LETTERS TO THE EDITOR

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3. Raghuveer TS, Srinivasa R, Chandrasekhar

Ensuring Correctness of Bone Marrow Reports in Infants: Role of the Pediatrician

Pediatricians generally have little to do with the way bone marrows (BM) are reported by pathologists. This communication is to emphasize the fact that BM of young children, particularly infants, constitutes a special group, where an active involvement of the pediatrician can ensure correctness of the report.

The BM of infants and young children differs from that of normal adults in having up to 75% (mean + 2 SD) lymphocytes and transitional cells(1). These cells, also called hematogones(2), have morphological features that cause them to be often wrongly interpreted as blasts. This occurs typically in two settings. The first is a BM examination done to look for a hematological or non-hematological malignancy. The second setting is evaluation for remission of a child on therapy for acute lymphoblastic leukemia (ALL). The error here is because of a phenomenon, not very common, known as rebound lymphocytosis, wherein lymphocytes and hematogones come to dominate the BM 6-24 months post-therapy(2,3).

In most of our cases where a mistake had occurred due to rebound lymphocytosis, the follow-up BM examination had been done outside in another hospital and the patient had returned to us after the marrow had been reported as being in relapse. Unfortunately, in all but an occasional of these cases, the pediatrician outside had accepted the wrong diagnosis. Morphological features of the BM cells, however, made it apparent to us that the cells were not blasts and a hematological follow-up resolved the issue.

Preventing such errors is obviously crucial. Providing full clinical information to the pathologist helps, as does firm dependence on ones clinical judgment. However, what would help most, is a high index of suspicion among pediatricians. The greatest problem in this, in our view, is a rather poor awareness because pediatrics texts generally do not discuss BM transitional cells/hematogones(4). Books that do, often do not emphasize these cells in the proper context where it matters most, i.e., in relation to management of hematological and non-hematological malignancies(5). Emphasis of great practical value is present only in a few monographs which are liable not to be consulted by many practising physicians.

The unique feature of bone marrow of
young children and infants and their practical significance therefore needs to be emphasized in the teaching of pediatrics so that pediatricians can interact meaningfully with pathologists in this somewhat difficult area of bone marrow interpretation.

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Megalocornea–Mental Retardation Syndrome

A 42-month-old girl presented with global developmental delay and an abnormally large right eye. She was the product of a consanguineous marriage. She had a history suggestive of hypotonia since infancy with delayed developmental of milestones. Examination revealed microcephaly and short stature. The right eye had megalocornea (corneal diameter >13 mm) and a normal iris. The left eye was normal. She had preaxial polydactyly of the right hand. Apart from hypotonia, the rest of the neurological and systemic examination was normal. Development assessment in the individual domains by the Early Developmental Profile were 19 months in gross motor, 23 months in fine motor, 21 months in cognition, 9.8 months in language, 24 months in social/emotional and 16.3 months in feeding. The development quotient was 44.9 with a disproportionate delay in language.

On investigation, her bone age was normal and there was no skeletal abnormality except the polydactyly. BERA revealed bilateral sensorineural deafness, Investigations revealed no other associated abnormalities. A diagnosis of Megalocornea-Mental retardation syndrome (type 5) was made.

Megalocornea-Mental Retardation syndrome (MMR) was first reported by Neuhaus, et al.(1). It is an extremely rare disorder with autosomal recessive inheritance. Most cases appear sporadically. The syndrome is characterized by distinctive ocular abnormalities,