Paediatric Head and Neck Lesions

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and
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Aim

• Spectrum of head and neck lesions in paediatric population
• Example cases – presented by Ming
Why

• Overall rarity of FNA in little people
• Head and neck the most common site
• Broad spectrum different to adults
• Often performed by adult cytopathologists
Children are not small adults!

• Age and location very important – different DDx
• Can be associated with genetic syndromes
• Important to have a good clinical history, examination, imaging, microbiology, viral serology etc.
Children are not small adults....ctd

• Morphologic similarities between paediatric malignancies
  -most common tumours are mostly small round blue cell tumours
  -both benign and malignant lesions can have spindle cell/ fibroblastic morphology

• Architecture on histology is critical for diagnosis of fibroblastic/spindle cell lesions
Cases
Primary FNAC of H&N lesion in a child is performed for mainly palpable superficial lesions.

1. Clinically suspicious LN/neck ‘lump’
2. Thyroid
3. Salivary Gland
4. Other miscellaneous (skin, soft tissue etc.)

*mostly benign
2 recent review articles summarizing the utility of FNA in paediatric population


<table>
<thead>
<tr>
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<tbody>
<tr>
<td>Total no. of cases</td>
<td>85 cases</td>
<td>748 cases</td>
<td>692 cases [584 cervical lymph nodes]</td>
<td>100</td>
</tr>
<tr>
<td>Age group</td>
<td>0-18 years</td>
<td>0-12 years</td>
<td>0-14 years</td>
<td>0-15</td>
</tr>
<tr>
<td>Adequacy of material</td>
<td>94%</td>
<td>93.4%</td>
<td>93%</td>
<td></td>
</tr>
<tr>
<td>Age predominance</td>
<td>6-10 years</td>
<td></td>
<td>10-15 years</td>
<td></td>
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<tr>
<td>Sex predominance</td>
<td>Male [69.4%]</td>
<td></td>
<td>Male : female ratio 1.5:1</td>
<td>Male 55%</td>
</tr>
<tr>
<td>Most common site of lesion</td>
<td>Lymph node cervical 69.4%</td>
<td>Lymph node cervical 81%</td>
<td>Lymph node cervical 84.3%</td>
<td>Lymph node cervical 87%</td>
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<tr>
<td>Other sites of lesions</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Skin &amp; subcut. tissue</td>
<td></td>
<td></td>
<td></td>
<td>2.1%</td>
</tr>
<tr>
<td>Miscellaneous [cystic &amp; soft tissue]</td>
<td>16 Case [Also in Bone]</td>
<td>7.6%</td>
<td></td>
<td>03.2%</td>
</tr>
<tr>
<td>Thyroid</td>
<td>3.2%</td>
<td></td>
<td></td>
<td>04.3%</td>
</tr>
<tr>
<td>Salivary gland</td>
<td>2 cases</td>
<td></td>
<td></td>
<td>01%</td>
</tr>
<tr>
<td>Orbital &amp; eye lid</td>
<td>0.2%</td>
<td></td>
<td></td>
<td>02.1%</td>
</tr>
<tr>
<td>Nature of Lesion</td>
<td>83%</td>
<td>98.5%</td>
<td>98.46%</td>
<td>88.17%</td>
</tr>
<tr>
<td>Malignant</td>
<td>17%</td>
<td>1.5%</td>
<td>1.54%</td>
<td>11.83%</td>
</tr>
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**[Table/Fig-6]: Comparative Study of FNAC From Similar Previous Studies**

<table>
<thead>
<tr>
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</thead>
<tbody>
<tr>
<td>Skin &amp; subcut. tissue</td>
<td><em>lymphangioma</em>haemangioma*lipoblastoma</td>
<td><em>Epidermal inclusion cyst</em>dermoid cyst<em>infected sebaceous cyst</em>chronic inflammations</td>
<td>2.1% *Epidermal inclusion cyst</td>
</tr>
<tr>
<td>Miscellaneous [cystic &amp; soft tissue]</td>
<td>16 Case [Also In Bone]<em>langhans cell histiocytosis</em>benign myxoid lesion<em>Spindle cell proliferation</em>osteosarcoma *Benign cystic teretoma</td>
<td>7.6% <em>Muscle fibromatosis coli</em>vascular hammartoma<em>lymphangioma</em>fibroma *neurofibroma</td>
<td>03.2% * Muscle fibromatosis coli<em>Hemangiomana</em>lymphangioma</td>
</tr>
<tr>
<td>Thyroid</td>
<td>1case of Papillary Carcinoma of Thyroid metastasis</td>
<td>3.2% total *Euthyroid colloid goiter [12case]*Thyroglossal duct cyst[11case]*Thyroid cyst [1case]</td>
<td>04.3% Total *Hashimoto’s Thyroiditis [01case]*Lingual thyroid [01 case]*Thyroglossal duct cyst [01case]*Papillary carcinoma of thyroid [01case]</td>
</tr>
<tr>
<td>Salivary gland</td>
<td>2cases*Pleomorphic adenoma [1case]*Mucoepidermoid carcinoma[ 1case]</td>
<td>2.1%[ total 15 cases]*Chronic sialadenitis [4 cases]*Mucus retention cyst [6 cases]*Pleomorphicadenoma [3cases] *Acute abscess[1case]*Normal [1 case]</td>
<td>01% of total *Pleomorphic adenoma [1case]</td>
</tr>
<tr>
<td>Orbital and eyelid</td>
<td>--</td>
<td>0.2%* Tubercular abcess [2case]</td>
<td>02.1% *Embryonal RMS[1case]*Small blue round cell tumor[1case]</td>
</tr>
</tbody>
</table>

**Table/Fig-8**: Comparative of Previous Studies in Head & Neck Lesions of Children

LN – mostly benign

- Mostly reactive (50-75%)
- Viral infections e.g. EBV
- Suppurative (bacterial)
- Necrotising granulomas (Atypical TB, TB, Cat scratch)
- Non necrotising granulomas (Toxoplasma, foreign body, Hodgkin and Non Hodgkin lymphoma)
- Necrotising (Kikuchi, SLE)
- Dermatopathic lymphadenitis
- Rosai-Dorfman
Table 1  Cervical lymph node drainage and selected causes of localised lymphadenopathy (partly adapted from Ferrer²⁸)

<table>
<thead>
<tr>
<th>Anatomical location</th>
<th>Lymphatic drainage</th>
<th>Selected causes of localised lymphadenopathy</th>
</tr>
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<tbody>
<tr>
<td>Anterior cervical</td>
<td>Throat, posterior pharynx, tonsils, thyroid gland</td>
<td>Local infections in the ear, nose and throat, infectious mononucleosis, cytomegalovirus infection, toxoplasmosis</td>
</tr>
<tr>
<td>Posterior cervical</td>
<td>Scalp and neck, thorax, cervical and axillary nodes</td>
<td>Local infections in the scalp, tuberculosis, lymphoma, head and neck malignancy</td>
</tr>
<tr>
<td>Tonsillar</td>
<td>Tonsillar and posterior pharyngeal regions</td>
<td>Infections of the throat</td>
</tr>
<tr>
<td>Submandibular</td>
<td>Floor of the mouth, submandibular gland, tongue, lips, conjunctivae</td>
<td>Dental disease, infections in the ear, nose, throat and eyes</td>
</tr>
<tr>
<td>Submental</td>
<td>Lower lip, floor of mouth, tip of tongue, cheek</td>
<td>Dental disease, local infections</td>
</tr>
<tr>
<td>Supraclavicular</td>
<td>Mediastinum, lungs oesophagus, abdomen via thoracic duct</td>
<td>Lymphoma, thoracic or gastrointestinal cancer</td>
</tr>
</tbody>
</table>
When to worry about a LN

- Duration - Persistently enlarged (>6/52 despite antibiotic treatment)
- Size
- Site – Left supraclavicular, posterior triangle
- Painless and firm/hard
- Non mobile
- Systemic symptoms
Malignancies in LN

• Primary haematolymphoid – Hodgkin, Lymphoblastic lymphoma, Burkitt lymphoma, Leukemic involvement of LN, ALK positive ALCL, DLBCL, Langerhans cell histiocytosis

• Metastatic (rare)- PTC, SRBCT, germ cell tumours, NPC (left supra clavicular LN from below diaphragm)
Common benign causes of paediatric lymphadenopathy

Benign Lymphadenopathy in Children

Some uncommon causes of paediatric lymphadenopathy

Langerhans cell histiocytosis

Acute lymphoblastic lymphoma

Hodgkin Lymphoma

Arrow = RS cell

Burkitt Lymphoma
Primary FNAC of H&N lesion in a child is performed for mainly palpable superficial lesions.

1. Clinically suspicious LN/neck ‘lump’
2. Thyroid
3. Salivary Gland
4. Other miscellaneous (skin, soft tissue)

*mostly benign
Thyroid

• Benign – Follicular or hyperplastic nodule, Hashimotos
• Malignant – PTC (80-95%), follicular ca, medullary ca, lymphoma

Risk factors - h/o radiation, DICER1 gene mutation for PTC and MEN2A and 2B for medullary thyroid ca
Classic papillary thyroid carcinoma
Medullary thyroid carcinoma. Calcitonin and NE markers positive. RET gene mutation.
Salivary

• Rare
• Benign – Sialadenitis, Lymphoepithelial cyst, mucocele, abscess, Pleomorphic adenoma
• Malignant (3-8%) – Mucoepidermoid, acinic cell ca, metastatic

• Mimics
- Intra parotid LN can be present
- Skin or soft tissue masses overlying the salivary glands
- Squamous metaplasia maybe present in benign inflammatory lesions. Don’t overcall MEC.
Primary FNAC of H&N lesion in a child is performed for mainly palpable superficial lesions.

1. Clinically suspicious LN/neck ‘lump’
2. Thyroid
3. Salivary Gland
4. Other miscellaneous (skin, soft tissue)

*mostly benign
Other Miscellaneous

• Benign (cystic or solid)
  -congenital/developmental
  -other skin, soft tissue, vascular/lymphatic, neural, bone/cartilage

• Malignant (mainly solid)
  -Neuroblastoma
  -Rhabdomyosarcoma
  -Teratoma (can be cystic)
  -other SRBCTs
Congenital/developmental

- Dermoid cysts
- Branchial cleft cyst/sinuses
- Nasolacrimal duct cyst
- Pre auricular pits/sinuses and cysts
- Haemangiomas and lymphatic malformations (generally diagnosed clinically)
- Glial heterotopia/nasal glioma
- Encephalocele
- Remnants
- Salivary gland heterotopias
Dermoid cysts

- Congenital - along lines of skin closure
- Have adnexal structures
- Epidermoid cysts are acquired

- Mimics
  - other cystic lesions with squamous cells
  - SCC unheard of in children unlike in adults
FIGURE 17.23 Frontal aspect of the face. A. 7-week embryo. Maxillary prominences have fused with the medial nasal prominences. B. 10-week embryo. C. Photograph of a human embryo at a stage similar to that in [A].
Figure 2. Normal embryologic development of the nasofrontal region. (a) Schematic illustrates the temporary fonticulus nasofrontalis and prenasal space. (b) Schematic illustrates how the fonticulus frontalis closes, the foramen cecum is formed, and a projection of dural diverticulum contacts the tip of the nose. (c) Schematic illustrates how the dural diverticulum retracts into the cranium and the prenasal space is obliterated. (Reprinted, with permission, from reference 5.)
Figure 11. Faulty development of the nasofrontal region leading to formation of various midface masses. (a) Schematic illustrates how dermal sinus tracts form when there is no involution or only partial involution of the dural diverticulum that extends through the foramen cecum to the columella. Dermoid and epidermoid cysts may form anywhere along the course of the dermal sinus tract owing to desquamation of tissue lining the tract. (b) Schematic illustrates how nasal gliomas form when the dural diverticulum that extends through the foramen cecum does not retract and involute normally, leaving sequestered neurogenic tissue that may be connected to the intracranial contents by a fibrous stalk. (c) Schematic illustrates how frontonasal encephaloceles form when the foniculus nasofrontalis remains patent. (d) Schematic illustrates how nasoethmoidal encephaloceles form when the foramen cecum fails to close and the pre-nasal space remains patent. (Reprinted, with permission, from reference 5.)
FIGURE 17.10  A. Development of the pharyngeal clefts and pouches. The second arch grows over the third and fourth arches, burying the second, third, and fourth pharyngeal clefts. B. Remnants of the second, third, and fourth pharyngeal clefts form the cervical sinus, which is normally obliterated. Note the structures formed by the various pharyngeal pouches.

FIGURE 17.14  A. Lateral cervical cyst opening at the side of the neck by way of a fistula. B. Lateral cervical cysts and fistulas anterior to the sternocleidomastoid muscle. Note also the region of preauricular fistulas. C. A lateral cervical cyst opening into the pharynx at the level of the palatine tonsil.
FIGURE 17.15 Patient with a lateral cervical cyst. These cysts are always on the lateral side of the neck in front of the sternocleidomastoid muscle. They commonly lie under the angle of the mandible and do not enlarge until later in life.
Fig. 4.14 Thyroglossal duct cyst (a. Diff-Quik stain, low power; b. Papanicolaou stain, high power). Abundant thick mucoid material with scant cells (a). Clusters of basal and columnar cells with cilia (b, arrow).
Lin et al. Bilateral cervical ectopic thymic nodules with accessory thyroid tissue and an ectopic parathyroid in the neck region. Journal of Dental Sciences (2011) 6, 61e64

Figure 2 Left ectopic thymic nodule with accessory thyroid tissue (A: 40×, B: 200×, arrowhead: accessory thyroid tissue in an ectopic thymus, arrow: Hassall’s corpuscle in an ectopic thymus); right ectopic thymic nodule with an ectopic parathyroid gland (C: 40×, D: 400×, thin arrow: an ectopic parathyroid).
Branchial cleft cyst
Other Miscellaneous

• Benign (cystic or solid)
  - congenital/developmental
  - other skin, vascular, neural, soft tissue, bone/cartilage

• Malignant (mainly solid)
  - Neuroblastoma
  - Rhabdomyosarcoma
  - Teratoma (can be cystic)
  - other SRBCTs
Miscellaneous
Benign nodules/lumps

• Mucocele
• Cystic hygroma
• Cervical rib
• Aabscess
• Schwannoma
• Skin lesions such as epidermoid cyst, pilomatrixoma
• Lipoblastoma
Clinically may present as a LN but ...........

**Table 3.5** Mimics of lymphadenopathy in children and adolescents

- Thyroid lesions/neoplasms
- Salivary gland lesions/neoplasms
- Cystic head and neck lesions (e.g., branchial cleft cyst, thyroglossal duct cyst, other developmental cysts)
- Extra nodal inflammatory lesions (e.g., abscess)
- Fibrous hamartoma of infancy
- Pilomatrixoma
- Mesenchymal lesions/neoplasms (e.g., lymphangioma/hemangioma, fat necrosis, lipoma, fibromatosis, solitary myofibroma, rhabdomyosarcoma)
- Neural neoplasms (e.g., schwannoma, neurofibroma, ganglioneuroma)
- Germ cell tumours (e.g., cervical teratoma)
- Odontogenic or bone lesions (e.g., odontogenic cyst, fibrous dysplasia)
- Thymic tissue or lesion (e.g., undescended thymus)
- Benign soft tissue elements (e.g., skeletal muscle, adipose tissue)
Lesions that Clinically Mimic Lymphadenopathy in Children
Lipoblastoma

Primary FNAC of H&N lesion in a child is performed for

Mainly palpable superficial lesions.

1. Clinically suspicious LN/neck ‘lump’
2. Thyroid
3. Salivary Gland
4. Other miscellaneous (skin, soft tissue)

*mostly benign

Malignancies............
Cases
Malignant

• Primary
• Secondary
Commonest Primary Malignancies in Paediatric Head and Neck

- Rhabdomyosarcoma
- Ewing’s sarcoma/PNET group (Classic Ewings and Atypical Ewings family of tumours)
- Extra renal rhabdoid tumour
- LCH
- Neuroblastoma
- Nasopharyngeal carcinoma

- Secondary/metastatic – PTC, SRBCT, NPC
Fig. 4.16 Rhabdomyosarcoma (a. Diff-Quik stain, high power; b. Papanicolaou stain, high power.) Pleomorphic cells with varying amounts of dense blue cytoplasm. A “tadpole” cell is seen at the bottom of the cluster (a). Pleomorphic nuclei with uneven granular chromatin (b).
**Fig. 4.19** Nasopharyngeal carcinoma, metastatic to lymph node (a. Diff-Quik stain, high power; b. Papanicolaou stain, high power). Large cells with mildly pleomorphic oval nuclei in cohesive clusters in a background of mixed lymphocytes.
DDx for rhabdoid morphology in an infant/child

- Extra renal rhabdoid tumour/ATRT in CNS/Renal rhabdoid tumour
- Epithelioid sarcoma
- Rhabdomyosarcoma
- Anaplastic large cell lymphoma
- Nasopharyngeal carcinoma

- Adults (Carcinomas, melanoma, extra skeletal myxoid chondrosarcoma, epithelioid MPNST, monophasic synovial sarc)
Figure 1  Malignant rhabdoid tumor in soft tissue arising in the paravertebral region of a 2-year-old girl. Rounded or polygonal tumor cells have eccentric nuclei and prominent nucleoli. Glassy eosinophilic cytoplasm containing hyaline-like inclusion bodies is evident.

Figure 2  Immunohistochemical findings of the same case as in Fig. 1. Tumor cells are positive for (a) vimentin and (b) cytokeratin in their cytoplasm. (c) Tumor cells also have positive reaction products for epithelial membrane antigen.
Epithelioid sarcoma

Differentiating extra renal Rhabdoid tumour from other DDx

- INI 1 immunohistochemistry – loss of nuclear staining.
- FISH shows loss of SMARCB1 at 22q11.2

- Other similar tumour in infants/kids is epithelioid sarcoma, chordoma
- Central necrosis, CD34+, beta catenin membrane +
Extra renal Rhabdoid Tumour

- Diverse IHC profile
- Positive – Vimentin, EMA, CK
- Less commonly S100, CD99, NSE
- Negative – Desmin, Myogenin, Beta catenin, CD34
Rhabdomyoblastic vs Rhabdoid

Primitive mesenchymal cells showing varying degree of differentiation towards skeletal muscle. Bright eosinophilic cytoplasm with cross striations. Can have variable shapes. Desmin, MyoD1 and Myogenin +
Conclusion

• Over to Ming