Abstract: Congenital cholesteatoma with multiple cranial nerve palsy is a rare condition. We hereby report a case of congenital cholesteatoma with multiple cranial nerve palsy. We had a 20-year-old male patient presented to us with multiple cranial nerve palsy (9, 10, 11, 12) on the right side. ENT examination was normal except a swelling over right parotid region. MRI showed a right sided parapharyngeal abscess with right mastoiditis. We proceeded with tympanomastoid exploration and removed a large cholesteatoma from the mastoid tip. Postoperatively, the patient recovered completely from cranial nerve palsy. We report this case because of its rarity.

Keyword: cholesteatoma, congenital cholesteatoma, cranial nerve palsy

A 20-year-old male patient admitted initially in neurology ward with change in voice 2 weeks and difficulty in swallowing for 2 weeks. History of fever was present. History of cough and choking spells were present. The patient was referred to ENT department as a case of multiple cranial nerve palsy (9, 10, 11, 12) on the right side with Internal jugular vein thrombosis.

On Examination:
- Patient had a Neck-swelling in the region of right parotid 4x5 cms
- Lifting the right earlobe - Skin over the swelling warm
- Ear - Both Tympanic membrane were intact and mobile
- Nose - normal
- Throat – oral cavity – tongue deviation to right
- Oropharynx – Soft palate movement restricted on right side
- Indirect Laryngoscopy – right vocal cord palsy present

Cranial nerve examination:
- Cranial nerves 1, 2, 3, 4, 5, 6, 7, 8 - normal
- Cranial nerves 9 & 10 – uvula in midline, on phonation uvula deviated to left. Gag reflex absent.
- Cranial nerve 11 - weakness of right trapezius and right sternocleidomastoid muscles.
- Cranial nerve 12 - deviation of tongue to right side - wasting of muscles present on the right side of tongue

PURE TONE AUDIOGRAM – normal hearing sensitivity in both ears.

A CASE REPORT ON CONGENITAL CHOLESTEATOMA WITH RARE PRESENTATION

SARAVANA SELVAN
Department of ENT, MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL
MRI REPORT

- Features suggestive of coalescent mastoiditis with abscess in the parotid and parapharyngeal spaces of neck on the right side.

A CASE REPORT ON CONGENITAL CHOLESTEATOMA WITH RARE PRESENTATION

Neurosurgery opinion

- A case of coalescent mastoiditis with abscess in right parotid and parapharyngeal region with right lower cranial nerve palsy. No neurosurgical intervention needed.

TREATMENT

Hence we planned for a tympano mastoid exploration. After raising tympanomeatal flap middle ear was inspected and was normal; hence proceeded with mastoid exploration. We did a mastoidectomy and was surprised to see a large cholesteatoma sac in the mastoid cavity arising from mastoid tip and was completely removed. In the post operative period patient had complete recovery.

- Congenital Cholesteatoma sites are recently classified into 4 sites:
  - Cerebello Pontine angle
  - Petrous pyramid
  - Jugular fossa
  - Middle ear

Origin of Cholesteatoma in our case would be jugular fossa.

CONGENITAL CHOLESTEATOMA (CC)

- **DERLACKI AND CLEMIS CRITERIA:**
  - Pearly white mass medial to intact tympanic membrane
  - Normal pars tensa and flaccida
  - No h/o otorrhoea, perforation or previous otological procedure

Levenson modified the above criteria by telling that Prior bouts of otitis media does not exclude CC.

Incidence - 1% to 5%

- Most common presentation of CC is a white retro tympanic middle ear mass in the antero superior quadrant.

THEORIES OF PATHOGENESIS OF CC

Embryonic cell rest theory: At weeks 10-33 of development there is epidermoid formation in the anterior epitympanum of the fetal temporal bone epidermoid cells should involute after week 33. Congenital cholesteatoma results from failure of this tissue to involute Only explains how they form in the anterior superior quadrant.

- Invasion theory: Misdirection of migrating ectodermal cells from the developing external auditory canal into the middle ear. Forms the building block for a cholesteatoma.

Metaplasia theory: Squamous metaplasia in the middle ear due to chronic inflammation of middle ear in CSOM.

- Implantation theory: Squamous epithelium is implanted into middle ear either from trauma or recognised healed perforation.

STAGING

- Stage I - only 1 quadrant involved, no ossicular involvement, no mastoid extension (40%)
- Stage II - involvement of multiple quadrants, no ossicular involvement, no mastoid extension (14%)
- Stage III - ossicular involvement/erosion but no mastoid extension (23%)
- Stage IV - mastoid extension (23%)

INVESTIGATIONS

Audiometry, Tympanometry

CT of temporal bone

REFERENCES:

1. HOUSE HP. AN APPARENT PRIMARY CHOLESTEATOMA ACASE REPORT. LARYNGOSCOPE 1953;63(3):712-3
3. PAPERELLA ET AL. CONGENITAL CHOLESTEATOMA. O.CNA 1978;11:113-20