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Cholesteatoma Complicated by Facial Nerve Palsy Associated with Ipsilateral Michel Aplasia: A Rare Case and Surgical Challenge

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Abstract **Case Report**

Introduction: Middle ear cholesteatoma is a locally destructive disease that may lead to serious complications, including facial nerve palsy. Its association with congenital inner ear malformations such as Michel aplasia is extremely rare. ***Case Presentation:*** We report the case of a 50-year-old patient presenting with chronic right-sided otorrhea complicated by a peripheral facial nerve palsy graded II according to the House–Brackmann classification. Temporal bone CT scan confirmed an extensive middle ear cholesteatoma with facial canal erosion and revealed an ipsilateral Michel aplasia. ***Management and Outcome:*** The patient underwent right canal wall down mastoidectomy with complete removal of the cholesteatoma. The facial nerve was carefully identified and preserved. Postoperative evolution was uneventful, with stable facial nerve function and good clinical recovery. ***Discussion:*** This rare association presents both diagnostic and surgical challenges due to the absence of inner ear anatomical landmarks and the potential for aberrant facial nerve course. ***Conclusion:*** Careful preoperative radiological assessment and meticulous surgical dissection are essential to achieve disease control while minimizing the risk of facial nerve injury.

Keywords: cholesteatoma; facial nerve palsy; Michel aplasia; temporal bone CT; otologic surgery.

INTRODUCTION

Cholesteatoma of the middle ear is a keratinizing squamous epithelium lesion characterized by progressive bone destruction. Although usually benign in histological terms, it may lead to severe complications including facial nerve palsy, labyrinthine fistula, and intracranial involvement.

Michel aplasia represents the most severe form of congenital inner ear malformation, resulting from an early arrest of otic development during the third week of gestation. It is characterized by a complete absence of cochlear and vestibular structures and is associated with profound congenital sensorineural hearing loss [1,2].

The coexistence of acquired cholesteatoma and Michel aplasia is extremely uncommon. We report a case illustrating the diagnostic and surgical challenges posed by this exceptional association.

CASE PRESENTATION

A 50-year-old male with no significant medical history presented with long-standing right-sided foul-smelling otorrhea and progressive hearing loss. He recently developed facial asymmetry. Clinical examination revealed a right peripheral facial nerve palsy graded II according to the House–Brackmann classification. Otoscopy demonstrated keratin debris within the external auditory canal, suggestive of cholesteatoma.

High-resolution CT of the temporal bone demonstrated opacification of the right middle ear and mastoid cavity with ossicular chain erosion and erosion of the facial canal (Figures 1 and 2). In addition, CT revealed a complete absence of the cochlea, vestibule, and semicircular canals on the right side, consistent with ipsilateral Michel aplasia.

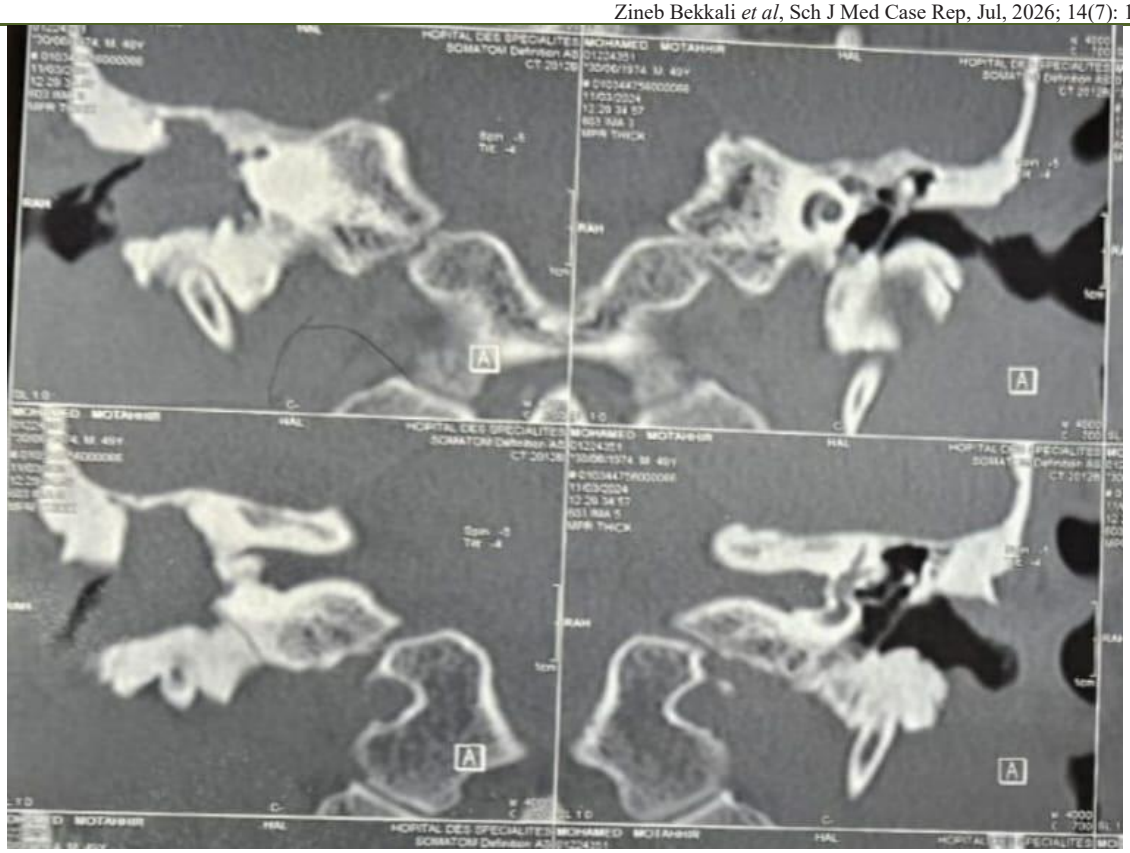

Figure 1: Coronal CT scan of the temporal bone showing a soft tissue density filling the right middle ear and mastoid cavity, consistent with cholesteatoma. Ossicular chain erosion and facial canal involvement are also noted

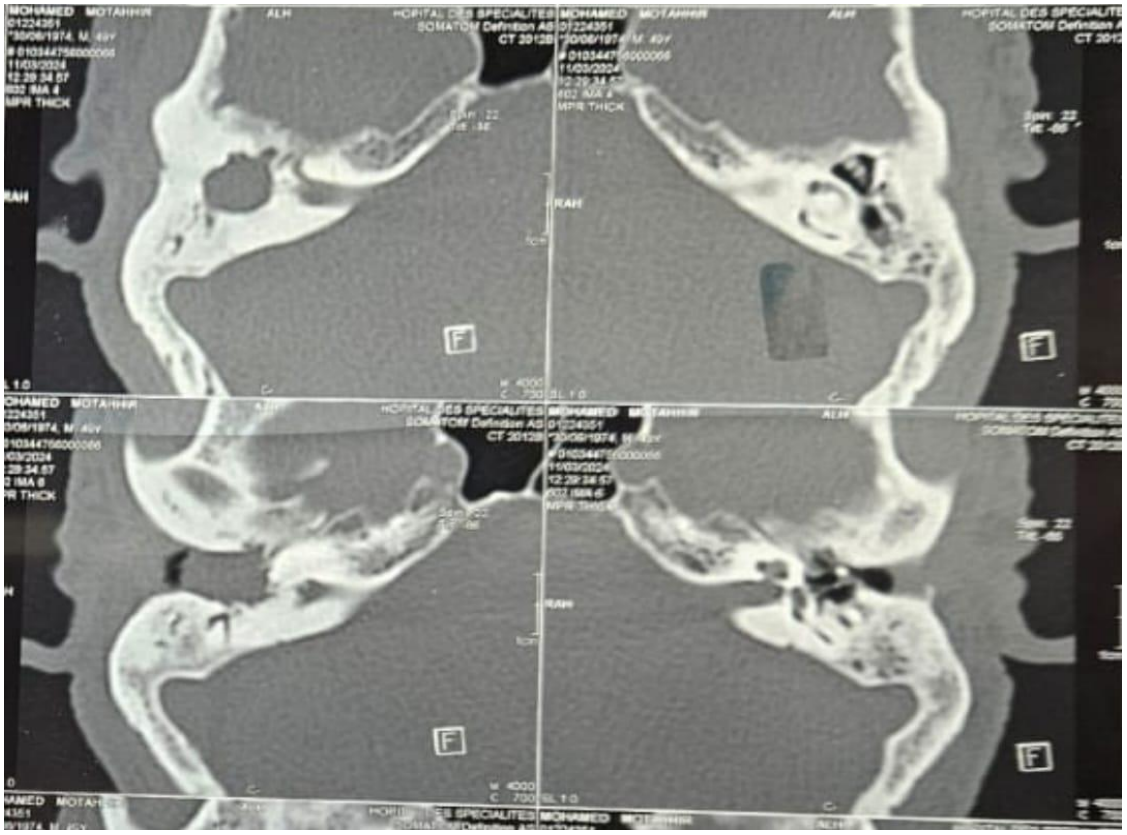

Figure 2: Axial CT scan showing complete absence of the cochlea, vestibule, and semicircular canals on the right side, consistent with ipsilateral Michel aplasia

The patient underwent surgical treatment under general anesthesia consisting of a right canal wall down mastoidectomy with complete excision of the cholesteatoma. Intraoperatively, careful dissection was performed given the altered anatomy, and the facial nerve was identified and preserved. Complete macroscopic removal of the cholesteatoma was achieved without intraoperative complications.

The postoperative course was uneventful. Facial nerve function remained stable at House–Brackmann grade II, with no deterioration. Early follow-up demonstrated good healing of the surgical cavity with no clinical evidence of residual disease. The patient was placed under regular clinical and radiological surveillance.

DISCUSSION

Cholesteatoma is a locally aggressive disease capable of causing bone erosion and both intracranial and extracranial complications. Facial nerve palsy is an uncommon complication, occurring in approximately 1–3% of cases, typically due to erosion of the fallopian canal [3,5]. High-resolution CT is essential for diagnosis, extension assessment, and detection of associated anomalies [6].

Michel aplasia is a rare congenital anomaly resulting from early developmental arrest of the inner ear, leading to complete absence of labyrinthine structures and profound deafness [2,7]. The coexistence of this malformation with acquired cholesteatoma is exceptional and adds considerable complexity to surgical management.

Surgical Challenge

The association of cholesteatoma with Michel aplasia creates significant surgical difficulty. The absence of inner ear structures eliminates key anatomical landmarks typically used for orientation during otologic surgery, particularly in identifying the facial nerve. Furthermore, the facial nerve may present an aberrant course in the context of congenital malformations, a risk further compounded by cholesteatoma-induced erosion of the facial canal, rendering the nerve more vulnerable to inadvertent injury.

Therefore, surgery relies on careful preoperative imaging analysis and meticulous dissection, with the primary objective of achieving complete disease removal while preserving facial nerve integrity [4,6].

CONCLUSION

This case highlights a rare and challenging association between middle ear cholesteatoma and Michel aplasia. It underscores the importance of thorough radiological evaluation and a well-planned surgical strategy to ensure safe and effective management of such complex cases.

Conflict of Interest

The authors declare no conflict of interest.

Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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