

## Paediatric Head & Neck Malignancy

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**What's most likely vs what's most deadly.  
Kids are not little adults.**

### A-Z of Paediatric Neck Masses

Arteriovenous Malformation  
Branchial Anomalies  
Craniopharyngioma &  
CNS tumours  
Dermoid  
Encephalocele  
Fibrous dysplasia  
Germ Cell Tumours/Teratoma  
Haemangioma  
Infectious mononucleosis  
Juvenile Nasopharyngeal Angiofibroma  
Kaposiform haemangioendothelioma  
Lymphoma, Leukaemia, Langerhans Cell  
Histiocytosis, Lymphatic Malformation  
Mycobacterium Avium Complex  
Neuroblastoma  
Odontogenic cysts & tumours  
Pilomatrixoma, preauricular cyst  
Quinsy  
Ranula & Rhabdomyosarcoma  
Salivary gland tumours  
Thyroid and Thyroglossal Duct Cyst  
Undescended lingual thyroid  
Vascular Anomalies  
Wilms & renal ca mets  
Xeroderma pigmentosum  
Y?  
Zoster

### 3x3 grid for paed neck lumps

Midline	Lateral	Non-specific
Congenital	Infective/ Inflammatory	Neoplastic
Mucosal	Cutaneous/ Soft Tissue	Glandular (Salivary, thyroid)

### Round blue cell tumours

Lymphoma (B-cell, T-cell, NK cell)  
Anaplastic large cell lymphoma  
Burkitt lymphoma  
Rhabdomyosarcoma  
Undifferentiated sarcoma

Clear cell sarcoma  
Small cell osteosarcoma  
Rhabdoid tumour  
Chondrosarcoma  
Ewing tumour  
Neuroblastoma

### Syndromes with oncologic association

T21 – leukaemia  
NF1 & 2 – leukaemia, phaeo, AN, meningioma  
Gorlin – BCC, medulloblastoma  
MEN 2A & 2B – MTC, phaeo  
Beckwith Wiedemann - Rhabdo, neuroblastoma

750 new dx / year in Aus

Leukaemia, CNS tumours, Lymphoma, Rhabdo &  
Others

### HL

- Teens, EBV.
- Classic HL (95%)
- Nodular Lymphocyte Predominant HL
- Reed-Sternberg cells
- Ann Arbor Staging I-IV
- Chemo +/- RTx

### NHL

- 7-11yo, 3:1 M:F
- Lymphoblastic lymphoma
- Peripheral B-cell lymphoma (including Burkitt lymphoma)
- Anaplastic large cell lymphoma
- Starry sky appearance
- St.Jude Staging System I-IV
- Chemo

### PTLD

- Transplant recipients
- Up to 45% after bone marrow transplant
- Up to 80% after solid organ transplant
- Kidney recipients lower risk. Heart-lung high risk.
- Looks like “infectious mononucleosis”
- Rx: reduce immunosuppression, add steroids and chemo

### Rhabdomyosarcoma

- 35% present with H&N mass.
- Half of head and neck RMS occur at parameningeal sites: middle ear/mastoid, nasal cavity, parapharyngeal space,

paranasal sinuses, or the pterygopalatine/infratemporal fossa.

- Subtypes: Embryonal, Alveolar, Undifferentiated
- Clinical Group Stage System by Intergroup Rhabdomyosarcoma Study Group
- Favourable vs unfavourable site, size (5cm), completeness of resection (Gp 1-4), histology, regional node, metastasis.
- Multimodal Therapy: Surgery then Chemo +/- Rtx
- Staging and re-staging after initial Rx
- Resection is the treatment of choice for nonparameningeal tumors of the head and neck.
- Combination chemotherapy is the mainstay of treatment and reduces risk of death because of distant metastasis.
- Surgery not recommended unless there is a strong likelihood that clear margins will be achieved. Debulking surgery does not improve outcomes and delays critical chemotherapy treatments in the recovery period.
- Prophylactic neck dissections are not recommended, but clinically evident cervical lymphadenopathy must be addressed.
- Vincristine, actinomycin, and cyclophosphamide are the mainstay chemotherapy

#### Neuroblastoma

- Infancy (80% <age 5)
- International Neuroblastoma staging system
- Low, intermediate and high risk group
- Surgery/Chemo/RTx

#### NPC

- 2 peaks (adolescence and 40-60)
- Geography: Southern China, South East Asia, Mediterranean
- Strong EBV association
- Painless neck mass, nasal obstruction, epistaxis, unilateral MEE
- TNM Staging, 80% of kids present with Stage IV disease
- WHO Type I – III, most common in kids is Type III Undifferentiated
- Radiation +/- Chemo

#### Thyroid

- 5% of paediatric H&N malignancy
- Paediatric thyroid nodule is more likely to be malignant
- 90% PTC: likely to metastasise early (70%), often multifocal, yet have good survival, central and lateral neck in some settings, RAI.
- FTC: unifocal, mets haematogenously, completion thyroid in select situation only, long term surveillance, Tg levels.
- MTC part of RET protooncogene, MEN 2A & 2B, total thyroid and central neck, RTx, chemo for advanced and mets, Calcitonin and CEA

#### LCH

- Formerly Hand-Schuller-Christian, Letterer-Siwe, Histiocytosis X.
- Skin rash, bone lesions (uni or multifocal involving H&N in up to 75% of patients), lymphadenopathy, etc
- Histiocyte Society for clinical trials

#### How do you investigate a neck lump?

- Green: less than 2cm, asymptomatic, benign on US. Discharge.
- Yellow: larger than 2cm, asymptomatic, benign on US. Observe.
- Red: greater than 2.5cm, persistent over 2 months, supraclavicular, history of malignancy, suspicious US, systemic symptoms. Incisional or excisional biopsy.

#### FNA vs Core Bx vs Incisional Bx vs Excisional Bx.