Wilms' Tumour

Synonym: nephroblastoma

Wilms’ tumours are the most common intra-abdominal tumours of childhood. A Wilms’ tumour is an undifferentiated mesodermal tumour of the intermediate cell mass (primitive renal tubules and mesenchymal cells). It may be sporadic or familial.

Epidemiology

- Wilms’ tumour is the most common renal tumour of childhood and affects about 1 in 10,000 children[1].
- The incidence is estimated at 7 in 1,000,000 children under 16 years of age[2].
- Wilms’ tumour is the second most common intra-abdominal cancer of childhood and the fifth most common paediatric malignancy overall (approximately 6% of all paediatric cancers)[3].
- Only 3% of Wilms’ tumours are reported in adults[4]. Most adult patients are diagnosed unexpectedly following nephrectomy for presumed renal cell carcinoma[5].
- In 5-10% of patients, both kidneys are affected at the same time (synchronous bilateral Wilms’ tumour) or one after the other (metachronous bilateral Wilms’ tumour)[6].

Associations[3]

Wilms’ tumours usually develop in otherwise healthy children but approximately 10% occur in children with recognised malformations; either:

- ‘Overgrowth syndromes’ (excessive prenatal and postnatal somatic growth resulting in macroglossia, nephromegaly and hemihypertrophy) - most commonly, Beckwith-Wiedemann syndrome or isolated hemihypertrophy. Others include Perlman syndrome, Sotos’ syndrome, and Simpson-Golabi-Behmel syndrome.
- No ‘overgrowth’ - associated with Edwards syndrome (trisomy 18), Bloom’s syndrome, Denys-Drash syndrome; or WAGR (Wilms’ with Aniridia, Gonadoblastoma (genitourinary malformations), and Retardation).

Familial Wilms' tumour[7]

Hereditary Wilms’ tumour (either bilateral tumours or a family history of the neoplasm) is uncommon. Several different families with Wilms' tumours have been identified. All are transmitted in an autosomal dominant manner, caused by mutations in one of at least three genes:

- One related to the WT1 gene on chromosome 11 (11p13) - (includes those patients with WAGR) - encodes a protein which is a transcriptional repressor downregulating IGF-II, an insulin-like growth factor.
- Other families (including those with Beckwith-Wiedemann syndrome) have a different mutation - of the WT2 gene on chromosome 11 (11p15.5).
- Other gene mutations, thought to be on chromosome 16 (WT3-16q) and/or chromosome 1p can also cause the tumour[8].

Presentation

Usually presents in the first five years of life but 3% of presentations are in adults[9].

95% are unilateral. Only 1-2% have a positive family history[10].

Clinical features[3]

- The most common presentation is an asymptomatic abdominal mass.
- Abdominal pain.
- Haematuria.
- Urinary tract infection.
- Hypertension, gross haematuria and fever may occur but are uncommon.
- Advanced disease may rarely present with respiratory symptoms due to lung metastases.

Screening

A recommendation for surveillance of children at high risk (familial or associated conditions) included[11]:

- Surveillance should only be offered after review by a clinical geneticist.
- Surveillance should be carried out by renal ultrasound every 3-4 months.
Surveillance should continue until 5 years of age in all conditions except Beckwith-Wiedemann syndrome, Simpson-Golabi-Behmel syndrome and some familial Wilms’ tumour pedigrees, when it should continue until 7 years.

Investigations[8]

- Useful laboratory tests include FBC, renal function and electrolytes and urinalysis.
- Genetic studies may reveal the chromosomal abnormalities consistent with the condition.
- Ultrasound and/or intravenous pyelogram (IVP) may show distortion of the renal pelvis; hydronephrosis. Dynamic imaging of the renal vein and inferior vena cava may be contributory.
- Renal angiography may help to show a more detailed view of the blood vessels.
- CT and MRI scanning may help to determine the nature of the tumour and may also reveal the degree of involvement of the lymph nodes, whether the other kidney is involved and invasion into blood vessels or the liver.
- Transcutaneous renal biopsy should be avoided, as this may make the condition worse.
- Chest CT to detect lung metastases.

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<th>Staging of Wilms' tumour[12]</th>
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Management[6]

- For most patients, nephrectomy followed by chemotherapy (regimes include vincristine, dactinomycin and doxorubicin, sometimes with additional cyclophosphamide) can be curative.
- Routine postoperative radiotherapy to the flank is beneficial in patients with a stage III tumour.
- Patients with massive, nonresectable unilateral tumours, bilateral tumours, or venacaval tumour thrombus above the hepatic veins should be considered for pre-operative chemotherapy because of the risk of initial surgical resection.

Prognosis[13]

- With treatment, over 90% of children diagnosed with Wilms’ tumour survive long-term[14].
- Long-term survival is above 75% for those with metastatic disease[1].
- There is an increased risk of second tumours in survivors of Wilms’ tumour. Second tumours include bone and soft tissue sarcomas, breast cancer, lymphoma, gastrointestinal tumours and melanoma. Acute leukaemias may also occur.

Further reading & references

6. Wilms Tumor and Other Childhood Kidney Tumors; US National Cancer Institute.
7. Wilms Tumor 1, WT1; Online Mendelian Inheritance in Man (OMIM)
8. Wilms Tumor 3, WT3; Online Mendelian Inheritance in Man (OMIM)
10. Krämer KH; Xeroderma Pigmentosum, Gene Reviews, Feb 2014

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