Pediatric Ocular Malignancy: A major challenge in developing countries

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Dear Friends,

Season's Greetings!!

Wide verities of intraocular and orbital malignant neoplasms are present in pediatric population. Like many pediatric cancers, ocular malignancy remains a major challenge in developing countries. Although pediatric ocular malignancies are rare, but delay in diagnosis and management can lead to deformity, loss of vision and mortality as well. It can occur primarily or as secondary and metastatic lesions. In developing countries, secondary metastatic lesions such as orbital retinoblastoma, neuroblastoma, Burkitt's lymphoma and myeloid sarcoma (AML) are more common orbital malignancies as compared to primary lesions[1]. The incidence of pediatric orbital malignancies ranges from 8-18% among all orbital lesions[2,3].

Rhabdomyosarcoma is the most common malignant orbital tumor of childhood. It accounts for about 5% of all cancers in pediatric population. Retinoblastoma (RB) is the most frequent and but highly curable malignant intraocular tumor in children. Estimated cumulative annual incidence of RB is 53-62 per million children younger than 14 years[4]. In developing countries like India such malignancies are reported late in advance stage, often requiring enucleation and associated with high mortality rate. The survival rate of RB reported in US reaches up to >90%[5], in China 81%[6], in India 48%[7] and in Africa 20-40%[8]. Management of pediatric ocular malignancy is complex and challenging, requiring coordinated, multidisciplinary specialized team work at a tertiary centre including ocular oncologist, pediatric oncologist, interventional radiologist and ocular pathologist. The treatment of retinoblastoma has undergone tremendous advancement in developed countries, over the past few decades by globe salvage therapy like chemo reduction by systemic chemotherapy, super selective ophthalmic artery chemotherapy infusion or intravitreal injection of Melphalan and laser treatment. Recent advancement in treatment of RB is not only targeted to neoplastic cells but it also target dynamic tumor microenvironment which consists of cytokines, growth factors, free radicals and tumor associated macrophages etc. which play important role in tumor growth, treatment resistance, metastasis and recurrence[9].

In developing countries, most of the children with retinoblastoma present with advanced stage disease and associated with high mortality[10,11]. The poor outcome is due to late diagnosis, inadequate infrastructures and lack of government support. In our country delayed diagnosis is due to illiteracy or lack of awareness and cultural and economical barrier. Improvement in survival in developing countries will requires certain measures like early reporting through large public health education campaign, and training of primary school teachers and rural health care workers, to formulate early and cheaper means of screening those at risk. Need of collaborative cancer research projects and clinical trials of cheaper and effective drugs with maximum participation by a permanent consortium of internet-linked university based and practice based ocular oncologist will felicitate management of RB. There is great need of national retinoblastoma registry, well organized national programme and development of retinoblastoma/pediatric cancer centre of excellence.

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