Giant cholesteatoma and concurrent fungal infection: A literature review and case report

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Background

Cholesteatoma is a non-neoplastic disease characterised by the proliferation of keratinising squamous cell cysts in the middle ear and surrounding areas. The incidence is 9 to 12.6 cases per 100,000 in adults and 3 to 5 per 100,000 in children. Giant cholesteatomas have been reported in literature however a formal review systematically assessing the variability in the modes of presentation and management and synthesising the results is not present. This study aims to undertake this whilst also reporting a case of giant cholesteatoma presenting to our department.

Methods

PROPSPERO registration number: CRD42018094560. Systematic searching was conducted using the key terms ‘Cholesteatoma’, ‘Skull base’, ‘Giant’, ‘Massive’, ‘Extensive’ limited to case reports and case series. Data was extraction was conducted with 2 independent reviewers. Data surrounding demographics of patients, mode of presentations and management was collected and synthesised in line with the Newcastle-ottawa/Murad classification. A single case of giant cholesteatoma from our centre was also reported.

Results

32 cases from 26 studies were identified. The cases reported were from 1985 to 2017 with an age of the patients ranged from 15 to 84. Presentations ranged from simple hearing loss and ear discharge to collapse and florid sepsis. There was a varying size corresponding to the amount of involvement of key structures within the middle ear and the cranium. 10 patients had a history of previous mastoid surgery and all aside from the studies with incomplete data, all patients underwent surgical intervention.

Discussion

Giant cholesteatoma has not previously defined. From the review we propose any cholesteatoma greater than 60mm in a single dimension can be considered a giant cholesteatoma. Regarding formal classification, a consensus should be reached on a single system encompassing all aspects. Surgical treatment was mostly successful even in the case of advanced disease. Surgical intervention should remain the gold standard with a modified radical mastoidectomy as the most favourable approach. There is no agreement on long term follow up with some centres opting for interval scanning and other centres conducting outpatient after follow up. Considering the cost of follow up – lifelong 6 monthly outpatient follow up (£2932) or yearly MRI scanning (£3956) in combination is more cost effective than having the patient suffer with a intracranial complication necessitating a craniotomy (£22,435).

Recommendations

• A consensus should be reached on a single system encompassing all aspects for classification of cholesteatoma.
• Giant cholesteatoma should continue to be treated surgically with mastoidectomy.
• Definitive guidance is required on the management and follow up giant cholesteatoma – a delphi study or nested case control in a cohort of a database of all giant cholesteatomas would be appropriate.
• Until more evidence is available 6 monthly follow up in the outpatient clinic with 5 yearly imaging may be the most appropriate method up to prevent large recurrence necessitating extensive surgery.

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