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An Unique Oral Manifestation In Dandy Walker Syndrome- A Case Report

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Abstract: *Dandy–Walker syndrome is one of the rare congenital abnormality affecting the cerebellum of the brain. It is associated with the hypoplasia of cerebellar vermis, a cystic dilatation of fourth ventricle resulting in an enlarged posterior fossa. Although, clinical findings like macrocephaly, ataxia, nystagmus hydrocephalus, increased intra cranial pressure etc may be the usual presentations due to cerebral malformation, oral findings are rare. Here, we report a case of 15 year old boy with Dandy Walker syndrome exhibiting multiple oral manifestations.*

Keywords: *Dandy Walker syndrome, odontoma, multiple retained deciduous teeth*

I. Introduction

A variety of malformations predominantly affects the cerebellum and its derivatives. Dandy-Walker syndrome is a rare congenital paleocerebellar malformation, characterized by the hypoplasia of cerebellar vermis, and the cystic dilatation of fourth ventricle. The disorder was originally described in 1887 by Sutton. Later, it was characterized by W. Dandy and K. Blackfan in 1914 and followed by Tagart and Walker in 1942. It was C. Benda, who designated the disease as Dandy Walker syndrome in 1954 (1,2).

The syndrome is characterized by enlarged fourth ventricle, aplasia or hypoplasia of the cerebellar vermis, cyst formation in the posterior fossa and agenesis of the corpus callosum. These abnormalities are diagnosed only by the clinical imaging modalities like CT and MRI and attribute to the following clinical findings like delayed motor development in infancy, hydrocephlaus, progressive macrocephaly leading to abnormal large skull, intracranial pressure in older children, leading to irritability, vomiting, and convulsions, cerebellar dysfunction causing ataxia and nystagmus, bulging occiput. Other associated findings are cervical nerve dysfunction, anomalies of the heart, limbs, digits, urinary system, face, cleft palate and lips(3).

II. case report

A 15 year old boy reported with the complaint of aching tooth on right side of the upper jaw.. Past medical history revealed that he was born with the large head and had been treated conventionally for hydrocephalus. There was a positive history of delay in the developmental mile stones with not a single reported history of epilepsy till date. Previous medical records reported the presence of arachanoid cyst and diagnosed as Dandy- Walker Malformation . Family history revealed that he was the second son of non-consanginously married parents. The maternal age was 41 years at the time of his birth and was born at full term. As per the history, he is the only member affected in the family.

On general physical examination, the patient was afebrile, conscious, well oriented, co-operative with normal congnition and motor ability.

On extra-oral examination, he had a large head measuring 61 cm at the periphery, 24 cm anteroposteriorly and 26 cm in width with occipital bulge. The facial features were characteristic showing, large head with frontal bossing. Hypertelorism, with deep sunken slanting eyes were present. Also, redness of both the eyes accompanied by reduced visual acuity was evident, for which the patient is under treatment. Wide bridge of the nose with anteverted nares and angulated ears were present (**Fig: 1A and 1B**).

Intra-orally, retained 53, 55, 62, 63, 65, 75, 74, 73, 72, 82, 83, 84, 85 and root stump 64 were present. Missing 15, 25, 42, 44, 45, 34, 35; crowding of maxillary and mandibular anterior teeth, high arched palate and ankyloglossia, were also evident(**Fig: 2A and 2B**). Dental Panoramic radiograph revealed the evidence of multiple retained deciduous teeth with the corresponding impacted succidaneous teeth. A radiopaque tooth –like mass measuring roughly about 1 cm was seen in the right posterior mandible, apical to retained 84 and 85 which was suggestive of odontoma (**Fig: 4**).

CT scan of the brain revealed, the presence of prominent basal cisterns and extra axial fluid filled cystic lesion in the infra-tentorial region extending above the tentorium (**Fig:3**).

After obtaining physical fitness for the patient, the dental treatment was initiated. Extraction of the root stump in 64 region was carried out, followed by oral prophylaxis 2 weeks later. Surgical removal of the odontome has been planned.

III. discussion

Dandy Walker syndrome(DWS) is a rare clinical entity which is detected early in life. It is one of the group of disorders that affects the development of hind brain and posterior cranial fossa. It is inherited as an autosomal dominant triat with the prevalence of 1 in 25000 to 35000 (2). DWS predominantly occurs in female. But, our case is a male, which is a rare gender predilection (5).Literature suggests that 40% individuals with DWM had normal intelligence(2). Our case had normal intelligence which is in accordance with the literature. Predisposing factors for DWS include gestational exposure to rubella during first trimester, cytomegalovirus, toxoplasmosis, warfarin, alcohol and isotretinoin(6).

As Dandy–Walker complex has several variants, and encompasses cystic dilatation of the fourth ventricle, complete or partial agenesis of cerebella vermis , with or without enlarged posterior fossa or mega-cisterna magna with normal cerebellar vermis and fourth ventricle, the clinical manifestations also vary widely ranging from mild to severe disability in motor control, learning, and even cognition such as development of language and other cognitive skills(7) . Despite of the malformation in the posterior cranial fossa, the present case differed in a way that his postural, motor, language skills and intelligence remained unaffected.

Other clinical manifestations included, aortic abnormalities, skull, nose, eye, dental and digital anomalies including cleft lip, cleft palate, enamel hypoplasia and microdontia. Although , this case had certain clinical features like, skull, nasal and eye abnormalities,similar to that described in the literature, it had additional manifestations such as, ankyloglossia and odontome in the mandible which are rare and not yet reported to our knowledge.

IV. Conclusion

Early recognition of this syndrome is essential for a dental practitioner, as this rare clinical entity affects the oral hygiene status of an affected individual, in a detrimental manner owing to the poor motor co-ordination. Since this patient had no severe morbidity due the cranial malformation, and was able to do all his daily activities, he was advised to maintain his at home oral hygiene practice regularly.

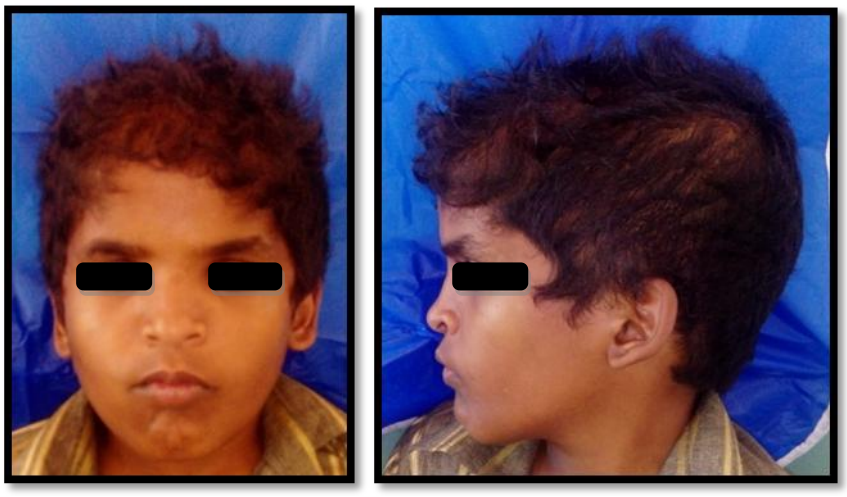

Figure: 1A **Figure: 1B**

Figure 1A: Extra oral photograph- frontal view showing macrocephaly,frontal bossing, depressed and wide nasal bridge **& Figure 1B-** left lateral view showing deep sunken orbits and low set ears.

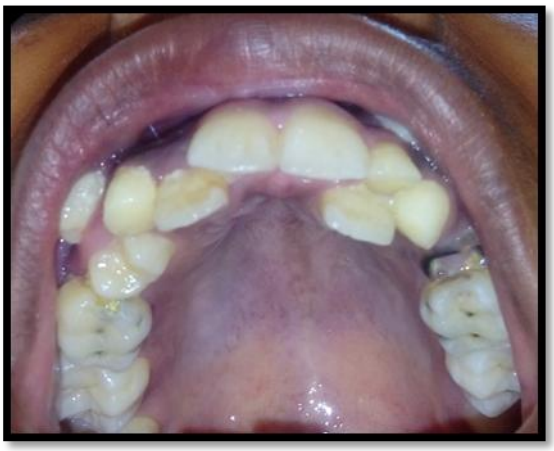

Figure 2 A: Intra oral phtographs showing high arched palate , retained deciduous and missing permanent teeth with anterior crowding.

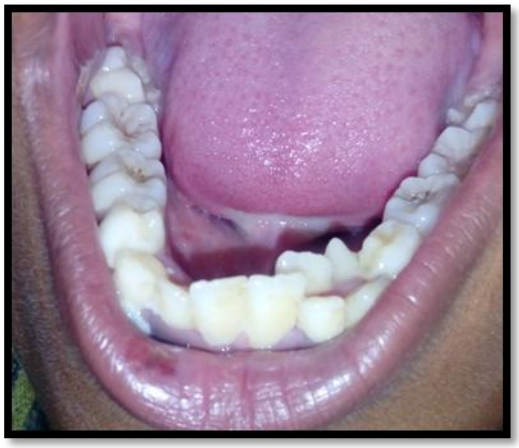

Figure 2 B: Intra oral phtograph showing ankyloglossia, multiple retained deciduous and missing permament teeth with anterior crowding.

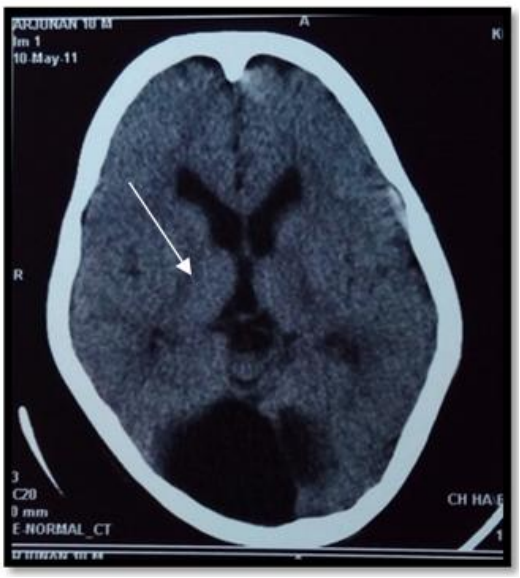

Figure 3 : Axial view of plain CT revealing cyst in the posterior cranial fossa (white arrow).

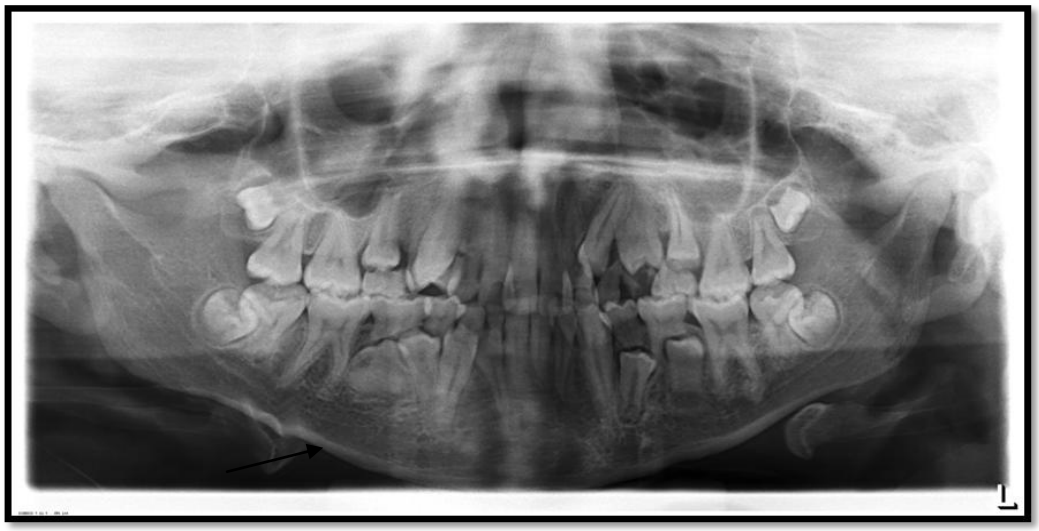

Figure 4: Orthopantomograph showing retained deciduous teeth and impacted permanent teeth with an odontome (black arrow) in relation to apices of 84 and 85.

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