AN UNUSUAL CASE OF STRIDOR

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1 year 8 months old boy toddler brought with :-

- History of noisy breathing since 8 months of age, aggravated over 4 days
- Wet cough, difficulty in breathing - 4 days
- Vomiting, poor oral intake - 3 days
- Fever - 1 day
• Apparently normal until 8 months of age

• **Noisy breathing** – initially whistling type, gradually progressive, later harsh quality, less in prone position and more in supine position / crying.

• Noisy breathing **aggravated** - 4 days associated with wet cough, chest indrawing.

• H/o frequent respiratory tract infections +

• No history of foreign body ingestion, oily/loose stools/ aspiration/ difficulty in feeding/ weight loss/ freq ear discharge / skin infections.
Admitted thrice in native place and evaluated. (? Narrow respiratory tract)
ECHO / Barium swallow – normal
No treatment given

Stridor persisted...

1st child for NCM parents
Antenatal/ perinatal period – uneventful
Mild delay in milestones +
Immunized for age
On examination,

- Alert, conscious, afebrile
- RR- 40/ min, HR- 130/min, SPO2 – 98% in RA,
  BP – 90/60 mmHg, good perfusion
- Stridor +
- Increased WO B - SCR + ICR +
- Decreased voice volume
- Frontal bossing +, AF - closed
- Hypertelorism + Mild pallor +
- Macroglossia +
- Multiple café u lait spots + in trunk, limbs, neck, axilla, inguinal region.
- Hemihypertrophy of left side of tongue +
Hyperpigmented patch
• Anthropometry
  Wt – 9.3 kg (normal for age acc to IAP cls)
  Ht – 83 cm (normal for age)
  HC – 49cm

• RS- Bilateral air entry equal, no wheeze
• CVS – normal heart sounds, no murmurs
• P/A – soft, non-tender, liver just palpable
• CNS – normal tone / activity
Basic investigations

- Hb – 9.2 g/dl, TLC - 5300
- PLT- 4.76 lakhs , CRP + (9)
- Serum creatinine/ electrolytes/ Ca / P / liver enzymes – normal
- Chest X ray – Bronchopneumonia

- IV antibiotic (Advent) started.
- A few hours after admission, SPO 2 dropped and distress with stridor worsened and he was shifted to ICU.
Working diagnosis

UPPER AIRWAY OBSTRUCTION

- ? Vocal cord papilloma
- ? Vocal cord palsy
- ? Subglottic stenosis
- ? Vascular ring
BRONCHOSCOPY

- Soft tissue mass in the supraglottic area – left side involving aryepiglottic fold, with external compression of laryngeal inlet, upper border extending into cricopharynx and lower border extending into upper oesophagus.
- External compression at the level of fourth tracheal cartilage anteriorly.

- All 4 previous possibilities ruled out !!!!
Cect chest
Indentation of trachea and tracheal narrowing
MRI + CECT brain/ neck/ chest
LYMPHOHEMANGIOMA / PLEXIFORM NEUROFIBROMA????

Most likely DIAGNOSIS - Laryngeal plexiform neurofibroma -
• Child underwent **Microscopic Laryngeal Biopsy** followed by Laser Excision of the mass.

• **Histopathology revealed** benign lesion.

• **Now 1 month post procedure. Child is doing well. No stridor.**
Pediatric laryngeal neurofibroma: Case report and review of the literature

- DOI: 10.1016/j.ijporl.2013.10.047
- SB Chinn, RM Collar, JB McHugh.

- Review of the world literature since 1940.
- Pediatric LNF was identified in 62 cases.

- MC presenting symptom - stridor and MC location of the tumor in the larynx - aryepiglottic fold.
- Recent reports - increased utilization of endoscopic resection with reduced need for tracheostomy
Plexiform Neurofibroma of the Larynx: A Challenging Management Dilemma

- Nerurkar NK, Kapre G.

- Reported 2 cases of pNFs in children
- Both children presented with large supraglottic masses which interfered with breathing and swallowing.

- Endoscopic resection better than open surgery.
- Initial devascularization with a laser followed by microdebrider-assisted debulking of the tumor mass.
DISCUSSION
• Type 1 is neurofibromatosis (NF1) also known as Von Recklinghausen’s disease in which plexiform neurofibromas may occur.

• Plexiform N eurofibroma (pNF) is a benign tumor of peripheral nerves - involves single or multiple nerve fascicles that arise from major nerve branches.

• pN Fs of the larynx are uncommon tumors with special predisposition for the supraglottic larynx.

• Rare cause of pediatric stridor.
Criteria for NF1

2 out of these 7 features must be present:
1) 6 or more café au lait macules over 5mm
2) Axillary or inguinal freckling
3) 2 or more iris Lisch nodules
4) 2 or more neurofibromas or 1 plexiform NF
5) Osseous lesion – sphenoid dysplasia/cortical thinning
6) Optic glioma
7) A first degree relative with NF1
- MC sites- Orbital/temporal region, paraspinal, anterior mediastinum, supraclavicular region.

- Age of a child correlates with the location of pNFs:
  1. Diffuse pNFs of the head and neck < 1year
  2. Diffuse pNFs of other parts of the body - before adolescence.
  3. Deep nodular pNFs infrequent in early childhood

- Pain, functional impairment, cosmetic deformity.

- Hyperpigmentation Overlying a Plexiform Neurofibroma (HOPN)

- Malignant transformation (5 – 10 % cases) – MPNS tumour. Tend to grow back at the site of removal.
Made up of a variety of cell types including neuronal axons, Schwann cells, fibroblasts, mast cells, macrophages, perineural cells and extracellular matrix materials such as collagen.
TREATMENT

- Mainstay – surgery (endoscopic resection > Open surgery)
- Retinoic acid, thalidomide
- Anti-angiogenesis drug – interferon alpha

PROGNOSIS

- pNFs can cause significant health problems and can be fatal if the tumor becomes malignant.
- Poor survival rate likely due to late detection.
Thank you
SPECIAL THANKS TO

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