Keywords: Congenital cystic malformation; Cranial fossa; Extracranial abnormalities; Gingivectomy; Central incisor

Introduction

Dandy-Walker malformation (DWM) is a congenital abnormality of the posterior cranial fossa with reported incidence of 1:25,000 to 1:35,000 live births [1,2]. e classic clinical trials of the DWM are absence of cerebellar vermis, cystic dilatation of the fourth ventricle, and the enlargement of posterior cranial fossa [3]. In approximately 80% of diagnosed DWM cases, the syndrome is accompanied by hydrocephalus and developmental delay [4]. Other posterior fossa malformations such as Dandy-Walker variant, mega-cisterna magna, Blake's pouch, and arachnoid cyst, show overlapped clinical features with DWM and are believed to be parts of the continuum of the syndrome [5-7]. e term Dandy-Walker Complex has been suggested to describe the diseases as spectrums opposed to separate entities [8].

e pathogenesis of DWM results from the disturbance of the

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breathing sound were monitored every 5 minutes throughout the treatment time.

e behavior of the patient during the treatment was very co-operative. He was awake, easily arousable, and was able to followed instructions well. ere was minimal movement during the treatment. e vital signs were stable during and a er the treatment. e post-operative instructions were given to parents regarding concerns of anesthesia and sedation. e patient met the discharge criteria according to AAPD guideline [28]. e evening follow-up call suggested that patient was able resume his normal activities and tolerate regular diets. ere was absence of pain, nausea, vomiting, bleeding, swelling and any other discomfort. e patient was instructed to follow up 1 month, 3 months and 6 months a er the procedure; however, he had not shown up due to long travel time and the unavailability of parents' schedule.

Discussion

Dandy-Walker malformation (DWM) is a rare congenital abnormality of posterior cranial fossa. A ected individuals show a range of clinical manifestations depending on the extent intracranial and extracranial anomalies. Previous studies suggest the dental management of DWM patients under general anesthesia and IV sedation [26,27]. In the current report, we presented the dental management of an 11-year-old patient with DWM, who required coronal restoration of the previously endodontic-treated maxillary right central incisor, under oral conscious sedation. Clinically, patient presented with auditory and visual impairment, as well as cerebellum dysfunction as shown in ataxic gait. Review of patient's medical history showed the absence of extracranial manifestations. Assessment of his airway and his behavior, as well as the amount of required dental restoration, suggested that he was a candidate for dental treatment under oral sedation. e patient tolerated the procedure well and was able to cooperate during the treatment. He recovered and met the discharge criteria a er the procedure. is report suggested that patient with Dandy Walker Malformation can be safely managed in the dental chair despite the medical complexity of the syndrome.

CNS tumors are the most common solid tumors in pediatric population with the highest incidence in the children under the age of 5 years old [29,30]. e 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 [31]. e development of the posterior fossa tumors has been found associated with genetic disorders and maternal exposure to teratogens. e clinical presentations of DWM vary depending on the severity of the a ected organs. e 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 a ected children with posterior fossa syndromes also presented with extracranial symptoms including cardiac anomalies, renal dysfunction, craniofacial malformations, and abnormal limb development [32].

In the current report, there was negative family history of known genetic abnormalities. e patient presented with the clinical signs and symptoms similar to the previous case report [17] including visual and hearing impairment, ataxic gait, developmental delay, brachycephaly, hypertelorism and attened nasal bridge. e absence of other extracranial abnormalities may be related to his early resolution of the hydrocephalus and the tumor mass by 1 year of age. e dental ndings of Angle's Class II malocclusion with anterior single tooth crossbite and crowding are similar to the dental ndings in DWM reported previously [27]. e increased overjet may be associated with the increased susceptibility to trauma as seen in the current patient.

e dental management of the patients with DWM should involve review of medical history, consultation with physicians for associated extracranial abnormalities, craniofacial examination, oral examination and behavioral assessment. Due to the clinical presentations of DWM as well as potential delay intellectual development, the dental treatment planning should emphasize on preventive strategies. Because of a wide range of the clinical presentations of DWM, the restorative treatment options, if necessary, should be considered based on assessment of individual's clinical presentations and behavior.

In the current report, although the patient required restoration of only one tooth, the patient's attention span was not adequate to allow for dental treatment in the regular dental chair. A er reviewing of his medical history, the decision was made to treat him with under oral sedative medications. e triple cocktail was chosen as the oral sedative regimen based on the patient's behavior and the length of treatment. e use of ber-reinforced composite as a mono block post system in place of the conventional post system allowed for greater adaptation and exibility to the canal, the incorporation of the restorative material into the post system, as well as better time management under the oral sedative medications.

To our knowledge, this is the rst report of the dental management of a pediatric patient who presents with Dandy-Walker malformation, using oral conscious sedation. Due to the complexity of the syndrome, as well as the heterogeneity of the Dandy-Walker complex, a ected individuals may present with distinct clinical presentations and varied intellectual development, which may be challenging to the dental management. e review of medical history of the current patient shows no contraindication for the dental treatment under oral sedative medications and the patient was successfully treated in the dental chair. Future studies on clinical signs and symptoms of DWM patients in the same and di erent age groups who require dental treatment will contribute to better understanding of dental management and clinical decision making.

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