LATE ONSET OF DIFFERENTIATION (RETINOIC ACID) SYNDROME – CASE REPORT

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Abstract

The differentiation syndrome (DS), previously called "all-trans retinoic acid syndrome" is a potentially fatal complication observed in patients with acute promyelocytic leukemia (APL) receiving induction therapy with all-trans-retinoic acid (ATRA) or arsenic trioxide (ATO). It is characterised by fever, peripheral edema, pulmonary infiltrates, respiratory distress, hypoxemia, hypotension, pleural and pericardial effusions, renal and hepatic dysfunction. Its pathogenesis is not completely understood, but it is believed that the release of inflammatory cytokines from malignant promyelocytes is the main phenomenon involved.

A bimodal incidence of DS has been observed, with a majority of cases occurring during the first week of ATRA treatment ("early" DS), but a substantial number of cases occurring during the third or even fourth week of ATRA treatment ("late" DS).

We report a patient with newly diagnosed APL treated with the AIDA regimen who experienced late DS, a situation where differential diagnosis was difficult because of patient medical history and absence of well defined diagnosis criteria for DS.

Due to the morbidity and mortality associated with DS, its early recognition and aggressive management are essential.