

To the best of our knowledge, this is the largest study till date describing SVCO at presentation in ALL [2,4]. We observed a very high incidence of SVCO at presentation (in contrast to 0.27% reported in a study from America) and identified a range of associated adverse and high-risk factors [4]. Additionally, SVCO had an inferior outcome in contrast to 67% and 50% survival reported earlier [2,4]. Further large scale and collaborative studies to confirm these observations and assessing the molecular and cytogenetic characteristics of this unique presentation are necessary. Reinforcement and implementation of standard management guidelines, educational initiatives, better supportive care and management of anticipated complications (respiratory compromise, thrombosis, tumor lysis syndrome, ICP) should improve the outcome [5].

Although uncommon, oncologists and pediatricians are likely to encounter a child with ALL presenting as SVCO and need to be aware of this potentially treatable medical emergency.

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Aggressive Natural Cell Leukemia in an Infant with Bilateral Testicular Mass

We read case report on long term survival in aggressive NK cell leukemia by Patel, *et al.* with great interest [1]. It is a very rare entity in pediatric population [2-4]. Recently, we also managed a similar case in an infant.

A 9-month old boy presented with recurrent respiratory tract infections since 2 months, swelling of both eyes since 2 months, bilateral testicular enlargement noticed since one month. At admission he had pallor, bilateral proptosis, hepatosplenomegaly and bilateral testicular enlargement. His complete blood counts showed pancytopenia (hemoglobin-7.7g/dL, total leukocyte count-5000/cumm, platelet-100,000/cumm,

absolute neutrophil count-350/cumm). CT abdomen did not reveal any mass. Bone marrow aspirate and biopsy showed presence of infiltration with MPO negative malignant cells, which could not be further characterized morphologically. Flow cytometry analysis failed to pick up the lineage of the cells. Testicular biopsy revealed diffuse infiltration by a round cell tumor with brisk mitotic activity. Tumor cells showed positive staining with CD45RO, CD43, CD99 and CD56. These were focally positive for LCA. Staining for CD3, CD20, CD10, MPO and ALK-1 was negative. It was opined to be aggressive NK cell leukemia (ANKL). Bone marrow cytogenetics showed trisomy of chromosome 8. FISH studies for MLL, BCR-ABL and TEL-AML were negative. Cerebrospinal fluid (CSF) was negative for any blasts. He received chemotherapy as per Interfant-99 protocol [5]. He was in clinical remission (CR) on day 33 of induction. He relapsed in bone marrow and CSF sixteen months from diagnosis during maintenance phase of therapy.

Diagnosing ANKL can be a challenge for the clinician, pathologist and haematologist. Our patient did not respond well to chemotherapy in contrast to report by Patel, *et al.* [1] although initial response to therapy was good. Infants with MLL gene rearrangement have poor prognosis [5], which was absent in our case. ANKL with bilateral testicular mass and trisomy 8 in an infant has not been reported previously. It is interesting to note that the outcome for the pediatric patients reported in the literature is somewhat better than that of their adult counterparts, with 7 of 13 children surviving at the time of last follow-up (54%) [3]. Most children undergo stem cell transplant (SCT) as salvage at relapse except one case who had SCT in CR1 and is alive at last follow up [2]. ICE chemotherapy has been suggested to be more effective for NK cell malignancy [3].

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