Editorial

Management of Retinoblastoma in Zambia

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Received: August 20, 2016, Accepted: April 05, 2017

The Cancer Diseases Hospital (CDH), which was opened in 2006, is Zambia's only oncology referral health facility offering health services for oncology, including pediatric oncology. It is Zambia's only radiation therapy center providing treatment to all cancer patients not only within Zambia but also from some of the other Sub-Saharan countries such as Democratic Republic of Congo, Ethiopia, Malawi, Angola, Zimbabwe, and Botswana among others. The hospital provides chemotherapy and radiation therapy services in addition to diagnostic imaging (computed tomography, magnetic resonance imaging, ultrasound, and X-ray) and laboratory facilities. It is a 252-bed facility with a pediatric oncology unit that has two dedicated pediatric wards (25 beds each) catering for all the pediatric oncology patients referred from various institutions across the country. However, there remain inadequate human resources to provide quality patient care, including a typical nurse to patient ratio of 1:15, only one subspecialty-trained pediatric hemato-oncologist with limited opportunities for doctors or nurses to obtain further subspecialty training abroad.[1]

Kitwe Central Hospital on Copperbelt Province of Zambia with a dedicated ophthalmology unit and St. Francis Mission Hospital in Eastern Province provide surgical treatment for retinoblastoma but not chemotherapy or radiation therapy. A number of patients therefore are referred to CDH from these hospitals after surgery for chemotherapy and radiation therapy.

Retinoblastoma is a malignancy affecting the retina of the eye and commonly occurs in children aged 3 years and below. It is a common childhood malignancy among African children,[2] and Zambia is no exception. According to Slone *et al*. in 2014[1] retinoblastoma made up 17% of all documented childhood malignancies seen at the pediatric oncology ward of the University Teaching Hospital (UTH).

The global incidence of retinoblastoma in children under 5 years is estimated at 11 cases per million.[3] Only about 5% of patients who develop this disease have a positive family history; the rest often have no positive family history of the disease. The most commonly occurring type is unilateral disease, with bilateral disease not being very common.[2] In Zambia, bilateral disease accounts for about a quarter of the cases referred to the UTH, most of whom are below 36 months at presentation. The average age at presentation is 32 months, with a few cases presenting aged 5 years and above.

Clinical Presentation

The clinical presentation of retinoblastoma is usually with leukocoria in more than half of the patients,

Access this article online	
Quick Response Code:	Website: www.apjon.org
	DOI: 10.4103/apjon.apjon_24_17

Cite this article as: Maliti BK. Management of retinoblastoma in Zambia. Asia Pac J Oncol Nurs 2017;4:184-6.

strabismus, deteriorating vision, changes in pupil size, proptosis as the disease progresses (especially in developing countries), and secondary changes such as glaucoma, retinal detachment, and orbital inflammation due to tumor necrosis.[3,4] In Zambia, most of the patients presented with proptosis having had leukocoria for several months up to more than a year in many cases. Proptosis, bleeding, ocular pain, an obvious fungating mass, and other sequelae are the reasons most parents get referred for further management after months of treatment with eye drops at the local clinic or district hospital. These points to a lack of information among many healthcare providers about the clinical presentation of retinoblastoma, resulting in delayed diagnosis and poor treatment outcomes.

Staging for retinoblastoma is usually according to the International Retinoblastoma Staging System and is staged from 0 to IV. Metastases occur in the central nervous system or systemically (commonly in the bones, bone marrow, and liver). The majority of patients presenting at UTH have stage IV disease at presentation, with up to 30% having central nervous or skeletal system involvement, unfortunately sometimes the stage of disease is not documented.

Treatment

In Zambia, the standard treatment for patients with retinoblastoma involves surgery (enucleation or exenteration) with neoadjuvant chemotherapy [Appendix I] in patients with extensive proptosis, laser treatment, chemotherapy [Appendix I], and/or radiation therapy with electrons (1.8–2 Gray per fraction to a maximum of 36–46 Gray) to the orbit. The treatments are lengthy, and for chemotherapy, the associated side effects sometimes result in treatment delays. The most common adverse effects include neutropenia, nausea and vomiting, anemia, diarrhea, and oral mucositis. Sometimes, children present with underlying malnutrition and anemia which further complicates their management. Many of these children require additional supplemental feeds with the formulations for malnutrition, in addition to blood transfusions in cases of severe anemia. In cases of severe neutropenia GCSF is given and, the child is placed in isolation until their neutrophil count recovers.

Outcomes

Retinoblastoma is >95% curable in developed countries but has <30% survival in developing countries where disease stage at presentation is usually advanced or metastatic which adversely affects outcomes of treatment and survival.[5,6] Slone *et al*.,[1] found that <10% of children actually completed any treatment protocol and a good number of them left against medical advice or just absconded their treatment. Mortality is more than 30%, mainly as a result of advanced disease at presentation and comorbidities with common diseases of childhood such as malnutrition and diarrhea.

Challenges and Barriers

Looking at the high numbers of patients not completing treatment, one of the major challenges is getting parents or caregivers to adhere to agreed therapy modalities. A number of parents could not consent to surgical interventions and often absconded. Some returned later after months or even a year with obvious disease progression and poorer outcomes for their charges.

Months of chemotherapy also resulted in several absconding treatments without completing the prescribed number of cycles. Distance from their home towns and villages also results in parents or carers staying at the hospital for months with disruption of their normal routines, family life, and productivity. This could be a contributing factor to abandonment of treatment and merits further study.

Many patients did not return when scheduled for radiation therapy, perhaps due to lack of understanding of the value of this treatment for their children.

Another major challenge is periodic stock outs of chemotherapy drugs, resulting in the delayed administration of scheduled regimens. Sometimes, the radiotherapy machine also breaks down resulting in a break in the treatment sessions until the machine is running again.

Barriers include access to primary care and prompt referral to specialist services. Healthcare providers at the primary healthcare level need to be conversant with the clinical presentation of retinoblastoma to promptly refer clients, instead of treating them with antibiotic eye drops for months as is the case in many instances. Deliberate capacity building in this regard is needed for the primary healthcare service providers to educate the communities on the signs and symptoms of retinoblastoma.

Many patients did not have the disease diagnosed histologically because histopathology services are not decentralized and are inefficient. Despite surgical intervention, results for the samples sent for histology are delayed or in some cases never available to the clinicians.

Conclusion

Pediatric cancers are a growing problem in Zambia, and management remains daunting with many challenges. The treatments are lengthy, and for chemotherapy, the associated side effects sometimes result in treatment delays. Improvement in patient management through increased diagnostic capacity, more consistent supply of drugs, and specialization of staff in pediatric oncology are some of the ways, in which management of children with retinoblastoma

could become more efficient and effective. One of the most fundamental and cost-effective ways to reduce the burden of retinoblastoma morbidity and mortality would be upscaling awareness and screening at the primary healthcare level within the communities where these children and their families live. This is because retinoblastoma is a cancer that has good outcomes if detected early enough for curative therapies to be implemented.

Building capacity for screening among nurses and clinical officers found in primary healthcare facilities will go a long way in making early detection a reality and improving treatment outcomes for the children and families affected by this disease.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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APPENDIX I

1. Alternative Retinoblastoma Protocol (repeated at 4-week intervals, for 9 cycles)
   a. For patients >10 kg body weight
      - Cisplatin 100 mg/m^2 day 1
      - Etoposide 100 mg/m^2 days 1–3
      - Vincristine 1.4 mg/m^2 day 15
      - Cyclophosphamide 800 mg/m^2 day 15
   b. For patients ≤10 kg body weight
      - Cisplatin 15 mg/m^2/day days 1–5
      - Etoposide 3.3 mg/kg/day days 1–3
      - Vincristine 0.05 mg/kg/day day 15
      - Cyclophosphamide 25 mg/kg day 15
2. Retinoblastoma Protocol Stage IV/Relapse (repeated at 3–4 week intervals, for 6 cycles)
   a. VEC: (Patients >3 years old)
      - Vincristine 1.5 mg/m^2 i.v (bolus) day 1
      - Etoposide 150 mg/m^2 i.v (1 h infusion) days 1, 2
      - Carboplatin 560 mg/m^2 i.v (1 h infusion) day 1
   b. VEC: (Patients <3 years old) drug dose route frequency
      - Vincristine 0.05 mg/kg/day i.v (bolus) day 1
      - Etoposide 5 mg/kg/day i.v (1 h infusion) days 1, 2
      - Carboplatin 18.6 mg/kg/day i.v (1 h infusion) day 1