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Lateral skull base surgery in a pediatric population: A 25-year experience in a referral skull base center

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ABSTRACT

Objective: To analyze the pathology and surgical outcomes of lateral skull base (LSB) procedures in a pediatric population.
Study design: Retrospective case review in a referral skull base center.
Methods: Charts of pediatric patients who underwent defined LSB procedures from 1983 to 2015 for various pathologies were evaluated at our center. A systematic review of literature was performed and our results were compared with the literature.
Results: 63 patients presented with 65 diseased ears. The mean age was 13 years. 29 (44.6%) presented with hearing loss and 28 (44.4%) and chronic otorrhea. The most common pathology was petrous bone cholesteatoma (27, 42.5%) followed by vestibular schwannoma (10, 15.8%). Subtotal petrosectomy (24, 35.8%) was the most common surgical procedure followed by, transotic (18, 26.8%). The facial nerve function was preserved in 45 (67.1%) and the hearing in 28 (41.7%) cases respectively. No major complications, including mortality was encountered in our series.
Conclusion: In rare and extensive pathologies involving the skull base in a pediatric population, the surgeon is posed with the dilemma of trying to achieve facial and hearing preservation while dealing with total tumor clearance. Mastery over LSB procedures can ensure complete disease clearance with optimal functional outcomes.
Level of evidence: 2b.

1. Introduction

Pathology that involves the deep parts of the LSB like the cochlea-vestibular system, facial nerve (FN), internal auditory canal and jugular bulb not only causes functional disturbances that can be devastating in children but also makes extirpation of such tumors a challenging proposition. Considering the early age of the patient, the treating practitioner will always be posed with the dilemma of whether to achieve functional preservation (hearing and FN function) or disease clearance. Fortunately, over the last few decades due to rapid advances in neuroradiology and neuroanesthesia, development of rational surgical approaches and better instrumentation, the objective of LSB surgery has moved from solely being focused on tumor removal to also preservation of cranial nerve functions. Children with LSB pathology have benefitted most from this development because in them, any functional deficit at that age and which remains over a very long period of their life, has serious social and psychological consequences.

Pediatric LSB surgery pathology and procedures are relatively rare and there are very few series dealing with this subject [1–8]. We present one of the largest series of pediatric LSB surgery published in English literature and discuss the special considerations in dealing with children with pathologies of the LSB.

2. Material and methods

Of the 4523 skull base surgeries performed from January 1991 to August 2015, charts of all patients less than 18 years of age undergoing LSB surgery in our center, were reviewed. The inclusion criterion was surgical treatment of extensive pathology that extended beyond the confines of the middle ear cleft and involved

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the petrous bone, the jugular bulb or destroyed large areas of the middle and posterior dural plates, that required at least a Subtotal Petrosectomy (STP) or any other procedure that involved a more extensive dissection. Exclusion criteria were: 1) Surgery for routine middle ear conditions, 2) LSB pathology in children that were not operated upon, 3) Malignant tumors (due to necessity of adjuvant treatment that could alter the outcome of the study), 4) Patients with incomplete case records, follow-up of less than one year or those lost for follow up.

All patients underwent complete preoperative otoneurologic evaluation followed by audiometric exam. FN function was graded preoperatively according to the House-Brackmann (HB) grading system. Audiometric studies included four-frequency (500, 1000, 2000 and 4000 Hz) pure tone average (PTA) for bone-conduction (BC), air-conduction (AC) and speech discrimination scores (SDS). The modified Sanna classification of hearing scores were used for documenting and analyzing audiological data [9,10]. A high resolution computed tomography scan of the temporal bone and magnetic resonance imaging were obtained as a part of diagnostic battery.

Petrous bone cholesteatomas (PBC), were classified according to the Sanna classification [11,12]. In Vestibular schwannomas (VS), the tumor was graded according to the Tokyo consensus meeting [10]. The surgical procedures included STP, transotic approaches (TOA), translabyrinthine approaches (TLA), transcochlear approaches (TCA), infratemporal fossa approaches (ITFA), transmastoid-infralabyrinthine approaches (TM-IL). In some cases, cochlear and auditory brainstem implants were performed along with the abovementioned procedures. All the procedures have been described in detail elsewhere [13–15].

Intraoperatively, operating time and complications were recorded. Postoperatively, average length of hospital stay and complications were recorded. FN and hearing status at the end of one year follow-up were obtained from the charts.

2.1. Review of the literature

A review of the literature was done using a PubMed Search using relevant search words. The demographic data, pathology, performed surgical procedures, duration of surgery, FN function and hearing status both pre- and postoperatively, other cranial nerve status, complication rates, and follow-up time were tabulated and compared with our data.

This study was approved by the Institutional Review Board of the hospital for ethical research.

3. Results

A total of 63 pediatric patients with 65 pathological ears were included in the study. 37 patients were males and 26 were females. The age ranged from 18 months to 324 months (mean, 156 months). All patients in this series were operated. The incidence of pathology in the LSB in children was 1.4% of all the 4523 patients who presented to us with such pathology at our center. The mean duration of follow-up was 43.6 months (range:12–125). Patient characteristics are shown in Table 1.

3.1. Symptomatology

The mean duration of symptoms at the time of presentation in our center was 26.5 months. 32.3% of the patients were referred to our center for surgical intervention after a prior surgery elsewhere (Table 1). On presentation, the most common symptoms were hearing loss (44.6%) and chronic otorrhea (43.1%). Preoperatively, FN was normal (HB I) in 53 (81.5%), and HB IV and above in nine (13.8%) cases. Clinically, 23 ears (35.4%) were totally deaf. The FN function and hearing status are shown in Table 3.

Table 1
Patient characteristics and symptoms of the study population.

Population characteristics	
Patients	63
Procedures	65
Mean age	13.0 (range 1.5–18)
Males, females	37, 26
Left side, right side	38, 29
Mean symptom duration (range)	25.6 days (range 2–360)
Mean duration of follow-up	42.8 months (range 12–125)
Symptoms	
Hearing loss	29 (44.6%)
Chronic otorrhea	28 (43.1%)
Dizziness/Vertigo	17 (26.2%)
Tinnitus	9 (13.8%)
Facial nerve palsy	7 (10.8%)
Trigeminal palsy	3 (4.6%)
Lower cranial nerve palsy	6 (9.2%)
Headache	4 (6.2%)
Recurrent meningitis	2 (3.1%)
Treatment details	
Patients previously operated elsewhere	21 (32.3%)
Revision surgeries in this series	2 (3.1%)

3.2. Pathology

25 (38.5%) cases were of tumoral pathology, mostly VS and FNTs. 31 cases were non tumoral pathology most common being PBCs seen in 26 (40%). Inflammatory and infective pathology was seen in four (6.2%) cases, the rest of them being surgeries for hearing related pathology (Table 2). Among the VS, five (50%) of the tumors were sporadic and the rest were NF II. The VS were extrameatal in all but one (90%) case. All NF II cases had bilateral and larger tumors (mean 3.6 cm) compared to sporadic type (mean 2.9 cm). Four (66.7%) of the FNTs were schwannomas and all of them involved multiple segments. Most of the PBCs, 14 (53.9%), were of the infralabyrinthine type [16].

3.3. Surgical approaches

STP was the most common approach performed in 23 (35.4%), followed by TOA and TLA in 18 (27.7%) and 13 (20.0%) cases, respectively. The surgical approaches are listed in Table 2.

STPs were used commonly in PBCs, in 17 (65.4%) cases. TLA was used mainly for VS in eight (80%) cases. Three cases of petrous apex cholesterol granulomas were treated by the TM-IL approach. In one of them, the procedure was abandoned due to a prominent sigmoid sinus and a high jugular bulb. An infracochlear approach was then attempted in the same sitting but this was also abandoned due to unfavorable anatomy. Two cases of class C1 tympanojugular paragangliomas (TJPs) were treated by the ITFA type A with complete tumor removal. In another case of extensive class C3Di2 a labyrinthectomy was added to the ITFA type A. The patient had a residual tumor and after five months a TCA was performed with a sural nerve grafting despite incomplete tumor resection.

In 55 out of 60 procedures (91.7%) gross total removal was achieved. Of the incomplete resections, one was a case a 4 × 3.7 cm NF II VS where the surgery had to be stopped due to bleeding. Other partial resections included a C3Di2 TJP, chordoma (involving the clivus, infratemporal fossa, orbit and the parapharyngeal space), posterior fossa meningioma (involving the parapharyngeal space and the internal carotid artery). Another case of unsuccessful drainage of petrous apex cholesterol granuloma is asymptomatic and stable on follow-up for the last 28 months.