**INTRODUCTION**

Oto-palatal-digital syndrome is a rare X-linked dysplasia with intermediate expression in females and autosomal dominant inheritance in males. There are two types of Oto-palatal digital syndrome type-I and type-II. Type-I is characterized by hypertelorism, prominent supra-orbital ridges, abnormal pinnae, broad nasal bridge with small nose and mouth, microstomia, micrognathia and malformed and or apparently low set ears. Fibulae are absent or severely hypoplastic. The cranium shows thick, sclerotic base and delayed ossification of the vault, poor peumatization of mastoids and thick ossicles. Ribs and clavicles are usually thick and irregular or wavy. Patients have hearing loss and mental retardation. Other rare malformations reported are omphalocoele, hydrocephalous and cryptorchidism.

**CASE REPORT**

A six-year-old male child presented with one day history of clear fluid otorrhea from right ear following ear cleaning. It was not associated with fever, neck rigidity or vomiting. The child had delayed milestones and had decreased hearing since birth. The child was third of four issues, other siblings were healthy. There was no history of consanguinity. Physical examination showed child was born with a weight of 17 kg, and height 106cm. The head was dolicho-cephalic with frontal bossing and prominent supra-orbital ridges. Down slanting palpebral fissures and ocular hypertelorism was present. The bridge of the nose was depressed and broad. The mouth was small and had the appearance of "fish mouth". The right ear had a small perforation in the posterior quadrant of the tympanic membrane. Biochemical tests confirmed CSF otorrhea and he was started on intravenous broad spectrum antibiotics and tablet diamox. The CSF otorrhea subsided on third day of admission.

**DISCUSSION**

The term Oto-palatal digital syndrome (OPD) was first quoted by Fitch in 1962. OPD’s key features are anomalies development of the ear, palate and digits. Two types of OPD, type I and type II, have been described. Both are X-linked, semi dominant traits with partial expression in female heterozygote. Type-I has been mapped to Xq28; type-II is not precisely mapped, however genetic and empiric studies have suggested that the two conditions may be allelic or possibly, closely linked genes. There is phenotypic overlap with type-I showing greater severity. Type-II is characterized by larger anterior fontanelle, broad face, tall and slender legs and a small perforation of the supra-orbital ridge, prominent supratrochlear ridges, abnormal pinnae, broad nasal bridge with small nose and mouth, prominent starts of palpebral fissure, hearing loss and mental retardation.

**CONCLUSIONS**

Cerebrospinal fluid otorrhea is not previously reported in oto-palatal-digital syndrome, and hydrocephalous and dandy walker anomaly are also rarely reported.