



## Case Report

# Retinoblastoma: A case study

K.Edvina Princy<sup>1,\*</sup>

<sup>1</sup>Dept. of Medical Surgical Nursing, Army Institute of Nursing, Guwahati, Assam, India



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### ABSTRACT

Retinoblastoma is also a rare malignancy of the eye affecting children, most typically four years recent and younger. Though therapy and radiation treatment aim to spare the eye, in some cases, Enucleation (i.e. removal of the eye) is needed to forestall cancer metastases or recurrence. Surgical procedures primarily performed at specialty establishments and may involve the surgical placement of an implant at intervals the orbit of the eye. It is vital part of the nurse to offer psychological support to the kid and members of the family.

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## 1. Introduction

Retinoblastoma is also a rare form of cancer that develops from the immature cells of a membrane, the light-detecting tissue of the eye. It is the foremost common primary malignant intraocular cancer in children, and it is nearly solely found in young children. Although most children survive this cancer, they will lose their vision at intervals the affected eye(s) or need to have the eye removed. Nearly 0.5% children with metastatic tumour have a hereditary genetic disorder associated with metastatic tumour. In alternative cases, it is caused by a innate mutation within the chromosome 13 13q14 (retinoblastoma protein).<sup>1</sup>

### 1.1. Etiology

Genes' mutation that is found in chromosomes, will have an effect on the method throughout once cells grow and develop at intervals in the body.<sup>2</sup> Once there is alterations in RB1 or MYCN, it will turn out metastatic tumour.

#### 1.1.1. RB1

In children with the genetic form of metastatic tumour, a mutation happens in tumour suppressor gene on chromosome 13. RB1 was the primary growth suppresser cloned. Although RB1 interacts with over one hundred cell proteins,<sup>2</sup> it is negative regulator impact on the cell cycle chiefly arises from binding and inactivation of the transcription issue E2F, therefore repressive the transcription of genes that square measure needed for the S part.<sup>2</sup> The defective RB1 chromosomes are often heritable from either parent; in some children, however, the mutation happens within the early stages of craniates development. The expression of the RB1 is chromosome dominant with ninetieth penetrance. Many ways are developed to discover the RB1 chromosomeS mutations.<sup>3</sup> makes an attempt to correlate cistron mutations to the stage at presentation haven't shown convincing proof of a correlation.

#### 1.1.2. MYCN

Somatic amplification of the MYCN transforming gene is answerable for some cases of non-heritable, early-onset, aggressive, unilateral metastatic tumour. Though MYCN amplification accounted for less than one, 4% of metastatic

\* Corresponding author.

E-mail address: [ephzina@gmail.com](mailto:ephzina@gmail.com) (K. E. Princy).

tumour cases, researchers know it in eighteen of infants diagnosed at however 6 months aged. Median age at diagnosing for MYCN metastatic tumour was 4-5 months, compared with twenty-four months for folks that had non-familial, unilateral malady with 2 RB1 cistron mutations.<sup>4</sup>

### 1.2. Symptoms

The primary clue and most blatant symptom is that the eye doesn't look right. Specifically, black pupil might look white. In a photo, instead of "red eye," a shaver with metastatic tumour can have one pupil that glows white once light shines there on. Other symptoms include:

1. Fixed eyes or eyes that seem larger than traditional
2. Cloudiness or discoloration within the centre of the attention
3. Eye pain
4. Eyes that cross or look in many directions
5. Redness within the white of the attention.<sup>4</sup>

### 1.3. Staging

Metastatic tumour staging is often variety of systems with numerous end-points and multiple systems square measure often used at the same time.

#### 1.3.1. Reese Ellsworth Classification

The Reese Ellsworth classification is assessed with fundoscopy and aims at predicting the prospect of protective the eye with external beam irradiation. The International Classification of metastatic tumour, that predict overall survival.<sup>1-4</sup>

Stage 0: Patients treated guardedly

Stage I: Eye enucleated, fully resected histologically

Stage II: Eye enucleated, microscopic residual growth

Stage III: Regional extension

A: Unconcealed orbital malady

B: Pre-auricular or cervical lymphoid tissue extension

Stage IV: Pathologic process malady

A: Haematogenous metastasis while not central nervous system involvement, single lesion & multiple lesions

B: Central nervous system pathologic process involvement, Prechiasmatic lesion, CNS mass, leptomenigeal disease.<sup>5</sup>

### 1.4. Diagnosis

To diagnose this cancer, doctor check the eye, for appearance closely at with a sturdy light and a magnifying lens. If it is like there's cancer, successive step is to search out however huge the growth is and whether or not it is unfolded. Child might have one in all these tests:

A MRI (magnetic resonance imaging) - powerful magnets and radio waves build careful pictures of the attention A CT scan (computed tomography)

- many X-rays taken from completely different angles measure place along to indicate additional info. The results facilitate doctors select the simplest course of action.<sup>6</sup>

### 1.5. Treatment

The priority of metastatic tumour treatment is to preserve the lifetime of the child, to preserve vision, then to attenuate complications or facet effects of treatment. The precise course of treatment depends on the individual case and is ready by the medical specialist in discussion with the paediatric specialist.<sup>4</sup> The varied treatment modalities for metastatic tumour includes:<sup>5</sup>

Enucleation: Most patients with unilateral malady gift with advanced intraocular malady thus typically bear surgery, that lands up throughout a cure rate of ninety fifth. In bilateral rubidium, surgery is usually reserved for eyes that have failing all proverbial effective therapies or while not helpful vision.

External beam irradiation (EBR): when a young child with bilateral metastatic tumour who has active or continual malady when completion of therapy is indicated and native therapies. However, patients with genetic defect who received EBR medical aid square measure rumoured to own a thirty fifth risk of second cancers.<sup>6</sup>

Brachytherapy: involves the placement of a radioactive implant (plaque), typically on the sclerotic coat adjacent to all-time low of a growth. It used as a result of the first treatment, or additional of in patients with little tumours or in people who had failing initial medical aid as well as previous EBR medical aid.

Thermotherapy involves the appliance of warmth on to the growth, typically within the sort of infra-red radiation. It is additionally used for little tumours. Optical maser surgical operation is typically suggested just for little posterior tumours.

Cryotherapy induces damage to the vascular endothelium with secondary thrombosis and infarction of the tumour tissue by rapidly freezing it. It may be used as primary therapy for little peripheral tumours or for little recurrent tumours previously treated with other methods.

Systemic chemotherapy is utilized in patients with unilateral disease when the tumours are small but can't be controlled with local therapies alone.

Intra-arterial chemotherapy: Chemotherapeutic drugs are administered locally by a thin catheter threaded through the groin, through the aorta, and thus the neck, directly into the optic vessels.<sup>7</sup>

Nanoparticulate chemotherapy is to reduce the adverse effects of systemic therapy; sub-conjunctival (local) injection of nano-particle carriers containing chemotherapeutic agents (carboplatin) has been developed.

Chemo reduction is a combined approach using chemotherapy to initially reduce the size of the tumour, and adjuvant focal treatments, such as trans-pupillary thermotherapy, to control the tumour.<sup>8</sup>

### 1.6. Complications

1. Recurring cancer
2. Risk of developing other types of cancer in any other body part.<sup>8</sup>

### 1.7. Prognosis

In the developed world, retinoblastoma has one among the simplest cure rates of all childhood cancers (95-98%), with quite 90% of sufferers surviving into adulthood. In the UK, around 40 to 50 new cases are diagnosed annually. In India, around 1500 new cases were reported annually. Good prognosis depends upon early presentation of the child in health facility. Late presentation is associated with a poor prognosis.<sup>9</sup> Survivors of hereditary retinoblastoma have a higher risk of developing other cancers later in life.

## 2. Nursing management: A case study

A case study of a child with Retinoblastoma is discussed with consent from her mother. Miss. X, 3-years child was admitted in eye ward at GMCH on 24.01.2020 with the complaints of diminished vision in her right eye for 7 months, watery discharge from the eye for 2 months, swelling (Protrusion) for 2 months and pain and severe itching for 2 weeks. She had a history of right eye redness which did not respond to topical medications. She was quite well till 2 years old. She has no past medical history except some cold and fever. In family history, her elder sister also developed same complaints at 3 years and she was found as 4th grade Retinoblastoma. On arrival, vital signs are stable. FNAC was done and report shows that negative for malignancy and chest X-ray was normal. MRI revealed Transaxial T2-weighted (TR/TE, 3,460/116 ms) (a) and T1-weighted (TR/TE, 374/14 ms) precontrast (b) and postcontrast (c) MRI of exophytically growing retinoblastoma with secondary retinal detachment. Retinoblastoma typically has low signal intensity compared to the vitreous body on T2-weighted images and intermediate signal intensity on pre-contrast T1-weighted images, and it demonstrates marked contrast enhancement. Uni-lateral retinoblastoma with focal bulging of the posterior eye segment (arrow) of the right eye and a shallow anterior chamber seen on T2-weighted (TR/TE, 4,430/102 ms) image. Patient underwent Enucleation. Child's vital signs are stable from POD1.

### 2.1. Nursing Care

Nursing care is discussed elaborately using nursing process approach.

#### 2.1.1. Nursing diagnosis

Acute pain related to itching from the right eye as evidenced by irritability and crying.

2.1.1.1. Expected outcome . The child will be able to express decreased pain, sleep and demonstrate decreased irritability..

#### 2.1.1.2. Nursing Interventions .

1. Assessed the severity and duration of a pain
2. Observed precipitating factors, recurrence, and progressive characteristics. The child's pain level measured using Wong Bakers faces scale as 6.
3. Provided basic comfort measures (e.g.: repositioning) and leisure activities (e.g. playing ball)
4. Provided toys and play things. Talked to families use distraction therapy, as well as other methods of pain relief.
5. Administered analgesics (Syp. Paracetamol 5mg tid) as per doctor's prescription.

### 2.2. Evaluation

The child expressed level of pain as 2 in Wong Bakers pain scale and she was comfortable and played with other child in the unit.

### 2.3. Nursing diagnosis

Disturbed sensory perception (visual) related to surgical removal of the eye

### 2.4. Expected outcome

The child will maintain visual acuity without further loss.

### 2.5. Nursing Interventions

1. Oriented the child to the environment, staff, everyone else in the area.
2. Removed extra furnitures around the child's bedside.
3. Arranged the things needed like toys, plate, spoon within reach.
4. Encouraged the child to express feelings of loss / possible loss of vision.
5. Assisted the child to use her limited vision to do her daily activities.
6. Assured the child by using one eye also she can manage the things.

## 2.6. Evaluation

The child managed with another eye.

### 2.6.1. Nursing diagnosis

Anxiety (Child & Mother) related to the disease suffered by the child.

2.6.1.1. Expected outcome . The mother & child will express decreased anxiety.

### 2.6.1.2. Nursing Interventions .

1. Assessed the level of anxiety, the degree of experience of pain / symptoms develop suddenly and the current state of knowledge.
2. Provided information that is accurate and truthful. Informed the family about supervision and treatment may prevent vision loss enhancement.
3. Encouraged the family to acknowledge the problem and express feelings.
4. Identified of sources / people who helped them like MSW.
5. Provided diversional therapy like toys, dolls.

## 2.7. Evaluation

The mother and child were happy and cope up with the condition.

### 2.7.1. Nursing diagnosis

Knowledge deficit (Mother) related to treatment and prognosis

2.7.1.1. Expected outcome . The mother will be able to understand the treatment procedure.

### 2.7.1.2. Nursing Interventions .

1. Encouraged the mother to ask her doubts.
2. Allowed her to express her feelings.
3. Discussed with the mother about the treatment and prognosis.
4. Encouraged mother to support the child physically and psychologically.

## 2.8. Evaluation

Mother understood the treatment process and cooperated well.

### 2.8.1. Nursing diagnosis

Risk for infection related to surgical procedure and hospitalization.

2.8.1.1. Expected outcome . The child remains free of infection

### 2.8.1.2. Nursing Interventions .

1. Assessed the risk and signs of infection like pain, discharge and fever.
2. Followed aseptic techniques during the procedure like administering injection etc.
3. Maintained in isolation when needed.
4. Taught about importance of hand washing to the child and mother.
5. Monitored temperature and reported elevation to physician.
6. Administer intravenous antibiotics like Inj.Amikacin 250mg twice a day as ordered.
7. Advised family members to avoid many visitors

## 2.9. Evaluation

She didn't develop any signs of infection like fever and discharge.

## 3. Conclusion

Retinoblastoma may be a tumour of the retina that's composed of undifferentiated neuroblastic cells and may be a malignant neoplasm of the retina in children. 40% of patients with retinoblastoma is hereditary disease. Retinoblastoma is a tumour that is autosomal dominant and an embryonic tumour. Most patients with active retinoblastoma discovered at the age of three years. . Information was given to the parents about the importance of regular follow up to minimize complications.

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## 6. Conflicts of Interest

None.

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### Author biography

**K.Edvina Princy** Tutor

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