Pediatric Parapharyngeal Tumors

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Parapharyngeal Space

- The layers of the deep cervical fascia split to envelop different structures of H&N, leading to the formation of different fascial spaces
- The suprathyroid area encompasses the deep hidden spaces between the skull base & hyoid bone
- Parapharyngeal space PPS is the centrally located suprathyroid space
- It is incompletely ensheathed by the deep cervical fascia
  - Can be easily displaced by surrounding lesions
- It is a fat containing space
  - Can be easily identified radiologically
Parapharyngeal Space

- Anatomically, it is divided into:
  - Prestyloid part: PPS proper
  - Poststyloid part: Carotid space CS
- The structures found in each space can give rise to different pathologies.
- PPS contains:
  - Minor salivary glands
  - Lymphoid tissue of the Waldeyer’s ring
  - Mucosal surface related to the nasopharynx
- CS: contains:
  - Carotid sheath: ICA, IJV, Cranial nerves

Parapharyngeal Space

*Pre-styloid*

*Post-styloid*
**Parapharyngeal Space**

- PPS is a relatively rare site of primary pathology specially in children
- Lesions are more commonly due to secondary extensions from the surrounding fascial compartments:
  - Carotid space: CS
  - Masseteric space: MS
  - Parotid Space: PS
  - Pharyngeal mucosal surface: Naso & Oropharynx
  - Retropharyngeal space: RP
  - Prevertebral spaces: PVS
- They can also be an extension from:
  - Infratemporal fossa
  - Skull base

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**Pediatric Neck Mass**

- **Congenital**
  - Thyroglossal Duct Cyst
  - Branchial cleft cyst
  - Cystic hygroma
  - Dermoid cyst
  - Thymus

- **Acquired**
  - Infectious/Inflammatory
    - Bacterial adenitis
    - Deep neck abscess
    - Mycobacteria
    - Viral adenitis
    - Cat scratch
    - IMN
    - Kawasaki
  - Neoplasms
Congenital Pediatric PPS Lesions

- **Teratoma, Hamartoma:**
  - Teratomas have origins in totipotential germ cells,
  - H&N teratomas are exceptionally rare comprising less than 2% of all congenital teratomas

- **Congenital cervical anomalies:**
  - Can rarely originate there: First branchial arch cyst and extend from parotid space
  - Extension from neck cysts to the PPS is more common: Second branchial arch cyst

- **Congenital vascular anomalies:**
  - Haemangiomas, AV malformations...
  - Lymphatic: lymphangioma, cystic hygroma, unilateral or bilateral, infra or suprathyroid
  - Venous and capillary malformations are mainly extension from surrounding spaces (parotid): lymphangioma
Congenital Pediatric PPS Lesions

- They are considered the most common lesions found in children
  - URTI are very common in children
  - Very rich lymphatic drainage of H&N: Waldeyer’s ring
- Deep neck space infections tend to follow the fascial planes of the neck
  - Can be primary PPS abscess:
    - Pharyngeal sepsis: Tonsillitis
  - Can be extension from:
    - Odontogenic infections in MS
    - Infective retropharyngeal node of Rouviere
**Inflammatory Pediatric PPS Lesions**

- PPS tumors in adults form only 0.5% of all H&N tumors
  - Most of them are benign lesions: 70-80%
- PPS neoplasms are even more rare in children
- However, there is still a wide variety of tumors that can be found in children, as uncommon miscellaneous case reports
- All tumors of PPS tend to present late due to the deep location of the space:
  - Lesion has to be more than 2cm to present at the retromandibular area
- The close relation of PPS to the skull base, can pose difficult problems during surgery, even if they are benign
- In comparison to adults, there are differences in the pathomorphologic spectrum of pediatric PP tumors
  - The salivary gland tumors & paragangliomas which dominate in adult populations, are extremely rare in pediatric populations
**Pediatric PPS Neoplasms**

- PPS tumors represent a heterogeneous group of uncommon neoplasms
- Benign tumors are generally very rare. A wide variety of tissues give rise to a wide variety of uncommon lesions.
- Primary malignant tumors of PPS in children are also rare
- More commonly they are an extension of malignancy from surrounding spaces
- Unfortunately, these tumors tend to be locally advanced by the time they are diagnosed mainly because the clinical presentation is non-specific

**Pediatric PPS Lesions**

- **Benign:**
  - Commonest: neurogenic: neuroblastic tumors, neurofibroma
  - Lipoma
- **Malignant:**
  - Lymphoma:
    - extra-nodal NHL, H&N most common site, Waldeyer’s ring
    - Burkitt’s lymphoma
  - Soft tissue sarcomas: rhabdo, chondro, fibro, hemangio, neuro
  - Rhabdomyosarcoma: H&N is the most common site of origin: orbit, nasopharynx, PNS
  - Neuroblastoma
  - Nasopharyngeal carcinoma: Undifferentiated
Pediatric PPS Neoplasms

I- Lymphoma:
- Lymphomas are the third most frequent type of childhood cancer
- Hodgkin lymphoma
- Extra-nodal NHL, H&N most common site with involvement of the Waldeyer’s ring
- Fever, night sweats and loss of weight
- Burkitt’s lymphoma

II- Sarcomas of the soft tissue and bone:
- Heterogeneous group of malignant diseases
- HN sarcomas are difficult to treat
- Most common site is masseteric space, from where it directly extends to PPS
- Any type of soft tissue sarcomas can be found: rhabdo, chondroma, fibro, hemangio, neuro, lipo...
- Rhabdomyosarcoma: common HN sites are: orbit, nasopharynx, PNS
Pediatric PPS Neoplasms

III- Nasopharyngeal carcinoma:
- < 1% of all childhood cancers
- Common 10-19 years
- Undifferentiated NPC is commonest: advanced locoregional spread and distant metastases
- PPS involvement may occur through:
  - Extension of the mucosal lesion to PPS occurs when the tumor breaches the pharyngobasilar fascia: T2b according to AJCC TNM classification 6th edition (2002):
    - Early detection, while the tumor is confined to the soft tissues of the nasopharynx is essential
  - The primary echelon of lymphatic drainage from the nasopharynx include the lateral retropharyngeal group

As adenoidal hypertrophy is by far the most common cause of a mass in the posterior nasopharynx in children, pediatric NPC is generally not suspected clinically until late into the disease process.

Asymmetric nasopharyngeal mass may arouse suspicion of NPC in children.

Subtle involvement of PPS may be difficult to appreciate:
Pediatric PPS Neoplasms

- IV- Sympathetic nervous system tumors: Neuroblastic tumors:
  - Derived from the primordial neural crest cells (sympathogonia) that form the sympathetic nervous system
  - Neuroblastic tumors are the third most common cause of solid tumors in early childhood
  - Cervical tumors account for only 1-5% of cases of neuroblastomas
  - Neuroblastic tumors are remarkable for their varied behavior
    - Differ in their degree of cellular and extracellular maturation
    - Precursor cells may remain undifferentiated = malignant, or they may mature = benign

Neuroblastic tumors

- They define a spectrum of sympathetic neuroectodermal tumors
  - Ganglioneuroma GN:
    - Composed of gangliocytes & mature stroma. occur in children ≈ 7 years and tend to behave in a benign fashion.
  - Ganglioneuroblastoma GNB:
    - Composed of both mature gangliocytes and immature neuroblasts and has intermediate malignant potential
  - Neuroblastoma NB:
    - Is the most immature, undifferentiated & malignant: aggressive and occur in younger patients < 2 years. It is considered the third most common pediatric malignancy, after leukemia and central nervous system tumors.
Case 1: SC GN

- 10 year old, painless right sided neck mass, 2 months duration
- FNAC: inconclusive
- CT scan shows:
  - A hypodense mass, 4x3cm, in the carotid space of both suprathyoid and infrahyoid neck
  - The lesion is not enhancing
  - It is deep to the carotid sheath and does not cause splaying of the ICA and IJV
Case 1: SC GN

- Intraoperatively, the mass was found posterior to the carotid sheath, originating from the sympathetic chain, which had to be sacrificed.
- Patient developed Right sided Horner’s syndrome postoperatively.
- Pathology proved to be: Ganglioneuroma.
Case 2: SC NB

- 7 month old female, presented with stridor, dysphagia and regurgitation of 3 months duration, followed by appearance of left sided painless cervical mass, of progressive in course.
- Examination revealed well circumscribed, firm, freely mobile, lobulated swelling, extending from PPS to the clavicle down.
- FNAC repeatedly: acellular and inconclusive
- CT scan: moderately enhancing 5x5cm solid mass with CA anterior displacement and IJV lateral displacement
Case 2: SC NB

- Intraoperatively: mass was posterior to the carotid vessels, pushing IJV & X laterally
- Complete surgical excision was done at the expense of sacrificing the sympathetic nerve trunk from which it was originating
- Pressure Symptoms were immediately relieved postoperatively
One characteristic feature of NB is the formation of Homer Wright rosettes.

Pathological examination aiming at prognostic evaluation include:
- The Shimada classification have unfavorable group or favorable group, on the basis of the combination of:
  - Patient age: <1.5 yr, 1.5-5yr, >5yr
  - MKI: Mitosis and karyorrhexis of the tumor cells
  - Cellular and stromal maturity and differentiation
- Pediatric Oncology Group (POG) classification: is based solely on the degree of differentiation of the different histologic elements.
- Bone is the most common site of metastasis, and bone metastases are present in up to two-thirds of patients at the time of diagnosis.
Neuroblastoma NB

- International Neuroblastoma Staging System (INSS): 1986
  - Based on clinical, radiologic, and surgical features
- 30% of all NBs display Myc-N amplification, a proto-oncogene, that indicates rapid tumor progression and a poor outcome
- By combining the Shimada classification, INSS stage, and some genetic features, it is possible to stratify NB into:
  - Low-, intermediate-, and high-risk groups.
- Treatment is based on risk group.

Case 2: SC NB

- Horner's syndrome developed after surgery but resolved spontaneously within 6 months
- Staging was done:
  - Lesion in Sacrum, +ve Myc-N amplification,
  - High risk group: Stage IV: Chemotherapy
Case 3: Teratoma

- A 15 year old male patient, presented with a left sided squint and diplopia of 3 months duration.
- On examination he had a left sided lateral rectus paralysis, left sided parapharyngeal swelling pushing the left tonsil medially, with a mass in the left submandibular area, 5x5cm, firm in consistency, non tender and non pulsatile.

CT scan & MRI: showed heterogenous densities of soft tissue, fluid and fat in PPS with widening of the foramen ovale and parasellar extension

FNAB inconclusive: How is diagnosis made?
Case 3: Teratoma

- The approach to such lesion should be tailored according to the extent and nature of the lesion.
- Both intra- and extra-cranial parts were tackled. The intracranial part was only decompressed. The extracranial part was completely removed without the need of mandibulotomy, due to the decompression of the cystic component of the lesion.
Pediatric Neck Mass

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    - Dermoid cyst

- **Acquired**
  - Infectious/Inflammatory
  - Neoplasms

**Diagnosis of Pediatric PPS Neoplasms**

- Sarcomas of the soft tissue and bone occur more commonly than malignant epithelial tumors within H&N region in children.
- Rhabdomyosarcoma or lymphoma is the more likely diagnosis in younger children whereas NPC is more common in adolescents.
- Solid tumor of the MS in a child should prompt the diagnosis of a sarcoma until proven otherwise.
**Diagnosis of Pediatric PPS Neoplasms**

- **A- Clinical Diagnosis:**
  - Due to the rarity of these tumors, the clinical experience in diagnosis and treatment of this condition is limited.
  - The clinical symptoms of PPS tumors are versatile and non-specific, depending on the exact origin, nature and extensions.
  - Presentation is usually delayed as PPS is deeply situated:
    - *Tumors must be at least 2cm in size before causing a detectable bulge.*
  - Unfortunately, malignant tumors tend to be locally advanced by the time they are diagnosed.
  - The therapeutic implications of a delayed diagnosis of malignant lesion in this area can be significant.

**Workup of Pediatric PPS Neoplasms**

- **B- Imaging:**
  - Imaging is a cornerstone in diagnosing PPS masses.
  - Complementary MRI and CT scan are necessary.
  - Imaging studies should answer the following questions:
    - Space of origin of the lesion
    - Tissue characterization for possible pathological correlation
    - Vascularity of the lesion
    - Relation to great vessels
    - Relation to surrounding structures:
      - Other spaces
      - Deep lobe of parotid
      - Skull base
      - Mandible
Workup of Pediatric PPS Neoplasms

- B- Imaging:
  - A spatial algorithm for the origin of the lesion should be thought of:
    - Localize the epicenter of the lesion
    - Define the direction of displacement of PPS fat displacement
    - Define the imaging findings to reduce the diagnostic possibilities
  - Combining the imaging characteristics of the lesion with a limited space-specific differential diagnosis, often allows a narrower differential diagnosis

- Radiological features suggesting malignancy should be looked for specifically:
  - Better done by MRI:
    - Extraparenchymal extension
    - Intracranial invasion
  - Better done by CT scan:
    - Bone expansion or destruction
    - Skull base and mandible destruction
  - Regional lymph node metastases: retropharyngeal or jugular nodes
  - Evidence of distant metastases
Workup of Pediatric PPS Neoplasms

- C- Pathological Diagnosis:
  - No attempt at biopsy should be done before imaging
  - FNAC can be done if the lesion is accessible in the neck
  - Suspicion of a nasopharyngeal mass on imaging should guide the endoscopic biopsy under GA
  - If possible, excisional biopsy is preferred over incisional biopsy
  - CT-guided FNAC of PPS tumors can be helpful for deeper lesions seen on imaging to predict the nature of the PPS tumors which allows the surgeon and patient to plan for treatment, accordingly.
  - Definitive diagnosis is usually delayed as it requires an elaborate histological examination and probable immunohistochemistry

Pediatric PPS Lesions

- The clinical symptoms of PPS lesions are versatile and non-specific, depending on the exact origin, nature and extensions
- Definitive diagnosis is usually delayed as it requires a histological examination
- Any non-inflammatory, firm neck mass, or solid mass in imaging in a child should be considered of potential neoplastic etiology until proved otherwise.
- Solid tumor of the MS in a child should prompt the diagnosis of a sarcoma until proven otherwise: rhabdo, osteo, chondro,…
- Awaraness of occurrence of NPC in children is important. A high index of suspicion will lead to biopsy and less delay in diagnosis
Pediatric PPS Neoplasms

Take Home Message

- Neck mass in children have a wide differential, age at presentation is important
- Although rare, malignant tumors of PPS does occur in children
- Pediatric malignant tumors of the head and neck are basically curable, if early discovered
- Awareness of occurrence of malignant tumors in H&N and PPS in children, by the clinician is crucial
- Careful evaluation for radiographic features of malignancy by the radiologist is essential

Pediatric PPS Neoplasms

Take Home Message

- Any non-inflammatory, firm neck mass, or solid mass in imaging in a child should be considered of potential neoplastic etiology until proved otherwise
- Rare PPS location of uncommon lesions, gives rise to difficulty in diagnosis, and final diagnosis is not made until excised tumor is examined pathologically
- A high index of suspicion will lead to less delay in diagnosis and better prognosis
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THANK YOU

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