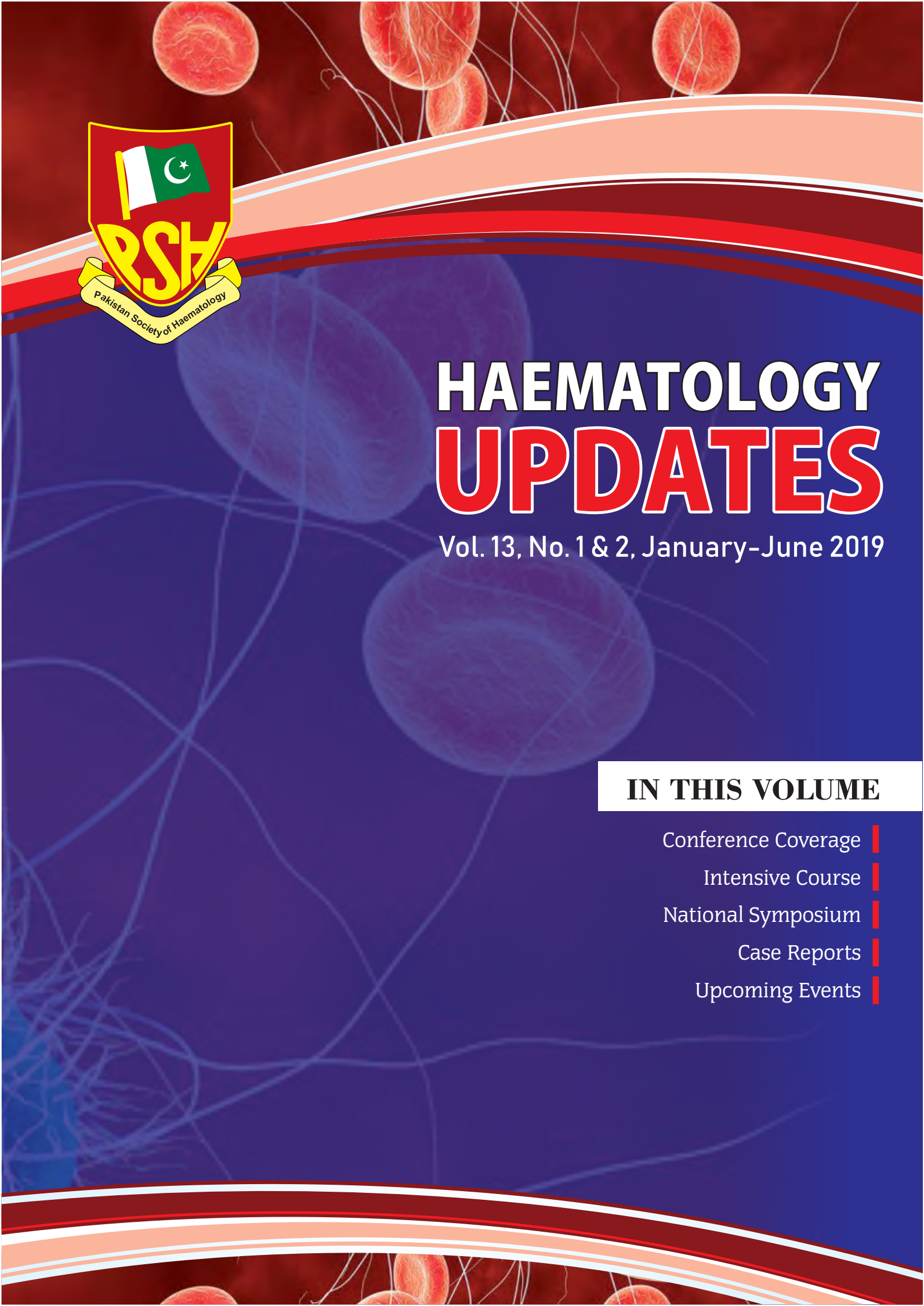

HAEMATOLOGY UPDATES

Vol. 13, No. 1 & 2, January-June 2019

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PRESIDENT COLUMN

I feel humbled, honored and privileged to assume the role of President, Pakistan Society of Haematology. PSH has completed 21 years as an organization and has a long list of achievements. As a professional body, PSH has expanded with a membership of over 500 and continues to grow. It has successfully created strong outreach and educational programmes and has been regularly holding annual conferences. Lately PSH has been trying to provide outreach and support in areas of the country with less facilities and resources with the goal of ensuring improvement in care for patients with blood diseases. After these achievements, however, a great deal of work still needs to be done to meet our objectives.

I am grateful to my predecessor, Professor Nisar Ahmed, for his outstanding contributions to PSH during his presidency, and the members of previous executive committee. I am inspired by their commitment to PSH and all of their achievements. Indeed, the growth of PSH has been boosted collectively by the individuals and all the past presidents and members of the PSH. I look forward to continuing this important work towards fulfilling the mission of our professional body. In this endeavor, I am joined by my colleagues on the Executive Committee- they have we all know distinguished themselves as a persons with dedication and professionalism. I am confident that they will continue to make outstanding contributions to PSH.

I am proud to be given this opportunity, and I will continue- with your help to develop PSH as a national authority in diagnosis and management of blood diseases including the ever-expanding field of haematopoietic stem cell transplantation. I see a true need to accelerate the pace of our work because of the escalating prevalence and complexity of haematological diseases. During the next two years PSH will continue to address the unmet national guidelines for important haematological diseases keeping in view our resource constraints. PSH will also work to promote education, research and advocacy reaching out to our member societies and other organizations.

I request all the members, especially our senior colleagues for their active help and involvement in PSH affairs to achieve our common goals.

On that note, I wish you all the very best for the coming seasons

Yours sincerely,

Dr Parvez Ahmad
President - PSH
Head Department of Clinical Haematology & BMT
Quaid-e-Azam International Hospital, Islamabad - Pakistan

JOIN US!

Dear Colleagues,

Assalam-O-Alikum

On behalf of new PSH Council, I extend my deepest gratitude to all PSH members for showing their trust & support.

My God Almighty bless us with the strength to honor this obligation. Your guidance and valuable suggestions would be required throughout.

Skype meeting was arranged for executive council for convenience of members and a warm response was received from all the participants with progressive ideas for the society.

Current PSH newsletter includes case reports of interesting/unusal haematological diseases and reports about recent haematology advances in our country.

Upcoming Haemcon2020 has been planned in February 2020. Your enthusiastic contributions are requested for the success of this mega event in historical city of Lahore. The vibrant budding haematologists should come forward and share their research with new ideas.

Let's follow the advice by Poet of East Dr. Allama Muhammad Iqbal.

جہانِ تازہ کی افکارِ تازہ سے ہے نمود
کہ سنگ و خشت سے ہوتے نہیں جہاں پیدا

خودی میں ڈوبنے والوں کے عزم و ہمت نے
اس آبِ جو سے کیئے بحر بیکراں پیدا

Wishing sincerely for the health, happiness & prosperity of all members.

Yours sincerely,

Dr Mehreen Ali Khan
Secretary - PSH
Consultant Haematologist and BMT Physician
AFBMTC/NIBMT, Rawalpindi - Pakistan

ABOUT PSH

Pakistan Society of Haematology (PSH) was formed in 1996 with the aim of promoting advancement of haematology, BMT and transfusion medicine in the country. Presently it has more than 500 members and we all should make efforts to enroll every haematologist in the country. We request all our members to take special interest in extending the membership to all those haematologists around you who have not yet registered with PSH. Website was launched and has been very active in recent past. We are trying to rejuvenate the website "https://www.psh.org.pk. The website would be interactive and provide on line forum for sharing views with other haematologists, and case discussion with the experts. Other features will be facility to download online membership form, newsletter, list and addresses of the members. Hopefully the website will be more operational within this month InshaALLAH.

HISTORY

PSH was raised as "Pakistan Society of Haematology/Transfusion Medicine (PASHT)" in 1991. A meeting was held at 5 pm on Friday November 22, 1991. Professor Dr Mohammad Khurshid, Brig (later Lt Gen) Muhammad Saleem, Dr Khalid Zafar Hashmi, Dr Nasim Siddiqui, and Dr Abdul Hayee attended the meeting as members in presence of Prof A. V Hoffbrand. In this meeting Dr Khurshid presented a brief outlay of the necessity to create such a society. He also pointed out that Dr. Abdul Hayee, Dr. Khurshid, Dr KZ Hashmi and Brig Saleem had met at Bahawalpur and agreed on the general principles that the first meeting would be held along with the International conference of Pathology

Though initial work was comprehensive, governing body and meetings of PASHT were not held regularly. In September 1994 it was proposed by Gen Muhammad Saleem to meet all PASHT members during Pakistan Association of Pathology (PAP) conference at Quetta. Dr. Muhammad Khurshid in consultation with Gen Saleem, Prof. Abdul Hayee, Dr. Khalid Zafar Hashmi proposed a provisional constitution of PASHT for the discussion in meeting.

Haematologists from all over the country met on Saturday 09 March 1996 at Hotel Pearl Continental Rawalpindi in order to form a society. It was unanimously agreed that official name of society will be "Pakistan Society of Haematology" with official abbreviation of "PSH". It was also decided that until elections for office bearers the society matters will be looked after by a committee as under

a. Dr. Muhammad Khurshid
b. Dr. Ehsan-ul-Allah
c. Dr. Abdul Hayee
d. Dr. Khalid Zafar Hashmi
e. Dr. Khalid Hassan
f. Dr. Masood Anwar will act as Co-ordinator

A general body meeting of PSH was held at Peshawar on 2 and 3 November 1996. Election for office bearers were carried out as follow:

ABOUT PSH

a. Dr. Muhammad Saleem President
b. Dr. Muhammad Khurshid as Vice President
c. Dr. Khalid Hassan as Secretary/treasurer

Later in October 1997 appointment of vice president was renamed as president elect:

List of past presidents includes:

1. Dr. Abdul Hayee
2. Dr. Abdul Khaliq
3. Dr. Muhammad Khurshid
4. Dr. Khalid Zafar Hashmi
5. Dr. Masood Anwer
6. Dr. Khalid Hassan
7. Dr. Suhaib Ahmed
8. Dr. Samina Naeem
9. Dr. Muhammad Ayyub
10. Dr. Nisar Ahmed

List of past secretaries includes:

1. Dr. Khalid Hassan
2. Dr. Massod Anwar
3. Dr. Fazle-e-Raziq
4. Dr. Salman Naseem Adil
5. Dr. Shaheena Kauser
6. Dr. Nadir Ali
7. Dr. Pervez Ahmed
8. Dr. Nadeem Ikram
9. Dr. Humera Rafiq
10. Dr. Tariq Mahmood Satti
11. Dr. Saima Farhan

PSH was registered with Govt of Pakistan on 08 August 1998 RS/ICT/298 dated 08 August 1998 as non political and non sectarian body to promote advancement of haematology including transfusion medicine through encouragement of research, teaching and technical methods. The body will also organize scientific meetings, publication of scientific material, and affiliation with other National and international organizations. Members of Governing body included:

a. Dr. Muhammad Saleem as President
b. Dr. Khalid Hassan as General secretary
c. Dr. Birgees Mazhar Qazi as member
d. Dr. Waseem Iqbal as member
e. Dr. Hassan Abbas Zaheer as member
f. Dr. Mobina Ahsan Dhodhy as member
g. Dr. Farah Yasin as member
h. Dr. Masood Anwar as member

It was also decided that first National conference will be held on 04 October 1998. Since then Annual conference is held regularly in all capital cities of Pakistan. The society is publishing a quarterly newsletter and providing a forum to the haematologists all over the country contributing as advisors in haematology, consultants, researchers and mentorship. Currently the Governing body includes:

Dr. Parvez Ahmed as President
Dr. Salman Naseem Adil as President Elect
Dr. Mehreen Ali Khan as Secretary/Treasurer

EXECUTIVE COMMITTEE

New Executive Committee was elected during 21st Annual Conference of Pakistan Society of Haematology (PSH) held at Karachi from 14th - 16th March 2019. Following are the office bearers of Executive Committee:

Dr. Parvez Ahmed
President
Cell: +92 300 8561288
Email: parvez101@yahoo.com

Dr. Salman Naseem Adil
President Elect
Cell: +92 300 9249027
Email: salman.adil@aku.edu

Dr. Mehreen Ali Khan
Secretary/Treasurer
Cell: +92 333 5164941
Email: mehreen35@hotmail.com

MEMBERS

Armed Forces
Qamar Un Nisa Chaudhry
Asad Mehmood Abbasi
Muhammad Sajid Yazdani

Islamabad
Nadeem Ikram

Punjab
Saima Farhan
Muniza Junaid
Muhammad Irfan Khan

Sindh
Syed Muhammad Irfan
Ikram Din Ujjan
Muhammad Nadeem

Khyberpakhtunkhwa
Shahtaj Masood

Baluchistan
Hayat Ullah

Azad Kashmir
Zahida Qasim

Office Coordinator
Imran Waheed
Tel: +92 51 9270076 Ext. 228 Mob: +92 322 5181302
Email: psh.org.pk@gmail.com

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RAWALPINDI/ISLAMABAD	KARACHI	LAHORE
Dr Asad Mehmood Abbasi asadabbasi739@yahoo.com 03215506566	**Dr Bushra Moiz** bushra.moiz@aku.edu 0300-2160765	**Dr Muneeza Junaid** dr.mjunaid10@gmail.com 0333-8029026

QUETTA	PESHAWAR	AZAD KASHMIR
Dr Hayat Ullah hayatdotani@yahoo.com 03337520574	**Dr Shahtaj Masood** **shahtajmasood@yahoo.com** **0333-9118335**	**Dr. Zahida Qasim** dr_qasim2011@hotmail.com 03335875122

NATIONAL ADVISORY AND STEERING COMMITTEE

- Dr. Suhaib Ahmed
- Dr. Samina Naeem
- Dr. Mohammad Ayyub
- Dr. Fazle Raziq
- Dr. Javed Asif
- Dr. Muhammad Amin
- Dr. Farooq Khattak
- Dr. Barjeez Mazhar Qazi
- Dr. Saeed Ahmed Malik
- Dr. Nughat Yasmin Ashraf
- Dr. Jalil Anwar
- Dr. Waseem Iqbal
- Dr. Syed Iftikhar Abdi
- Dr. Ehsan Alvi
- Dr. Muhammad Saleem
- Dr. Abdul Hayee
- Dr. Muhammad Khurshid
- Dr. Abdul Khaliq
- Dr. Khalid Zafar Hashmi
- Dr. Masood Anwar
- Dr. Khalid Hassan
- Dr. Yasmin Lodhi
- Dr. Tahiq Jameel Ghazi
- Dr. Qaiser Husnain
- Dr. Ghulam Rasool
- Dr. Farzana Amjad
- Dr. Nouman Malik
- Dr. Fozia Butt
- Dr. Zahoor Ur Rehman
- Dr. Luqman Butt
- Dr. Farhat Abbas Bhatti
- Dr. Nadir Ali
- Dr. Muhammad Ashraf
- Dr. Tahira Zafar
- Dr. Zeba Aziz
- Dr. Madoodul Manan
- Dr. Mahadev Harani
- Dr. Zahoor ul Latif
- Dr Mian Muhammad Sharif
- Dr. Mussarat Niazi
- Dr. Muhammad Saeed Talpur
- Dr. Atifa Shoaib

SCHEDULE OF MONTHLY MEETING

City	Date	Time	Coordinator
Rawalpindi/ Islamabad	Last Thursday of the Month	03:00pm – 05:00pm	Dr Asad Mahmood Abbasi
Karachi	Last Friday of the Month	08:00am – 09:00am	Dr Bushra Moiz
Lahore	2nd Tuesday of the Month	09:00am – 10:00am	Dr Muneeza Junaid
Quetta	Last Friday of the Month	09:00am – 10:00am	Dr Hayat Ullah
Peshawar	3rd Thursday of the Month	12:00pm – 01:00am	Dr Shahtaj Masood

HIGHLIGHTS

21ST PAKISTAN SOCIETY OF HAEMATOLOGY
ANNUAL INTERNATIONAL CONFERENCE 2019
March 14 - 16, 2019 | Karachi - Pakistan
HAEMCON-2019

The 21ST Annual Pakistan Society of Haematology conference was successfully conducted at Aga Khan University Hospital Karachi, from 14-16th March 2019. There were seven pre-conference workshops which were very well attended by participants on March 14, 2019. The conference days were March 15-16 2019. The first day of the conference was commenced with the recitation of verses from Holy Quran followed by the national anthem of Pakistan. The chairperson of HAEMCON-2019. Our chief guest for the conference was Prof. Dr. Masood Anwar. The inaugural session had two plenary lectures by international speakers namely Dr. Muzaffar Qazilbash (Progress in immunological and cellular therapy for blood cancer) and Dr Shahrukh Hashmi (The role of artificial intelligence in stem cell transplantation). Dr Nisar Ahmed (ex-president PSH) presented the Ibn-e-Sina lecture on "Childhood Myelodysplastic Syndrome". The inaugural session ended with vote of thanks by Dr. Izza Hussain, Secretary HAEMCON-2019.

On the first day of the conference there were sessions on red cell and white cell disorders which consisted of state of the art lectures on various topics by national and international speakers. The topics included aspects of G6PD deficiency, neurological effects of anemia, new treatment options in thalassemia major, CLL, CML and the entity of double hit lymphoma. The second day started off with

HIGHLIGHTS

HAEMCON-2019 KARACHI

session on platelet and coagulation disorders where thrombotic and bleeding disorders in neonates was discussed. We also had a presentation on quality assurance in coagulation laboratory by one of our international speaker – Dr Catherine Hayward (President of the North American Special Coagulation Laboratory Association).

The next session was on transfusion medicine and important topics of voluntary blood donation, molecular testing, quality assurance and massive transfusion in trauma were debated. This year, we introduced experimental haematology as a theme where ground breaking studies from bench to bedside were presented e.g. musculoskeletal ultrasound in haemophilia, apoptosis in CD34+ EPCs by vitamin E in anemia, CAR T cell therapy and NK cell immunotherapy in AML. The last session was on stem cell transplant where speakers talked on current status and challenges of transplant in Pakistan and future of haploidentical transplantation. Each thematic session had oral presentations by residents on the original work done by them, supervised by respective faculty (a total of 10 presentations were included in the programme). Each oral presenter received travel grant if they were stationed outside Karachi.

The general body meeting and banquet dinner was held on March 15, 2019. There were four awards for best presented in the banquet dinner. The conference ended with concluding remarks by Prof. Dr. Mohammad Khurshid.

HIGHLIGHTS

14TH FCPS HAEMATOLOGY INTENSIVE COURSE 2019

The 14th FCPS Haematology Intensive course was held at The Indus Hospital (TIH), Karachi from 26 -28 March 2019. The course was organized by TIH in collaboration with Pakistan Society of Hematology (PSH); it was the first effort of TIH team to play its role in this crash course that helps residents in quick revision of syllabus. The successful completion of this 3-days program was possible because of the support of Haematology faculty who participated in this course from all over the country.

This course was intended for haematology residents particularly those appearing in exam therefore designed according to CPSP pattern, focused on self-assessment of residents followed by interactive discussions with examiners. The course was led by Dr. Fatima Meraj, Section Head Haematology (TIH) with new perspectives introduced by her team i.e. case based discussions which also covered commonly encountered problems arise during exam preparation. It was a unique experience for residents as they were exposed to extensive morphology, exam oriented dry challenges, wet part (benches) of coagulation and transfusion medicine followed by the detailed discussion of each given case by senior faculty.

The course was inaugurated by Dr. Fatima Meraj, Welcome address was delivered by Dr. Saba Jamal, Director Laboratory and Blood centre (TIH). During three-days course, besides renowned speakers conducted case based discussions, also shared exam tips with candidates. As morphology is the most important component, Day 1 was dedicated to morphology to cover maximum variety of cases; hands on workshop was followed by case based discussions. Dr. Neelum Mansoor (TIH) was the session co-ordinator for morphology. Topics covered in day 1 discussion was approach to pancytopenia, myelophthisic bone marrow infiltration and acute leukemia. An extensive discussion on above mentioned topics was done by Dr. Fatima Meraj, Prof. Dr. Syed M Irfan and Dr. Muhammad Nadeem. Day 2 was started with enlightening talks on MPN/MDS by Prof. Dr. Tahir Shamsi and Dr. Nida Anwar.

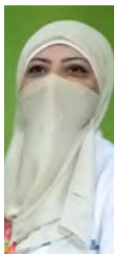
HIGHLIGHTS

14TH FCPS HAEMATOLOGY INTENSIVE COURSE 2019

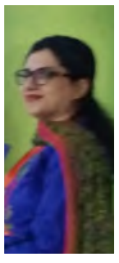

Red cell disorders, hemoglobinopathy and chronic leukemias alongwith interpretation of flowcytometry was elaborated by Prof. Dr. Salman Adil, Prof. Dr. Nisar Ahmed and Dr. Fatima Meraj respectively. Speakers and trainees both were enthusiastic as this course provided them a platform to put forward their queries.


In hemoglobinopathy session, the candidates were provided with real time cases of hemoglobinopathies for interpretation and then challenges in interpretation were discussed by Prof. Dr. Nisar Ahmed. In 2nd half of day 2, residents performed hands on workshop on transfusion medicine and also solved dry challenges covering all important topics of blood banking. This session of transfusion medicine was coordinated by Dr. Uzma Ata (TIH).


Day 3 started with hands on workshop on coagulation, an essential part of haematology. Dr. Izza Hussain (TIH) was the session co-ordinator for bleeding and thrombotic disorders. Candidates were also given the opportunity to practice dry challenges of coagulation, thromobosis and platelet function disorders. Later queries of candidates were answered by transfusion experts Dr. Saba Jamal and Dr. Ayesha Junaid. Case based discussion on ABO discrepancies and detection and identification of antibodies was carried out by Dr. Saba Jamal and Dr. Zulqarnain Imran respectively. Blood collection, product preparation and quality assurance in blood banking, an integral part of blood banking was highlighted by Dr. Ayesha Junaid. A brief interactive session on fetomaternal hemorrhage (FMH) and case based calculation of FMH alongwith its monitoring and management was conducted by Dr. Neelum Mansoor.




Dry challenges of bleeding coagulopathy was solved by candidates in an interactive session conducted by Dr. Munira Borhany. Assoc. Prof. Dr. Muneeza Junaid elaborated interpretation of platelet function disorders and enabled candidates to understand diagnostic approach of these disorders. Last session was a case based discussion on thrombotic disorders, conducted by Prof. Dr. Bushra Moiz. She provided an insight and understanding of diagnostic approach as well as management of thrombophilia in a detailed way. The questions asked were answered in an elaborated and informative manner hence maintained the interest of candidates till the end.




It was the first event of PSH hosted by TIH and great opportunity for candidates to get maximum benefit from all respected seniors. At the end, Dr. Saba Jamal thanked all the distinguished guests and distributed shields and certificates of appreciation to all speakers, organizers and participants.


SYMPOSIUM

11TH NATIONAL SYMPOSIUM, TIMERGARA - KPK

11th PSH National Haematology Symposium took place in DHQ Hospital Timergara – Dir Lower, KPK on 20th April, 2019. After registration and sitting of Guests the first session began with recitation from Holy Quran. Dr. Fazal Raheem Ex-Senior Pathologist from DHQ Hospital Timergara welcomed the participants. The Chief Guest of the Symposium was Dr. Muhammad Ali, who is the Medical Superintendent of the Hospital. He address the participants and was pleased on having the first ever Haematology Symposium in their city. He further said, that such events should be held frequently so that sharing of knowledge at under privileged areas of Pakistan like Timergara can be possible. The first lecture was on Pancytopenia workup which was delivered by Prof. Dr. Parvez Ahmed who is the President of Pakistan Society of Haematology (PSH). This was followed by a talk on Management of Thalassemia by Prof. Dr. Tariq Mahmood Satti, Commandant of AFBMTC/ NIBMT, Rawalpindi.

Menorrhagia, which is a common problem among women, its management is covered by Dr. Lubna Tahir, Consultant Gynecologists of DHQ Hospital Timergara – Dir Lower, KPK. Here the first session ended all the participants were served with tea and during this time, participants meet the experts of hematology and discussed clinical cases.

The second session started with the lecture by Prof. Nisar Ahmed, Head of Pathology Department, The Children's Hospital & Institute of Child Health, Lahore on the Management of Menorrhagia: Haematological Perspective. This was followed by two another informative lectures, related to Blood Transfusion, Vein to Vein Safety and rational use of Blood Products by Prof. Dr. Saba Jamal from Indus Hospital, Karachi and Prof. Dr. Shahtaj Masood from Hayatabad Medical Complex, Peshawar.

The next distinguished speaker was Dr. Usman Ahmed from Shaukat Khanum Cancer Hospital and Research Center, Lahore, who enlighted the participants about low platelet count, and how to deal with it in pregnant females. Bleeding disorders and how to approach these bleeding disorders was told by Dr. Saima Farhan, Assistant Professor of Paediatric Haematology, The Children's Hospital & Institute of Child health, Lahore. The last presentation was on the most common Hematological disorder, i.e. workup of Anaemia. This topic was covered by Dr. Saima Mansoor, Assistant Professor at The Children's Hospital & Institute of Child Health, Lahore

The souvenirs and shields were presented to Chief Guest, Speakers and organizers. The symposium was concluded by vote of Thanks by Dr. Muhammad Ali, Medical Superintendent. It is anticipated that such workshops and symposiums should be organized in future as well.

CASE REPORT

CHRONIC GRANULOMATOUS DISEASE

Dr Ayesha Iftikhar
AFBMTC/NIBMT, Rawalpindi

A 39-year-old male, resident of Bhakkar, ex- chain smoker, and farmer by profession, initially presented in 2017 with complaints of fever and cervical lymphadenopathy. CBC revealed bicytopenia (TLC 4.5x10^9/L, hemoglobin 9.1g/dL and platelets 80x10^9/L), bone marrow examination was suggestive of chronic granulomatous inflammation. Cervical Lymph node biopsy was consistent with tuberculosis. CECT Abdomen (29/12/2017) showed hepatosplenomegaly (liver 18.7 cm, splenic index 1597) and few enlarged partially calcified mesenteric lymph nodes. He took anti tubercular treatment (ATT) for 06 months for tuberculous lymphadenitis but bicytopenia persisted. On further investigation he was found to have positive Coombs test, was diagnosed as Evan Syndrome and started on steroids and mycophenolate mofetil (MMF) in Dec 2017. His bicytopenia worsened. Repeat Bone marrow examination done in March 2018 and Aug 2018 showed reactive changes. In July 2019 he presented in CMH Lahore with complaints of severe lower back pain and fever for 4 days. Blood CBC a showed pancytopenia (TLC 3.3 x10^9/l, Hb 8.8g/dl, Platelet 26 x10^9/l). Liver function tests showed raised bilirubin (Bilirubin 88 umol/l, ALT 53 IU/l, ALP 918 IU/l). Renal functions were within normal limits. Coombs test was negative (DAT+IAT). Ultrasound Abdomen revealed hepatomegaly and dilated gut loops. MRI Lumbosacral spine (4/7/2019) showed infective spondylodiscitis with edema in right psoas-likely Potts disease. He was again started on ATT (Tab Myrin–P Forte 4-tab OD), MMF 500mg BD and prednisolone 5mg BD continued. The patient was referred to AFBMTC for further management. He presented in AFBMTC OPD on 22 July 2019. Physical examination revealed pallor, CBC showed anemia and thrombocytopenia. Bone marrow examination was repeated and was consistent with Chronic Granulomatous Inflammation. CECT chest, abdomen and pelvis was suggestive of spondylodiscitis with associated right psoas abscess; likely secondary to carries spine and splenomegaly. MMF was stopped and prednisolone were gradually tapered off. He was referred to Infectious disease department for further management of tuberculosis and exclude MDR/XDR TB and advised weekly follow up with blood counts.

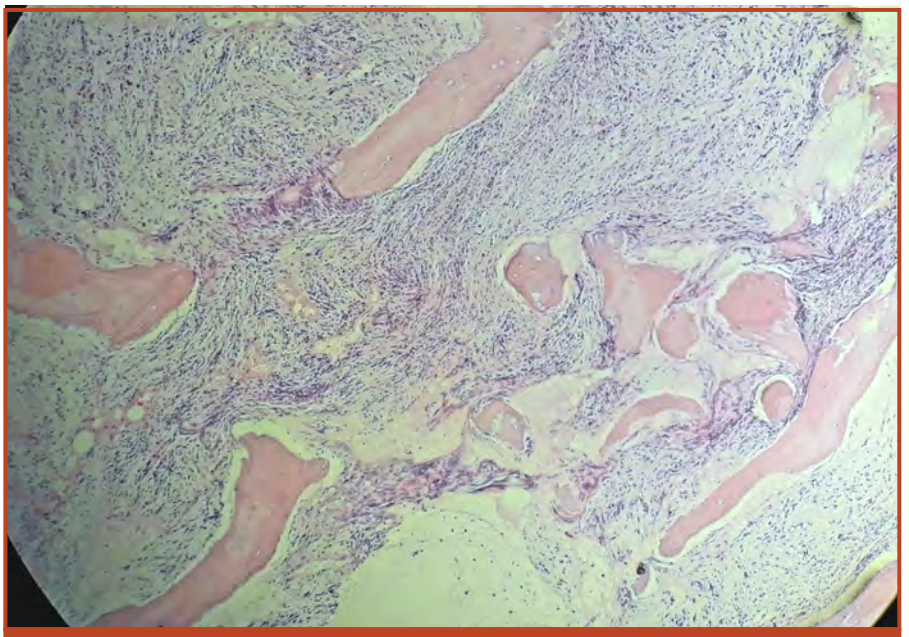


Figure: Bone marrow trephine biopsy showed effaced architecture with ill-defined extensive granulomas and marked fibrosis.

Granulomatous diseases commonly involve multiple systems including the lungs, lymph node, liver and spleen. The finding of granulomas in the bone marrow, however, is uncommon with a

reported incidence between 0.3% and 2.2%[1]. This nonspecific finding is indicative of a systemic process with an extensive list of possible causes including viral, bacterial and fungal infections; malignant diseases; autoimmune diseases; drugs and sarcoidosis. The vast majority of cases of bone marrow granulomas are attributed to infectious sources and hematologic malignancies. In a study carried out in India over a span of 5 years (2012-2016) also found that maximum number of cases were associated with granulomas in the bone marrow were infectious (HIV & TB) followed by sarcoidosis[2]. In another study carried out in China in January 2018 out of 11,339, 110 cases showed granulomatous lesions in the bone marrow biopsies (0.97%)[3]. Etiologies were identified in 80 cases (72.8%), with infections being the most common (64.5%), followed by malignancies (4.5%) and autoimmune diseases (3.6%)[3]. Among infectious cases, 87.32% (62/71) cases were diagnosed as TB, a positive acid-fast stain or/and polymerase chain reaction (PCR) result for mycobacterium TB DNA fragment amplification was obtained for 35 cases and in 30 cases (27.27%), a definite diagnosis could not be established.[3]

All hematologic cell lines may be affected. Bone marrow fibrosis may result from proliferation of macrophages (which are abundant in normal marrow) after they engulf mycobacteria[4]. This proliferation damages the marrow microenvironment with a resultant diminution in cell lines, and this is termed myelophthisis[4]. Myelofibrosis with myelophthisis can accompany miliary tuberculosis, cavitary pulmonary tuberculosis and granulomatous involvement of the spleen, lymph nodes and liver[4]. Myelophthistic anemias are characterized by the presence of tear-drop erythrocytes, nucleated red cells and early granulocytes on the peripheral blood smear[4]. Bone marrow aplasia may complicate miliary tuberculosis[4]. However, the anemia or pancytopenia seen in this setting is usually secondary to marrow dysfunction rather than aplasia[4]. Additional mechanisms include direct bone marrow infiltration with granulomata, and the development of amyloidosis of the marrow[4].

References:

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CASE REPORT

LANGERHAN CELL HISTIOCYTOSIS

Dr Maryam Khan
AFBMTC/NIBMT, Rawalpindi

A 43 years male, soldier developed soft erythematous, cystic scalp eruption in October, 2018. He visited a local doctor who prescribed some topical medication and it reduced in size. This was followed by another lesion at the same site in May, 2019. He also complained of disabling back pain, headaches, excessive thirst, mood swings and weight gain. The patient had past history of traumatic nasal fracture, right knee medial meniscal tear and mixed anxiety depressive disorder. He did not smoke or drink. He was on antidepressants and mood stabilizers. On physical examination he was obese and had two 0.5 x 0.5 cm lesions on right parietal area of scalp. His blood results? were unyielding, skeletal survey showed a well-defined oval shaped exophytic lytic lesion in right iliac bone causing cortical thinning. Scalp lesion biopsy showed a dermal nodular growth with overlying thinned out epidermis, epidermis surrounding this area was hyperplastic. The nodule was bordered by hair follicles and comprised of Langerhan cell histiocytes dispersed in sheets with numerous eosinophils, macrophages and lymphocytes in between. Immunohistochemistry showed CD 1a, S 100, CD 68-Positivity in Langerhan cells. He was diagnosed as Langerhan Cell histiocytosis. He underwent staging bone marrow and Contrast enhanced CT scan of chest, neck and abdomen. Bone marrow showed CD1a and Langerin positive cells. CECT showed no organomegaly or any organ involvement, MRI brain and pituitary function studies were normal. A diagnosis of Langerhan Cell histiocytosis (High risk) was made.

Literature Review: Langerhans-cell histiocytosis (LCH) is the most common histiocytic disorder. Granulomatous lesions comprising langerin-positive (CD207+) histiocytes and an inflammatory infiltrate can arise in virtually any organ system but have a particular affinity for bone, skin, the lungs, and the pituitary gland. The annual incidence of LCH has been reported to be 4.6 cases per 1 million children under 15 years of age, with a male-to-female ratio of 1.2:1 while estimated incidence among adults is 1 to 2 cases per million, though LCH is probably underdiagnosed in this population. LCH has a widely variable clinical presentation, ranging from single indolent lesion to explosive multisystem disease. Children with liver, spleen, or bone marrow involvement are at highest risk for death from LCH and are therefore classified as having high-risk LCH. Studies for newly diagnosed LCH patients include skull series, skeletal survey, chest radiograph, complete blood count, liver enzymes. Ultrasound or MRI may identify lesions in patients with suspected liver involvement. CT scanning is useful to assess lesions in the orbit, mastoid, sphenoid, and temporal bones. MRI is effective in evaluating lesions in brain, pituitary, vertebrae, spinal cord, and pelvis. PET scans are also effective to screen for the lesions. Bone marrow biopsy and aspirate is indicated in patients younger than 2 years old or any patients with cytopenias. Endoscopy with biopsy on patients with evidence of malabsorption may be useful.

LCH in adults can arise as a component of mixed histiocytic disease Erdheim-Chester disease (ECD), with mixed phenotype in the same lesion or as a distinct phenotype of separate lesions. ECD generally has poorer outcome than LCH and may benefit from alternative initial therapy. PET scan, MRI of chest (cardiac involvement), abdomen (kidney and aortic involvement), and legs (tibial involvement), and biopsy of multiple accessible lesions in adults with LCH may be informative to identify patients with

CASE REPORT

simultaneous ECD. Therapies for skin limited LCH include topical steroids, nitrogen mustard, or imiquimod; surgical resection of isolated lesions; phototherapy; systemic methotrexate, 6-mercaptopurine, vinblastine/vincristine, thalidomide, cladribine, and/or cytarabine. Isolated bone lesions can be effectively treated with limited curettage and/or corticosteroid injection.

Large pelvic lesions or vertebral lesions not amenable to curettage may be treated with systemic therapy. Radiation therapy may be effective in older children with single vertebral lesions that have not caused complete collapse of the vertebra or with a lesion in the greater trochanter of the femur at risk for pathologic fracture. Patients with single bone lesions have only a 10% chance of reactivation. Gross total resection of LCH bone lesions is not only unnecessary but harmful, impeding the remodeling that occurs when margins remain intact. The current standard of care as determined by results from the LCH-III trial is to treat these patients with vinblastine/prednisone for 1 year. The frequency of reactivation was significantly decreased by one-year vs 6 months of therapy, suggesting that further prolongation of this mild treatment may further reduce reactivations. This is being tested in the LCH-IV trial. Adults with either multifocal bone disease or lesions in risk organs are treated with cytarabine (100 mg/m^2 per dose × 5 days per month × 12 months) Patients with CNS lesions, CNS-risk lesions, or progressive neurodegeneration are treated with a higher dose (150 mg/m^2 per dose). Adults treated with Vinblastine/prednisone according to standard pediatric protocol had near universal toxicity and sub optimal efficacy.

UPCOMING

UPCOMING

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VIEWS & NEWS

The PSH "Haematology Updates" is
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