ORIGINAL RESEARCH PAPER
Volume-8 | Issue-12 | December - 2019 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

NEUROBLASTOMA : A FIGHT TO LIVE

Nursing

Jatinder Kaur RN, RM, MSN, Clinical Instructor/tutor, Paediatric Nursing.

ABSTRACT

Care of the newborn diagnosed with a malignancy is a challenge for the neonatal intensive-care unit nurses. Malignancies found in infants differ from those found in 'older children. Nursing care of the neonate suspected or diagnosed with a malignancy includes problem identification and standard practices as well as interventions unique to the patient with cancer. This article reviews the incidence, diagnosis, treatment, and nursing management of neuroblastoma diagnosed in neonates.

The most common neonatal malignancy is neuroblastoma, accounting for 40-50% of malignancies diagnosed in neonates.

KEYWORDS

INTRODUCTION

In the discipline of pediatric cancer biology, neuroblastoma signifies an oncologic conundrum given the clinical range with which it presents. Neuroblastoma is a type of childhood cancer that starts in immature nerve cells in the sympathetic nervous system. These cells are called neuroblasts. Sometimes neuroblasts change and no longer grow or behave normally. Sometimes these changes may lead to non-cancerous, or benign, tumours such as ganglioneuroma. In some cases, these changes in neuroblasts can cause neuroblastoma. It can develop anywhere in the sympathetic nervous system, but it starts most often in the abdomen in the adrenal gland that lies just above the kidney,near the spinal cord in the neck, chest, periorbital areas. Neuroblastoma most commonly develops in babies and young children. The best prognosis is awarded to newborn followed by infant and toddler. The children over age five are subjected to poor prognosis.

Epidemiology

Neuroblastoma is the most common extra-cranial solid tumor in infants and children, representing 8%-10% of all childhood tumors. It accounts for approximately 15% of all cancer-related deaths in the pediatric population.1 The incidence of

neuroblastoma is 10.2 cases per million children under 15 years of age, and nearly 500 new cases are reported annually. While 90% of cases are diagnosed before the age of 5, 30% of those are within the first year. The median age of diagnosis is 22 months. Rarely does it present in adolescence and adulthood, but outcomes are much poorer in this age group. There is a slight predilection for males (1.2:1).1

Risk Factors

Family history: About 1% to 2% of children diagnosed with neuroblastoma have a family history of the disease. The risk for neuroblastoma seems to be highest for siblings or an identical twin of children who already have the disease.

Certain genetic conditions: Hirschsprung's disease is a condition in which nerves are missing from part of the intestines. As a result, the large intestine doesn't work properly and can get blocked.

Congenital central hypoventilation syndrome (CCHS) is also called primary alveolar hypoventilation. It is a rare disorder that affects breathing. Patient takes shallow breaths, especially when they are sleeping.

Neurofibromatosis type-1: affects the development and growth of nerve cells and causes tumors' to develop in the nerves and skin including neuroblastoma.

Beckwith-Wiedemann syndrome :affects how different parts of the body grow. Patients grow and gain weight at an unusual rate during childhood and have a higher risk of developing certain cancers, including neuroblastoma.

Li-Fraumeni syndrome : includes mutation in tumor suppressor gene that greatly increases risk of developing several types of cancer, including neuroblastoma.

Costello syndrome: causes delayed development, intellectual disability, a characteristic facial appearance, loose folds of extra skin and small growths and unusually flexible joints These children have a higher risk of developing neuroblastoma.

Clinical Presentation

The symptoms of neuroblastoma vary depending on where the cancer is and whether it's spread.The early symptoms can be vague and hard to spot, and can easily be mistaken for those of more common childhood conditions. Symptoms can include:

- a swollen, painful tummy, sometimes with constipation .
- breathlessness and difficulty swallowing
- a lump in the neck
- blueish lumps in the skin and bruising, particularly around the eyes
- weakness in the legs and an unsteady walk, with numbness in the lower body.
- fatigue, loss of energy, pale skin, loss of appetite and weight loss
- bone pain, a limp and general irritability
- rarely, jerky eye and muscle movements

Diagnostic Evaluation:

- A Urine Test – To check for certain chemicals produced by neuroblastoma cells .
- Scans –Ultrasound Scans, CT Scans And MRI Scans of various parts of the body to look at these areas in detail
- MIBG Scan – This involves the injection of a substance taken up by neuroblastoma cells
- Biopsy – the removal of a sample of cells from the tumour tissue for examination under a microscope
- Bone Marrow Biopsies – To see cancer cells in the bone marrow
- Tumour marker tests:
- Tumour markers are usually proteins shed by the tumour cells.
- Neuron-specific enolase (NSE). is an enzyme made by neural cells and neuroblastoma tumours.
- GD2 is a cell membrane ganglioside found on the surface of a neuroblastoma cell to make tumours grow faster.
- Chromagranin is a protein made by neuroblastoma cells.
- Lactate dehydrogenase (LDH) levels reflect the overall amount of tumor present in the child and whether it is actively growing.

Staging Of Neuroblastoma

- Stage L1 – the cancer is just in one place and has not spread, and can be removed by surgery
- Stage L2 – the cancer is in one place and has not spread, but cannot be removed safely by surgery

- Stage M – the cancer has spread to other parts of the body
- Stage Ms – the cancer has spread to the skin, liver or bone marrow in children aged less than 18 months.

Treatment For Intermediate-risk Neuroblastoma.:
Chemotherapy:
It may be given after surgery to treat any tumour that was left behind. Before surgery to shrink the tumour and make it easier to remove. A combination of chemotherapy drugs is often given for 4–8 cycles (12–24 weeks). The most common drugs used are:

- carboplatin (Paraplatin) or cisplatin (Platinol AQ)
- cyclophosphamide (Cytoxan, Procytox)
- doxorubicin (Adriamycin)
- etoposide (Vepesid, VP-1

Surgery:
is often used to remove as much of the neuroblastoma as possible. It may be used before or after chemotherapy. If surgery is done before chemotherapy, a second-look surgery may be done again after chemotherapy to remove if there is any cancer left if possible. For infants who have no symptoms, surgery may be the only treatment used for intermediate-risk neuroblastoma. It is followed by watchful waiting, or observation.

Radiation Therapy:
It may be used to treat a tumor that is causing serious symptoms like spinal cord compression or difficulty breathing (if the liver is enlarged and crowding the lungs).

Treatment For High-risk Neuroblastoma:
is often aggressive. It usually includes a combination of therapies, including chemotherapy, surgery, radiation therapy, stem cell transplant, retinoids and immunotherapy. Treatment is often divided into the following 3 phases.

Induction is done to try to get the cancer from remission by destroying or removing as much of the cancer as possible.

Consolidation is done to try to get rid of any cancer cells that are still in the body.. High-dose chemotherapy is given and then followed with stem cell transplant. Radiation therapy may be given to the primary tumor before, during and after high-dose chemotherapy and stem cell transplant.

Maintenance is done to try to lower the chance that the neuroblastoma will come back, or recur often involves retinoids and immunotherapy.

Chemotherapy is often used to treat high-risk neuroblastoma by shrinking the tumour. It also shrinks the blood vessels that supply the tumour. This process make the tumor easier to remove.The drugs include:
Carboplatin (Paraplatin, Paraplatin AQ)
Cyclophosphamide (Cytoxan, Procytox)
Vincristine (Oncovin)
Doxorubicin (Adriamycin)
Etoposide (Vepesid, VP-16)
Cisplatin (Platinol AQ)
Topotecan (Hycamtin)

Surgery:
is often used to get tissue samples to diagnose the tumor. Surgery is done again after chemotherapy to remove as much of the tumor as possible.

Stem Cell Transplant:
The child is then given high-dose chemotherapy to destroy any cancer cells in the body. Sometimes radiation may be given as well. After high-dose chemotherapy with or without radiation therapy, a stem cell transplant is done to replace the stem cells that were damaged or destroyed by high-dose chemotherapy. The child is given the stem cells that were collected before chemotherapy (called autologous peripheral blood stem cell transplant).

Radiation Therapy:
It may be given in preparation for stem cell transplant, after stem cell transplant or to treat emergencies, such as spinal cord compression, or to treat areas where the cancer has spread that did not respond well to other therapies. After surgery, radiation therapy is sometimes given to the tumor bed to destroy any cancer cells that may be left behind after surgery. Radiation therapy to the tumor. It may be given before, during and after high-dose chemotherapy and stem cell transplant.

Sometimes targeted radiation with radioactive metaiodobenzylguanidine (MIBG) is used to treat high-risk neuroblastoma with high-dose chemotherapy and stem cell transplant.

Retinoids and immunotherapy: may be given after stem cell transplant to treat high-risk neuroblastoma. A combination of drugs is usually given, including 13-cis-retinoic acid, interleukin-2, sargramostim and dinutuximab.

Treatments For Stage 4s Neuroblastoma
Watchful waiting: in children with stage 4S neuroblastoma who don't have any symptoms. Often these tumors will disappear on their own or will turn into a non-cancerous ganglioneuroma. If symptoms develop, then chemotherapy :

Chemotherapy: drugs include:
Etoposide (Vepesid, VP-16)
Cyclophosphamide (Cytoxan, Procytox)
Carboplatin (Paraplatin, Paraplatin AQ)
Doxorubicin (Adriamycin)
Vincristine (Oncovin)

Follow-up After Treatment For Neuroblastoma
Oncology Group provides long-term follow-up guidelines for childhood cancer survivors.Advise not to wait until the next scheduled appointment to report any new symptoms and symptoms that don't go away such as:
Any lump, growth or swelling
Difficulty breathing or swallowing
Nausea and vomiting
Loss of appetite
Weight loss
Pain
Limping or weakness in legs

The chance of neuroblastoma coming back, or recurring, is greatest within 2 years, so close follow-up is needed during this time. It is very rare for relapses to occur more than 5 years after treatment.

Schedule For Follow-up Visits
Every 2–6 months for the first few years after treatment
Less often over the next several years

Prognostic Factors:
A predictive factor influences how a cancer will respond to a certain treatment..The following are prognostic and predictive factors for neuroblastoma.

Age : Children younger than 12 months of age usually have a better outcome.

Stage: The earlier the stage at diagnosis, the better the prognosis.

Tumor histology: is what the cancer cells look like under the microscope and done to determine if a tumor has a favourable or unfavourable histology**.**

DNA ploidy : Diploid tumors tend to be less responsive to treatment and often need more aggressive therapy. than Hyperdiploid tumors that usually have a better prognosis.

MYCN status: is a gene that regulates cell growth. Many copies of the gene are usually seen in more aggressive tumors.

Chromosome changes: Children with a greater number of chromosome changes often have a poorer prognosis or higher risk of recurrence.

Side Effects:
Bone Marrow Suppression, Weakened Immune System, Low Platelet Count, Low White Blood Cell Count,Anemia,Nausea And Vomiting, Sore Mouth Or Throat, Fatigue, Loss Of Appetite, Hair Loss, Diarrhea,

Constipation, radiation enteritis ,infection, graft-versus-host disease.

CASE STUDY

The case study of a male child aged 7 years with complain of left eye swelling x 1 week with stage IV neuroblastoma who received palliative care at a tertiary care Hospital, Ludhiana in 2017. Physical Exam: reveled palpable soft tissue mass in lateral aspect of left orbit with fullness of upper lid. Significant (6-8mm) proptosis /exophthalmos of left globe. Some restriction in upward gaze in left eye. Full motility in right eye. Fixes and follows with each eye. Developmental issues relevant to child. He completed intensive treatment for neuroblastoma (i.e., chemotherapy, and irradiation) before the disease progresssed. Pain management was difficult in this child but achieved with parenterally-administered morphine (via a central line. Child received palliative radiotherapy to try and alleviate pain symptoms. Child came from a rural location which provided extra challenges in his care. In spite of the appropriate treatment with intensive regimen, the patient failed to improve and died of progressive disease.

CONCLUSIONS

Nursing care of the child with a solid tumor is complex and demanding. Children with solid tumors receive multimodal therapy including surgery, chemotherapy, and radiation, and some will receive high-dose chemotherapy with stem cell reinfusion. Side effects of treatment are common, most often caused by toxicity related to chemotherapy and radiation. Nurses who care for these children need to be aware of potential side effects, and what actions to take if they occur.

Nursing Diagnosis:

- Potential for infection related to marrow suppression
- Respiratory compromise related to poor perfusion.
- Immobility related to tumor mass
- Potential for injury related to bleeding tendencies
- Alteration in skin integrity related to venous access device (VAD)
- Potential for alteration in nutrition related to mucosal injury, nausea,vomiting
- Nausea and vomiting related to chemotherapy
- alteration in comfort related to tumor mass, chemotherapy.

REFERENCES:

1. Park JR, Eggert A, Caron H. Neuroblastoma: biology, prognosis, and treatment. Hematol Oncol Clin North Am. 2010;24:65–86. [PubMed] [Google Scholar]
2. Maris JM. Recent advances in neuroblastoma. N Engl J Med. 2010;362:2202–2211. [PMC free article] [PubMed] [Google Scholar]
3. Esiashvili N, Anderson C, Katzenstein HM. Neuroblastoma. Curr Probl Cancer. 2009;33:333–360. [PubMed] [Google Scholar]
4. Brodeur GM. Neuroblastoma: biological insights into a clinical enigma. Nat Rev Cancer. 2003;3:203–216. [PubMed] [Google Scholar]
5. London WB, Matthay KK, Ambros PF, et al. Clinical and biological features predictive of survival after relapse of neuroblastoma: A study from the International Neuroblastoma (NB) Risk Group (INRG) Database. J Clin Oncol. 2010;28:9518. [Google Scholar]
6. American Society of Clinical Oncology. (2014, 04). Neuroblastoma.
7. Bartholomew, J. Neuroblastoma. Baggott C, Fochtman D, Foley GV & Patterson Kelly, K (eds.). (2011). Nursing Care of Children and Adolescents with Cancer and Blood Disorders. (4th Edition). APHON. 30: pp. 1038-1053.
8. Brodeur GM, Hogarty MC, et al. Neuroblastoma. Pizzo, P. A. & Poplack, D. G. (Eds.). (2011). Principles and Practice of Pediatric Oncology. (6th Edition). Philadelphia: Lippincott Williams & Wilkins. 30:886-922.
9. Duffey-Lind, E. Neuroblastoma. Kline, N. E. (Ed.). (2004). Essentials of Pediatric Oncology Nursing: A Core Curriculum. (2nd Edition). Association of Pediatric Oncology Nurses. 2:6: pp. 35-30.
10. Neuroblastoma. Janes-Hodder, H. & Keene, N. (2002). Childhood Cancer - A Parent's Guide to Solid Tumor Cancers. (2nd Edition). O'Reilly. pp. 124-136.
11. Lacayo NJ. (2012, 6/6). Medscape: Pediatric Neuroblastoma Treatment and Management.
12. Hendershot, C. Neuroblastoma. Tomlinson, D. & Kline, N. E. (2005). Pediatric Oncology Nursing: Advanced Clinical Handbook. Germany: Springer. 2.6: pp. 50-57.