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ENT manifestations of Klippel-Feil syndrome: Report of two cases and literature review

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Abstract

Aim: Klippel-Feil syndrome (KFS) is a rare embryopathy characterized by fusion of cervical vertebrae with classical triad : low posterior hairline, short neck, and decreased cervical range of motion . KFS is commonly associated with many congenital malformations. ENT anomalies represent a common feature of them. Our purpose is to describe ENT manifestations of this rare syndrome through tow observations, as well as a of literature review.

Materials and methods: Retrospective study over ten years of 55 children with syndromic deafness seen in the ENT department of Tlemcen, Algeria between 2014 and 2024. We recorded 2 cases, described all ENT manifestations, various complementary examinations and treatment.

Results: Case1: A 3 years old girl, growth retardation, sleep apnoea and poor speech. Triad of KFS. Sprengel's anomaly and dorsal kyphosis. ENT features: adenoid facies, high-arched palate, low-set ears and bilateral secretory otitis media with flat tympanogram. ABR / ASSR: slight conductive deafness on the right (40 dB) and profound on the left. petrous bones CT-scan did not reveal any hearing malformations. A cervicothoracic CT-scan confirmed fusion of the C1-C2 vertebrae. Management includes adenoidectomy, myringotomy with T tube insertion allowing normalization of hearing on right. Speech therapy and audiometric monitoring every 6 months were recommended.

Case2: 4years old boy, poor speech, classic triad of KFS, adenoid facies, nasal dorsum cyst with fistula, pretragic fistula, low-set ears and secretory otitis media. ABR/ASSR: slight conductive hearing loss on left and a moderate on right. CT-scan of the face: superficial nasal dorsum cyst without intracranial extension. In addition to the cystectomy, the management was identical to previous case.

Conclusion: ENT manifestations of KFS should be known and sought by practitioners. Deafness is frequently encountered in KFS and should be screened. The management of ENT manifestations helps to reduce morbidity's patients of KFS.

Keywords

“Klippel-Feil syndrome; Deafness; ABR / ASSR, fusion's cervical vertebrae; embryopathy”.

Biography

-MEHTARI NASRINE SIHAM, born on December 13, 1981 in Oran Algeria

- ENT doctor since 2010
- Assistant professor at the Tlemcen University Hospital since 2011
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