ABSTRACT

PTCD is a newly discovered brain malformation with only 15 cases previously reported in the literature. In our series, all three patients had an absent cochlear nerve in the facial nerve. In addition, mild cochlear dysplasia was seen in 2 patients. An accessory canal (or duplicated IAC) for the facial nerve was present in all patients, though most severe in patient 3. There is also a lipoma present within the interpeduncular cistern (white arrow). No previous case series have looked specifically at the course of the cranial nerves and the status of the labyrinth. We describe 3 new cases of PTCD and look specifically at the course of the cranial nerves in these patients, in an effort to anatomically explain sensorineural hearing loss in these patients.

RESULTS

Children with PTCD have consistent MR imaging findings, including absent 8th cranial nerves, duplicated IACs and aberrant course of cranial nerves. Furthermore, all patients had a normal vestibular labyrinth. The entity should be recognized and prognosis for successful cochlear implantation is poor.

CONCLUSIONS

REFERENCES