PROGNOSIS

HTSL is an extremely aggressive NHL with rapid progression. Most patients die within two years of diagnosis despite aggressive, multiagent chemotherapy, due to resistance, relapses and advance stage at presentation. Degree of thrombocytopenia is the only established adverse prognostic factor, directly linked to relapse and poor overall survival. Post solid organ transplant patients have worse prognosis with dismal overall survival[1,3].

TREATMENT

The disease shows marked chemoresistance to most of the currently used multiagent chemotherapeutic regimens, none of which have been established as a standard. Anthracycline based regimens and CHOP/CHOP- like regimens have been used for satisfactory early induction followed by a combination of cytarabine and cisplatin. Newer agents like Pentostatin and Alemtuzumab have shown promise with adhesion molecule CD44, emerging as a potential for targeted therapy. All these cases should be evaluated for eligibility for hematopoietic stem cell transplantation (SCT), which remains the only potential cure[1].

CONCLUSION

HTSL is a rare, extremely aggressive form of NHL, with a median overall survival of 11 months. This variant of NHL warrants intensive induction chemotherapy followed by early high-dose therapy and hematopoietic stem cell transplantation which remains the only potential curative therapeutic option. Despite extensive treatment, complete remission is uncommon and relapses which occur, usually present with chemoresistance. A proper knowledge of this entity is necessary for an early and prompt diagnosis, so that the patient can be aggressively treated and considered for SCT. A standard treatment regimen hasn't yet been established and a proper documentation of such cases can be effective in standardising a validated, uniform therapeutic approach for patients with HTSL.