Wilms Tumour

Definition:

Wilms tumour, also known as nephroblastoma, is a type of kidney cancer primarily affecting children. It is a rare cancer, accounting for about 5% of all childhood cancers.

Pathophysiology:

The pathophysiology of Wilms tumour involves the abnormal proliferation of immature kidney cells, which results in the formation of a solid mass within the kidney. The tumour typically arises from the nephrogenic blastema, a group of embryonic kidney cells that give rise to the different cell types within the kidney.

In Wilms tumour, the normal process of differentiation, where embryonic cells mature into specific cell types, is disrupted. As a result, the tumour contains a mixture of different cell types, including undifferentiated blastemal cells, immature epithelial cells, and stromal cells. These abnormal cells grow and divide uncontrollably, leading to the formation of a tumour mass.

The pathophysiology of Wilms tumour also involves the loss of function of specific tumour suppressor genes, including WT1 and WT2, which normally regulate cell growth and division. Mutations in these genes can result in the abnormal proliferation of kidney cells, leading to the development of Wilms tumour.

Types of Wilms Tumour:

Favourable histology: This is the most common type of Wilms tumour and accounts for about 90% of cases. It is characterised by a well-defined border and a mixture of blastemal, epithelial, and stromal cells. The cells appear relatively normal and do not show aggressive features under the microscope.

Anaplastic histology: This is a rare type of Wilms tumour that accounts for about 5–10% of cases. It is characterised by the presence of anaplastic or abnormal-looking cells, which may have large nuclei, an irregular shape, and other features that suggest aggressive behaviour. Anaplastic Wilms tumour is associated with a poorer prognosis compared to favourable histology Wilms tumour.

Diffuse anaplasia: This is a subtype of anaplastic Wilms tumour that is characterised by the presence of anaplastic cells throughout the tumour. It is associated with a poorer prognosis compared to focal anaplasia, where anaplastic cells are present only in certain areas of the tumour.

Clear cell sarcoma: This is a rare subtype of Wilms tumour that is characterised by the presence of clear cells, which have a distinct appearance under the microscope. Clear cell sarcoma is associated with a poorer prognosis compared to favourable histology of Wilms tumour.

Rhabdoid tumour: This is a rare and aggressive subtype of Wilms tumour that is characterised by the presence of rhabdoid cells, which have a distinct appearance under the microscope. Rhabdoid tumours are associated with a poorer prognosis compared to other types of Wilms tumours.

Causes of Wilms Tumour:

The cause of Wilms tumour is not well understood, but there are genetic mutations that have been associated with its development. These mutations can occur spontaneously or be inherited from a parent. Wilms tumour is more common in children with certain genetic syndromes, such as WAGR syndrome, Beckwith-Wiedemann syndrome, and Denys-Drash syndrome. Wilms tumour is slightly more common in girls than boys and is more common in African-American children than in other racial or ethnic groups.

Clinical Manifestations of Wilms Tumour:

The most common symptom of Wilms tumour is abdominal swelling or a mass that can be felt in the abdomen. Other symptoms may include abdominal pain, fever, nausea, vomiting, loss of appetite, and blood in the urine.

Diagnostic Criteria:

The diagnosis of Wilms tumour is usually made based on imaging studies such as ultrasound, CT scan, or MRI. A biopsy may be done to confirm the diagnosis. Specific laboratory tests are not usually used to diagnose Wilms tumour, but blood and urine tests may be done to evaluate kidney function and assess overall health.

Treatment:

Treatment for Wilms tumour usually involves surgery to remove the tumour, followed by chemotherapy and sometimes radiation therapy. The specific treatment plan will depend on the size and location of the tumour, the stage of the cancer, and other factors. Chemotherapy drugs commonly used to treat Wilm's tumour include doxorubicin, vincristine, and actinomycin-D. Radiation therapy may be used in some cases to help destroy any remaining cancer cells after surgery.

Overall, the prognosis for children with Wilms tumour is generally good, with a five-year survival rate of over 90% for children with favourable histology. However, the prognosis is less favourable for children with unfavourable histology or those with advanced-stage disease at the time of diagnosis.

Contraindications/Cautions:

There are no specific contraindications or cautions related to Wilm's tumour, but treatment may be adjusted based on the patient's age and overall health status.

Gender and Age Differences:

Wilms tumour is more common in children, with most cases diagnosed before the age of 5. It affects both genders equally.

Nursing Asssessment:

Nurses should assess the patient for any signs or symptoms of Wilms tumour, including abdominal swelling, pain or discomfort, blood in the urine, and high blood pressure. They should also monitor the patient's overall health status and response to treatment.

Nursing Diagnoses:

- Risk for impaired physical mobility
- Risk for infection
- Disturbed body image
- Anxiety

Nursing Management:

- Provide emotional support to the patient and their family, including education on the diagnosis and treatment options.
- Monitor the patient's vital signs, laboratory values, and response to treatment.
- Administer medications as prescribed, including chemotherapy and pain management.
- Provide education on self-care and management of side effects of treatment, such as nausea and vomiting.
- Prepare the patient for surgery, including preoperative teaching and postoperative care.
- Promote physical activity and mobility as appropriate while ensuring adequate rest and recovery time.