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REFERENCES

1. Mathur C, Stigler MH, Perry CL, Arora M, Reddy KS. Differences in prevalence of tobacco use among Indian urban youth: The role of socioeconomic status. *Nicotine Tob Res.* 2008;10:9-16.
2. Singh V, Pal HR, Mehta M, Dwivedi SN, Kapil U. Pattern of tobacco use among school children in National Capital Territory (NCT). *Indian J Pediatr.* 2007;74:1013-20.
3. Madan Kumar PD, Poorni S, Ramachandran S. Tobacco use among school children in Chennai city, India. *Indian Journal of Cancer.* 2006;43:127-31.
4. Ashraf A, Quaiyum MA, Ng N, Van Minh H, Syed AR, Ahmed M, *et al.* Self-reported use of tobacco products in nine rural INDEPTH Health and Demographic Surveillance Systems in Asia. *Global Health Action* 2009 Sep 28; Supplement 1:19-27.
5. Narain R, Satyanarayana L. Tobacco use among school students in India: the need for behavioral change. *Indian Pediatr.* 2005;42:732-3.

Superior Vena Cava Obstruction in Childhood Acute Lymphoblastic Leukemia

Etiology, complications and management protocols for superior vena cava obstruction (SVCO) in children are well recognized [1]. However, there is paucity of literature addressing SVCO at presentation in childhood acute lymphoblastic leukemia (ALL), its association with risk factors, prognostic impact and outcome [2]. Herein, we describe our management experience of 24 (3.14%) children with ALL presenting with SVCO at diagnosis out of 762 patients managed at our center from January 1990 to December 2005 and followed until January 2009 [3]. Therapy protocols were modified United Kingdom ALL (UKALL) X and XI.

Dyspnea, orthopnea, stridor, facial puffiness with plethora and venous engorgement along with constitutional symptoms of fever, pallor, fatigue and bleeding were the most common presenting features. The diagnosis of ALL was confirmed in all patients by bone marrow examination. The extent of disease was delineated by computed tomography in patients presenting after 2000 with cautious use of procedural sedation where needed.

Four had frank superior mediastinal syndrome (SVCO with significant and manifest tracheal compression with respiratory distress or failure) while 1 presented with raised intracranial pressure (ICP). Emergency radiotherapy was not used in any of these patients. SVCO was managed as per standard guidelines [1].

On comparison with other ALL children and those in Continuous complete remission, children presenting with SVCO at diagnosis had significantly higher incidence of hepatomegaly, splenomegaly, lymphadenopathy, bulky disease, mediastinal adenopathy, overt testicular disease and high total leukocyte count at presentation. Ten (47.1%) had hyperleukocytosis. In contrast, age, gender, symptom diagnosis interval, hemoglobin and platelet counts at diagnosis were similar in the 3 groups.

Seventeen patients opted for therapy while 7 cited socioeconomic reasons for therapy refusal. 4 (2 combined, 1 bone marrow and 1 central nervous system) relapsed, 4 died (within 1 day to 3 weeks from diagnosis), 4 were lost to follow up or defaulted therapy. There were 5 (29.4%) survivors at median follow up periods of 41 months (range: 29-84 months). The survival outcome was significantly inferior as compared to the entire cohort ($n=762$) by univariate log-rank analysis ($P=0.045$) (3). However, SVCO at presentation did not have independent prognostic impact ($P=0.12$) in multivariate Cox-regression analysis.

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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REFERENCES

1. Arya LS, Thavaraj V, Kulkarni KP. Oncological emergencies. *In: Chaudhary P, Bagga A, Chugh K, Ramji R, Gupta P, editors. Principles of Pediatric and Neonatal Emergencies: 3rd ed. New Delhi: Indian Pediatr. (In press)*
2. Arya LS, Narain S, Tomar S, Thavaraj V, Dawar R, Bhargawa M. Superior vena cava syndrome. *Indian J Pediatr. 2002;69:293-7.*
3. Kulkarni KP, Marwaha RK, Trehan A, Bansal D. Survival outcome in childhood ALL: experience from a tertiary care centre in North India. *Pediatr Blood Cancer. 2009;53:168-73.*
4. Ingram L, Rivera GK, Shapiro DN. Superior vena cava syndrome associated with childhood malignancy: analysis of 24 cases. *Med Pediatr Oncol. 1990;18:476-81.*
5. Hon KL, Leung A, Chik KW, Chu CW, Cheung KL, Fok TF. Critical airway obstruction, superior vena cava syndrome, and spontaneous cardiac arrest in a child with acute leukemia. *Pediatr Emerg Care. 2005;21:844-6.*

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.