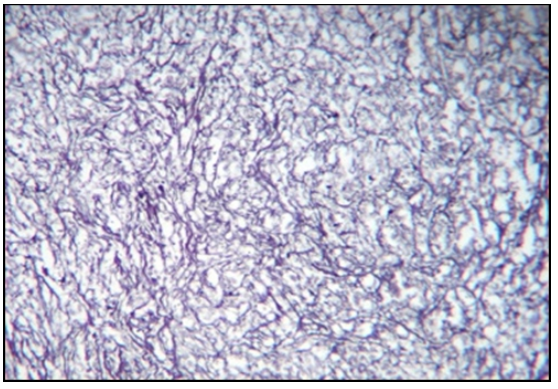

Fig .2: NHL Lymph node shows loss of architecture with pericellular fibrils

DISCUSSION

MRBCT comprise a diverse group of diagnostically challenging primitive and undifferentiated neoplasms. Although classical histological features are highly suggestive of a tumor type, on occasion these tumors are indistinguishable by light microscopy, making a definitive diagnosis difficult. Accurate diagnosis of MRBCT has become increasingly crucial, as disparate approaches to therapy are used for distinct tumor types.

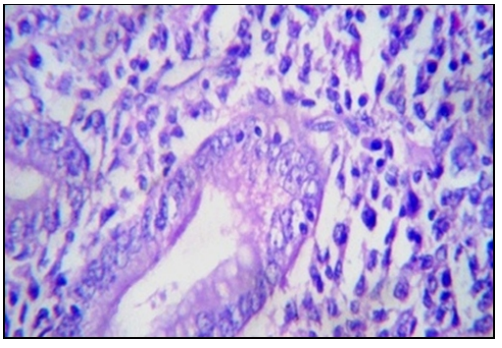

Fig 3:NHL stomach: Shows mucosal glands infiltrated by lymphocytes
(Lymphoepithelial lesion)

In this present study, the age of the patients with MRBCT ranged between 3 months to 92 years, 23 cases in children with 32.9% of incidence and 47 cases in adults with 67.1% of incidence.

NHL is common in adults than in children with ratio of 4:1.The mean age at diagnosis of NHL is 44.9 years. Surveillance Epidemiology and End Results (National Cancer Institute.USA) showed 67 years as the median age at diagnosis of NHL[6].

Ewing's/PNET is predominantly seen in adolescent age group, which is similar to the study conducted by Carvajal R et al[7]. In the present study, Rhabdomyosarcoma (RMS) is common in children than adults. Most of the RMS cases are below 21 years with the mean age at diagnosis of 16.9 years which correlates well with Michael et al[8].

Median age at diagnosis of Wilm's tumor as 53 months (4.5 years). Mir Mahmood Seyed Ahadi et al[9] found that mean age at the time of diagnosis as 45.2 months (4years) which is slightly lower than that found in the present study. Norman Breslow et al[10] found that median age at diagnosis of Wilm's tumor is 39.5 months. 80% cases of Neuroblastoma are seen under the age of 6 correlating with Stiller C A et al[11].

Malignant Round Blue Cell Tumors show a male: female ratio of 1.8:1. This study shows a male preponderance of Non Hodgkin's lymphoma with a male: female ratio of 2.2:1, which correlates with the Surveillance Epidemiology and End Results (National Cancer Institute.USA)6 which also shows a male preponderance with a male: female ratio of 1.4:1.

Carvajal R et a[17] found slight male predominance of Ewing's sarcoma/ PNET. This current study correlates with the above study with a male: female ratio of 2.3:1.

Simona Ognjanovic et al[12] found a male preponderance for Rhabdomyosarcoma with a male: female ratio of 1.37:1 which correlates with ourstudy..

Mir Mahmood Seyed Ahadi et al9 found that Wilm's tumor is common in males with a male: female ratio being 1.2:1. This present study correlates with the above study with a male: female ratio of 1.3:1.

Norman Breslow et al[10] found that slight preponderance of females in the National Wilm's tumor study registered between October 1969 and December 1985.

Gregory Hale et al[13] found a male preponderance in Neuroblastoma with a male to female ratio of 1.3:1. In contrast, this present study shows distinct female preponderance with the male: female ratio being 1:4.

Site

In this study, extra nodal NHL constitutes about 27% of the total NHL cases recorded. This can be compared with Antoria M.S.Muller et al[14] who found that extra nodal NHL constitutes 20-30% of all NHL in his study.

Sandeep Agarwala et al[15] found that most common site of occurrence of Rhabdomyosarcoma is the head and neck region. This present study correlates with the above study.

Kissane et al[16]found that Ewing's sarcoma has a predilection for lower segment of skeleton as this current study.

Neuroblastoma commonly present as retroperitoneal mass which correlates with the study conducted by DeLorimer et al based on California Tumor Registry.

Reticulin stain was done for all the cases of nodal lymphoma to confirm the effacement of architecture, which is characteristic of

Lymphoma.

PAS stain was done to confirm for all the cases of the Ewing sarcoma/PNET to confirm the presence of glycogen in the cytoplasm. Although these two special stains are not specific for the diagnosis they had a limited role in categorizing the MRBCT.

CONCLUSION

MRBCT comprise a wide range of neoplasms varying considerably in regard to etiology, site of origin, involvement of skeletal system and prognosis.NHL is the most common type of tumor in this study which is about 51.5% (36 cases) and MRBCT other than NHL constitute 48.5% (34 cases). MRBCT other than NHL is common in children and adolescence whereas NHL is common in the older age group and have a male preponderance. The common clinical presentation of NHL is lymph nodal enlargement. Ewing's sarcoma/PNET affects lower segment of the skeleton whereas Rhabdomyosarcoma involves the head and neck region. Neuroblastoma usually presents as retroperitoneal mass. Reticulin and PAS stains are supportive to the diagnosis of NHL and Ewing's sarcoma /PNET respectively in the study.

Accurate histopathological diagnosis of MRBCT is essential for effective therapeutic management and prognosis of the patients.

References:

1. Vincent J. Vigorita, Orthopaedic Pathology. 2nd Edition, Volter Kluwer / Lippincott Williams & Wilkins, 2008
2. Meera Hameed, Small Round Cell Tumors of Bone, Arch Pathol Lab Med – Vol.131, February 2007
3. Sharon W. Weis, John R.Goldblum, Enzinger and Weiss's Soft Tissue Tumors fifth edition, Mosby An imprint of Elsevier science
4. Handbook of Histopathological Technique by culling CFA, 1957. Butterworth & Co., (Publishers) Ltd
5. Juan Rosai. Rosai and Ackerman's Surgical Pathology 10th edition Volume I & II, Mosby An imprint of Elsevier. 2004.
6. Surveillance Epidemiology and END results. National Cancer Institute. U.S. National Institutes of Health / www.cancer.gov
7. Carvajal R, Meyers P. Ewing's sarcoma and primitive neuroectodermal family of tumors. Hematol oncol Clin North Am 2005; 19:501-525
8. Michael P. La Quaglia, Glenn Heller, Freshteh Ghavimi, Ephraim S. Casper, Vaia Vlamis, Steven Hajdu, Murray F. Brennan. Cancer
9. Mir Mahmood Seyed – Ahadi, Ahmad Khaleghnejad –Wilm's tumor: A 10 year Retrospective study. Arch Iranian Med 2007; 10 (1): 65-69
10. Norman Breslow, J.Bruce Beckwith, Marcia Ciol and Katrina Sharples. Age distribution of Wilm's Tumor: Report from the National Wilm's Tumor study. Cancer Research 1988; 48 (15): 1653-165
11. Stiller CA, Parkin DM. international variations in the incidence of neuroblastoma. International Journal of Cancer. 1992 Oct 21; 52(4):538-543.
12. Simona Ognjanovic, Amy M Linabery, Bridget Charbonneau and Julie A. Ross. Cancer 2009; September 15:115 (18): 4218-4226.
13. Gregory Hale, Mary J. Cula and Julie Blatt. Pediatric Hematology – Oncology 1994, Volume 11, No.1, Pages 91-97
14. Antoria M.S.Muller, Gabriele Ihorst, trends, geographic distribution, and etiology. Annals of Hematology (2005) 84: 1-12
15. Sandeep Agarwala. Pediatric rhabdomyosarcomas and non rhabdomyosarcomas soft tissue sarcoma. Journal of Indian Association of Pediatric surgeons 2006; 11 (1): 15-23
16. Kissane JM, Askin FB, Foulkes M, ewing's sarcoma of the bone: Hum Pathol.1983;14:773-779.