ABSTRACT

Dandy–Walker syndrome (DWS) is a rare congenital cystic malformation of the posterior cranial fossa. Patients show signs and symptoms of complex clinical manifestations, ranging from cranial nerve and cerebellar dysfunctions to extracranial abnormalities, which may pose challenges in dental management. This article represents a rare case of a 12-year-old girl with DWS along with the involvement of the oral cavity.

KEYWORDS: Dandy–Walker syndrome, dental consideration, poor motor coordination

Introduction

Dandy–Walker malformation is a congenital abnormality of the posterior cranial fossa and the incidence of Dandy–Walker syndrome (DWS) is 1:25,000–1:35,000 live births.[1] The disorder was originally described in 1887 by Sutton. Later, it was explained by W. Dandy and K. Blackfan in 1914 followed by Tagart and Walker in 1942, and finally, C. Benda in 1954 designated this disorder as DWS.[2,3]

DWS comprises a wide range of findings from anatomically to clinically. Clinical manifestations are usually present at infancy which includes delayed motor development, nystagmus, apnea, and hydrocephalus. Hydrocephalus is often not seen at birth, but it is found in approximately 90% of the diagnosed case. DWS is also associated with other congenital anomalies which include hypertelorism, cardiac, renal, skeletal malformations, syndactyly, polydactyly limb, and vertebral abnormalities.[4]

Previous literature has shown that DWS is also associated with the high-arched palate, cleft lip/palate, retrognathia, malocclusion, and poor oral hygiene.[5] A DWS is a rare finding. This article presents a case of a DWS with grossly decayed posterior teeth which required extraction.

Case Report

A 12-year-old female reported in the Department of Pedodontics and Preventive Dentistry at UCMS and GTBH Hospital with the chief complaint of pain in the upper left tooth for past 6 months and decayed lower teeth. Mother gave the history of some viral infection in the 6th month of her pregnancy. The patient was born at the 34th gestational week with a birth weight of 2100 g. At 10 months of age, the patient presented with hydrocephalous and cystic dilatation of the right lateral ventricle; hence, the diagnosis of DWS was made. Mild nodular lesions present in both lateral and third ventricles [Figure 1]. DWS had caused impairment in

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vision and ataxic gait with left-sided imbalance. Her intellectual development was slightly delayed and was not attending any school. At present, she was not under any medications. After taking the parents’ consent, the clinical examination was performed.

On physical examination, the patient appeared to have ataxic gait and left-sided imbalance as she walked. The extraoral examination showed brachycephaly, frontal bossing, thick lower lip, and deviation of the chin to the right side [Figure 2]. The patient exhibited Frankel’s behavioral rating scale III. Patient’s mother gave the history of dental restoration which was done 2 years back and also mentioned pain sometimes in the lower jaw.

Intraoral examination showed dental age-appropriated dentition with the high-arched palate. Twenty six and 46 were grossly carious. Sixteen were having Class I Caries and 36 were already restored [Figure 3]. The behavior of the patient during the examination was cooperative; hence, restoration of 16 and extraction of 26 and 46 were planned under local anesthesia as the patient already had a history of dental treatment in the normal dental operatory. Routine blood investigations, electrocardiographic evaluations, and chest X-rays disclosed no abnormal findings. Pediatric and neurological consultation was done, and clearance was obtained regarding carrying out treatment in the routine dental operatory. Vital signs were regularly monitored. In the next appointment, glass ionomer cement (GIC) (GC Type 9) restoration of 16 was carried out, and atraumatic extraction of 26 and 46 was done in subsequent appointments. The patient showed good compliance throughout the procedures. Healing was satisfactory in both extractions with no postoperative complications after 1 week. The patient was instructed to follow-up at regular intervals; however, she had not shown up due to the unavailability of the parents’ schedule.

Discussion

DWS is a rare congenital abnormality of the posterior cranial fossa. It is an inherited autosomal trait and predominantly found in the female. The exact etiology of DWS is unclear but is believed that it is the combination of environmental and genetic factors. Environmental factors including prenatal exposure to alcohol, teratogenic agents, viral infection, and maternal diabetes.[6]

Recently, the genes FOXC1 on human chromosome 6p25 and linked ZIC1, ZIC4 on chromosome 3q24 have been identified which is found to be the causative agent of DWS.[7,8]

In our case, the patient had involvement of posterior permanent dentition along with recurrent swelling. DWS is associated with other craniofacial abnormalities such as the short neck, microcephaly, brachycephaly, hypertelorism, antimongoloid slant of palpebral fissures, globulus large nose, large mouth with downturned corners, poorly lobulated ears, high-arched palate, cleft palate, small hands and feet, and clinodactyly.[4]
Central nervous system (CNS) tumors are the most common solid tumors in the pediatric population with the highest incidence in children under the age of 5 years old. The location of the primary CNS tumors in children is commonly found in the posterior cranial fossa as opposed to the supratentorial tumors in cerebral hemispheres in adults. The development of the posterior fossa tumors has been found associated with genetic disorders and maternal exposure to teratogens. The clinical presentations of DWS vary depending on the severity of the affected organs. The increased intracranial pressure due to the tumor mass and hydrocephaly accounts for cerebellar and cranial nerve dysfunction, neurological complications, as well as intellectual impairment. Nearly half of the affected children with posterior fossa syndromes also presented with extracranial symptoms including cardiac anomalies, renal dysfunction, craniofacial malformations, and abnormal limb development.[9]

The syndromes associated with DWS include posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, and eye abnormalities and Ellis–van Creveld syndrome.[10,11]

The diagnosis varies depending on the degree of anomalies. Although nonspecific, the early signs of DWS including macrocephaly, enlargement of the skull in the occipital area and apnea, could be observed in the newborns. In 80% of cases, the diagnosis of DWS is made by the 1st year of life using computed tomography and/or magnetic resonance imaging. Undiagnosed DWS in the adult is usually detected by incidental findings during diagnostic imaging following head trauma, or in the presence of other neurological symptoms including headaches and seizures. The available treatment options for DWS are the traditional shunt placement, posterior fossa craniectomy, and endoscopic third ventriculostomy.[9]

The dental management of the patients with DWS should involve the review of medical history, consultation with physicians for associated extracranial abnormalities, craniofacial examination, oral examination, and behavioral assessment. Due to the complexity of the syndrome, affected individuals may present with distinct clinical presentations and varied intellectual development, which may be challenging to dental management. The review of the medical history of the current patient showed no contraindication for the dental treatment, and the patient was successfully treated in the dental chair.

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 poor motor coordination. Extensive research is required to identify or diagnose the disease at the earliest.

Previous reports showed the dental management of DWS patients under general anesthesia. Here, in our case report, it is the first time done on the dental chair under local anesthesia.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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