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Case Report

Look into their eyes and save their vision: role of primary health care provider in early diagnosis of retinoblastoma

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ABSTRACT

Childhood cancer is not amenable to preventive or screening strategies. The most effective strategy to reduce the cancer burden and improve outcomes, is to focus on early, correct diagnosis followed by evidence-based therapy. When diagnosed early, they are responsive to appropriate therapy and increases survival rates, thereby reducing the need for intensive treatment and reduces the expenditure per child. Retinoblastoma is the most common intra ocular malignancy and is one of the most curable cancers in children. If diagnosed early and treated optimally, not only they are completely cured but a vast majority can have ocular salvage and retain vision. Lack of awareness among general population, diagnostic delays, delays in referral to proper treatment centre, compounded by socio economic factors attributes to the poor outcome in such children. We report a case of 2 years 10 months old girl child whose initial presentation of leukocoria was missed and later presented with advanced retinoblastoma with CNS metastasis.

Keywords: Primary health care provider, Red eye reflex, Retinoblastoma, Early diagnosis

INTRODUCTION

India is currently in a state of transition, economically and epidemiologically in terms of health. We have made enormous strides over past few decades. Still communicable diseases continue to remain a major health problem. Non communicable diseases contribute to 62% of total deaths in India, but with limited resources and infrastructure, focus of health care policy is still onwards communicable diseases, reproductive health and malnutrition.

Low middle income countries including India account for 80% of childhood cancer burden.[1] In India, non-communicable diseases accounts for half of all deaths in age group of 5-14 years.[2] Childhood cancer in India accounts for 0.7-4.4% of total cancer diagnosis.[3] In spite of significant improvement made in childhood cancer services in last few decades, outcome when compared to global standards still remains modest in India. Significant barriers are delays in recognition, diagnosis and cure.

Among all the childhood cancers retinoblastoma provides a unique opportunity of identifying it at an earlier stage when the disease is curable. Retinoblastoma is the most common intraocular malignancy of infancy and childhood. The overall survival rate of retinoblastoma is reported to be more than 95% when diagnosed in intraocular phase.[4] But delayed diagnosis and treatment leads to extraocular metastasis, visual loss and death. The mortality rate in advanced cases, is reported to be as high as 50-90%, in developing countries.[5]

We report a case of CNS metastatic retinoblastoma who had facial disfigurement and fungating bleeding ulcer of left eye. Palliative chemotherapy caused significant

reduction in size of the lesion and improved the quality of life of child. The case highlights the need for increasing the awareness among public and primary health care providers for early recognition of retinoblastoma.

CASE REPORT

A 2 year 10 months old girl child, was noticed to have white reflex in left eye when she was 1 year old. She was taken to a nearby primary health centre and advice nutritional supplements for the same. But with no clinical improvement, she was taken to nearby higher ophthalmology centre, 8 months later and was managed as traumatic cataract and was on regular follow up. Six months later, she developed progressive swelling of left eye and was taken to back to same centre and advised CT scan, which could not be done due to covid lock down. The child was later taken to an ophthalmology centre elsewhere in October 2020. Imaging study done revealed metastatic retinoblastoma of left eye. Child was advised high dose chemotherapy. The family has been struggling to organize funds to initiate chemotherapy when the swelling involved entire left face causing disfigurement and foul smell emanating from the ulcerated lesion. From first week of December, she became lethargic, had poor oral intake and was bed ridden. The parents were withdrawn due to the social stigma of having a child with foul smelling and pus discharging ulcerated lesion (Figure 1). With the help of local community service organization, who arranged some funds, she brought to tertiary care ophthalmology centre in Chennai. MRI brain done showed left eye orbital retinoblastoma with metastatic deposits in mandible, maxilla, zygoma, frontal, temporal and parietal epidural spaces. CSF analysis showed positive malignant cells. Family was counselled about the poor prognosis. In view of physical suffering of child, case was discussed in a multi-disciplinary meet and after approval from tumor board planned for palliative chemotherapy with family's consent. Chemotherapy and supportive care were provided with fund support from the hospital. After the first cycle of chemotherapy, there was significant reduction in swelling of eye (Figure 2). Child's general condition and appetite improved and she became active once again. After 3rd cycle, proliferative growth was restricted to eye (Figure 3).

Figure 1: Ulcerated, pus discharging lesion of left eye.

Figure 2: After first cycle of chemotherapy.

Figure 3: After 3rd cycle of chemotherapy.

DISCUSSION

Around 43% of global burden of retinoblastoma children live in 6 countries of Asia-pacific region-India, China, Indonesia, Pakistan, Bangladesh and Philippines. Around 1500 new retinoblastoma cases are diagnosed in India, every year. But the awareness about retinoblastoma is very low in developing countries. The poor survival rates in underdeveloped nations are being attributed to inadequately trained primary health care workers, lack of diagnostic facilities at regional centres, delays in referrals, families' belief in indigenous medicines, availability of specialized treatment facilities only in cities and financial constraints.[6] In developing countries,

orbital extension is a major cause of death in children with retinoblastoma.

Leukocoria remains the most common presentation of retinoblastoma followed by strabismus, proptosis, visual loss and red eye.[7] Lack of awareness among parents and primary health care providers about leukocoria results in seeking medical management, when the child has already lost vision or has an advanced stage of disease.

As early diagnosis and timely treatment are vital for better prognosis retinoblastoma, increasing public awareness, organizing education programmes for pediatricians and ophthalmologists and improving health care accessibility are the need of the hour. The level of awareness of first contact health provider in identifying and making appropriate referral is critical. Hence the need to conduct awareness programmes among general physicians about early signs of retinoblastoma. Posters depicting various symptoms of retinoblastoma can be displayed at all primary health care centres and private clinics to increase the awareness among general public. Red eye reflex examination should be made compulsory in all newborns and all children up to 5 years of age, when they are brought for immunization or for any minor illness.

In some of the developing nations, when the educational programme was linked with vaccination programmes, the results were encouraging, as the rate of orbital retinoblastoma cases reduced to half in the post campaign period. [8]

CONCLUSION

As orbital retinoblastoma is more common in developing countries, implementation of cost-effective measures like increasing awareness among public and health care physicians will help in reducing the mortality of this highly curable pediatric malignancy.

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REFERENCES

1. Bhakta N, Force LM, Allemani C, Atun R, Bray F, Coleman MP et al. Childhood cancer burden: a review of global estimates, Lancet Oncol. 2019;20(1):e42-53.
2. Fadel SA, Boschi-Pinto C, Yu S, Reynales-Shigematsu LM, Menon GR, New Combe L. et al. Trends in cause-specific mortality among children aged 5-14 years from 2005 to 2016 in India, China, Brazil, and Mexico: an analysis of nationally representative mortality studies. Lancet. 2019;393(10176):1119-27.
3. Three Year Report of the Population Based Cancer Registries 2009-2011: Report of 25 PBCRs, National Cancer Registry Programme, Indian Council Medical Research, Bangalore. 2013.
4. Huang D, Zhang Y, Zhang W, Wang Y, Zhang P, Hong L et al. Study on clinical therapeutic effect including symptoms, eye preservation rate, and follow-up of 684 children with retinoblastoma. Eur J Ophthalmol. 2013;23:532-8.
5. Badhu B, Sah SP, Thakur SK, Dulal S, Kumar S, Sood A et al. Clinical presentation of retinoblastoma in Eastern Nepal. Clin Exp Ophthalmol. 2005;33:386-9.
6. Chawla B, Kumar K, Singh AD. Influence of socioeconomic and cultural factors on retinoblastoma management. Asia Pac J Oncol Nurs. 2017;4:187.
7. Kaliki S, Patel A, Iram S, Ramappa G, Mohamed A, Palkonda VKR. Retinoblastoma in India: clinical presentation and outcome in 1,457 patients (2,074 eyes), Retina. 2019;39(2):379-91.
8. Leander C, Fu LC, Pena A. Impact of an education programme on the late diagnosis of retinoblastoma in Honduras. Pediatr Blood Cancer. 2007;49:817-9.

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