

T-Lymphoblastic Leukaemia (T-ALL)

Pages with reference to book, From 358 To 260

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Cases with this association have been reported under different names such as Steinberg sarcoma, childhood lymphoblastic or T-cell lymphoma, lymphoma of the convulated lymphocytes, ALL with an anterior mediastinal mass and acid phosphatase-positive acute lymphoblastic leukaemia. To quote here from an excellent review by Cooke¹ : On reviewing the individual clinical and pathological reports it is noticeable that the only feature by which they differ from a similar series of cases of acute leukaemia is by the presence of the mediastinal tumour. Clinical manifestations, blood picture eventual glandular and splenic enlargement, extensive infiltration of the bone marrow, liver and kidneys by non-granular mononuclear cells are identical whether or not a mediastinal mass is present. Cooke¹ described the main features of the disease, namely predominance of males over females, an incidence almost always below the age of 30 years (mean 16 years), and in two-thirds of the cases where haematological data were available WBC count was over 100x10⁹/Litre. She also noted that the thymic tumour may precede by several weeks the development of ALL, or that both features may be present ab initio. It is now acknowledged that an initial presentation as an acute leukaemia or with lymphadenopathy but without mediastinal enlargement is also possible^{2,3,4}

The speed with which the leukaemic picture may develop is graphically illustrated by one of Cooke's cases where a WBC count of 21x10⁹/Litre with 72% neutrophils changed a week later to 140x10⁹/L with 89% lymphoblasts. A high frequency (70%) of leukaemic transformation in children with malignant lymphomas (non-Burkitt type) has been reported⁵ , particularly in those cases with mediastinal involvement meningeal leukaemia was seen in half of their cases with bone marrow involvement. The marked predominance of males and the higher incidence of T-ALL in older children as compared with Null-ALL has been confirmed by most of the recently published series^{6,7} A'so splenomegaly is a more constant feature in T ALL than Null ALL⁸.

T-ALL thus appears to be associated with factors known to be of bad prognosis in ALL in general. Meningeal leukaemia, presumably related to the high initial WBC counts, is commonly early event Interesting observations have been made in relation to the worse prognosis in T-ALL⁷. The remission rate to conventional ALL treatment was only 40% in T-ALL compared with 89% in other ALL cases. When comparing groups of similar WBC counts and organomegaly (except for mediastinal mass) similar drug schedules resulted in earlier relapses and shorter survivals in T-ALL, thus suggesting an intrinsically more malignant nature for T-cell neoplasm apparently not accounted for only by the noted differences in WBCs. ALL with a thymic mass appears to have in addition to high WBC at presentation, higher haemoglobin and platelet counts than Null-ALL, thus suggesting a better preservation of the normal bone marrow and that the tumour cells may not have arisen from the bone marrow itself as they seem to in the common type or Null-ALL.

The importance of recognising T-ALL as a distinct clinicopathological and immunological variant of ALL, is emphasised from the fact that in some studies on cell markers in ALL, cases presenting with an anterior mediastinal mass were excluded⁹, however some months later the same group of workers included such with positive T-markers as examples of immunoblastic acute lymphoid leukaemia. It is perhaps only of semantic interest to decide whether the disease could be considered primarily a leukaemia or a lymphoma and which is the right nomenclature. This in fact may depend on whether the initial diagnosis (which may depend on the presenting clinical features) is made by a histopathologist, a haematologist or even an immunologist. The almost inevitable development of ALL in patients presenting with thymic tumours, the fact, perhaps equally is that as many will already have ALL at the

time of diagnosis, and the knowledge that some may not have a mediastinal mass at all, suggest that the disease should be considered primarily a leukaemic process-hence the suggested name: 'T-ALL'. As commented above, the leukaemic manifestations are even more florid than in other cases of ALL. The 'T' preceding the abbreviation 'ALL' is necessary as it denotes the likely thymic origin and the lymphoid nature of the cell target for the malignant process.

References

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