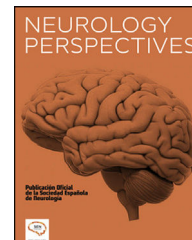




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## ORIGINAL ARTICLE

# Radiological examination of cranial, ear-nose-throat, and maxillofacial system findings of Dyke-Davidoff-Masson syndrome in 28 patients

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### KEYWORDS

Dyke–Davidoff–Masson syndrome;  
Ear–nose–throat;  
Maxillofacial surgery;  
Facial asymmetry;  
Frontal sinus

### Abstract

**Objectives:** It was aimed to detail the intracranial and ear–nose–throat–maxillofacial (ENT–MF) system findings of Dyke–Davidoff–Masson Syndrome (DDMS), and to examine the association in a larger sample according to current literature.

**Patients and methods:** The study was designed retrospectively. Cranial magnetic resonance (MRI) and computed tomography (CT) imaging records of patients were re-examined. Outcome parameters were identified as patients' demographics, the emergence of the central nervous system (CNS) pathologies, the asymmetric pneumatization of the skull bones, and ENT–MF system presentation. The correlation of the cranial and ENT–MF system findings was evaluated with Spearman correlation analysis.

**Results:** Radiological images of 28 patients were examined. The number of men was 21 (75.0%) and the number of females was 7 (25.0%). The mean age of the patients was  $23.6 \pm 10.6$  (min. 1, max. 44). DDMS involvement detected on the right side in 15 (53.6%) of the patients. Cerebral atrophy ( $n = 28$ , 100%), lateral ventricular dilatation ( $n = 26$ , 96.3%) and corpus callosum damage ( $n = 19$ , 67.9%) were most frequent findings respectively. Cerebellar atrophy ( $n = 4$ , 14.3) and venous sinus dominance ( $n = 4$ , 14.3) were the rarest pathologies. Facial asymmetry ( $n = 15$ , 53.6%) and frontal sinus hyperpneumatization ( $n = 15$ , 53.6%) were the most frequent findings in the ENT–MF system. Increased calvarial thickness showed a high positive correlation with facial asymmetry and frontal sinus hyperpneumatization ( $p = 0.002$ ,  $r = 0.571$  and  $0.024$ ,  $r = 0.424$ , respectively).

**Conclusion:** DDMS may occur with different radiological findings depending on the level of cerebral damage. Facial asymmetry often accompanies the disease and it should be considered in clinical practice of facial plastic and maxillofacial surgery.

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**PALABRAS CLAVE**

Síndrome de Dyke-Davidoff-Masson;  
Oído-Nariz-Garganta;  
Cirugía maxilofacial;  
Asimetría facial;  
Seno frontal

## Exploración radiológica de los hallazgos del sistema craneal, oído-nariz-garganta y maxilofacial del síndrome de Dyke-Davidoff-Masson en 28 pacientes

**Resumen**

**Objetivos:** Se tuvo como objetivo detallar los hallazgos del sistema intracraneal y del sistema oído-nariz-garganta-maxilofacial (ENT-MF) del síndrome de Dyke-Davidoff-Masson (DDMS) y examinar la asociación en una muestra más amplia de acuerdo con la literatura actual.

**Pacientes y métodos:** El estudio se diseñó de forma retrospectiva. Se volvieron a examinar los registros de imágenes de los pacientes por resonancia magnética craneal (IRM) y tomografía computarizada (TC). Los parámetros de resultado se identificaron como la demografía de los pacientes, la aparición de patologías del sistema nervioso central (SNC), la neumatización asimétrica de los huesos del cráneo y la presentación del sistema ENT-MF. La correlación de los hallazgos del sistema craneal y ENT-MF se evaluó con el análisis de correlación de Spearman.

**Resultados:** Se examinaron imágenes radiológicas de 28 pacientes. El número de hombres fue 21 (75,0%) y el número de mujeres fue 7 (25,0%). La edad media de los pacientes fue de  $23,6 \pm 10,6$  (mín. 1, máx. 44). La afectación del DDMS se detectó en el lado derecho en 15 (53,6%) de los pacientes. La atrofia cerebral ( $n = 28$ , 100%), la dilatación ventricular lateral ( $n = 26$ , 96,3%) y el daño del cuerpo calloso ( $n = 19$ , 67,9%) fueron los hallazgos más frecuentes, respectivamente. La atrofia cerebelosa ( $n = 4$ , 14,3) y la dominancia del seno venoso ( $n = 4$ , 14,3) fueron las patologías más raras. La asimetría facial ( $n = 15$ , 53,6%) y la hiperneumatización del seno frontal ( $n = 15$ , 53,6%) fueron los hallazgos más frecuentes en el sistema ORL-MF.

**Conclusión:** la EMDD puede presentarse con diferentes hallazgos radiológicos según el nivel de daño cerebral. La asimetría facial suele acompañar a la enfermedad y debe considerarse en la práctica clínica de la cirugía plástica facial y maxilofacial.

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**Introduction**

Dyke–Davidoff–Masson syndrome (DDMS), also known as cerebral hemiatrophy, is a rare condition characterized by convulsions, facial asymmetry, contralateral hemiparesis/hemiplegia, and mental retardation. Due to the loss of cerebral volume, thickening of calvarial and diploe on the same side, and hyperaeration in the paranasal sinuses and mastoids occur.<sup>1</sup> Cerebral hemiatrophy can be partial or widespread.<sup>2</sup> Depending on the extent of involvement, clinical symptoms may occur such as epilepsy, facial asymmetry, hemiplegia/hemiparesis, speech and language disorders, learning difficulties, and mental retardation. Therefore, the beginning and the process of the disease can be different. Diagnosis is based on clinical findings and radiological imaging.<sup>2</sup>

Magnetic resonance imaging (MRI) and computed tomography (CT) are frequently used in diagnostic imaging. Unilateral loss of cerebral parenchyma, thickening of the calvarial bones on the same side, ventriculomegaly, hyperpneumatization in the paranasal sinus and mastoids, basal ganglia, or brainstem atrophy can be found on radiological examination.<sup>3</sup>

The syndrome was first described by Dyke, Davidoff, and Masson in 1933.<sup>4</sup> The most important findings are unilateral cerebral atrophy, contralateral hemiparesis, and epilepsy. Psychiatric or behavioral disorders such as schizophrenia can be seen in some cases.<sup>5</sup> The extension and severity of the symptoms vary according to the affected brain development. Prenatal or perinatal brain injury may have a more severe clinical condition than at early age's trauma.<sup>3–5</sup>

Etiology of the disease depends on an injury during the development of the central nervous system. Causes in the prenatal period are congenital malformation, infection, and vascular insufficiencies, birth trauma, anoxia, hypoxia and intracranial hemorrhage. Postnatal causes are trauma, tumor, infection, and prolonged febrile seizures. Congenital type DDMS shows an enlargement of the calvarium, diploic space, and paranasal sinuses, in contrast to acquired DDMS. These compensatory cranial changes occur relative to the atrophied cerebral hemisphere.<sup>3,4,6</sup>

The literature of DDMS is generally based on single case reports or series with a limited number of cases. In this study, it was aimed to detail the cranial and ear–nose–throat-maxillofacial (ENT-MF) system findings in a larger sample.

**Material and method**

The study was designed retrospectively. The cranial magnetic resonance (MRI) and computed tomography (CT) imaging records of patients were re-examined between January 2010 and January 2020. The study conducted at a tertiary referential university hospital. The definitive diagnosis of the DDMS was made with a correlation of clinical history and radiological findings of the patients. Outcome parameters were identified as patients' demographics, the emergence of the central nervous system (CNS) pathologies, the asymmetric pneumatization of the skull bones, and ENT-MF system coalescence. The presence of the findings was

examined on the side where DDMS was located. Incidental counterparty findings were not taken into analysis.

### Radiological examination

All cranial or maxillofacial CT images were obtained as standard 0.5 mm thick high-resolution sections (Toshiba; Activion, 64 Multislice CT, Japan). Also, cranial and maxillofacial findings were evaluated with T1, T2 sequences and diffusion MRI (Siemens; Magnetom, 3T MR, Germany).

### Inclusion and exclusion criteria

Patients with clinical and radiological diagnosis of the DDMS included in the study. Craniofacial malformation (etc. Craniosynostosis), congenital malformations, and any sign of cranial-maxillofacial trauma or surgery were examined and excluded.

### Statistical analysis

Findings were expressed as number and frequency. The correlation of cranial and maxillofacial findings was evaluated with Spearman correlation analysis. SPSS 22.0 (Statistical Package for Social Sciences, SPSS Inc., Armonk, NY) program was used for statistical analysis.

### Ethics committee approval

The study was conducted in accordance with the 1964 Helsinki Declaration and subsequent amendments. The institutional review board of Dicle University has approved the study protocol.

### Results

Radiological images of 28 patients were examined. The number of men was 21 (75.0%) and the number of females was 7 (25.0%). The mean age of the patients was  $23.6 \pm 10.6$  (min. 1, max. 44). DDMS involvement detected on the right side in 15 (53.6%) of the patients. Cerebral atrophy ( $n = 28$ , 100%), lateral ventricular dilatation ( $n = 26$ , 96.3%), and corpus callosum damage ( $n = 19$ , 67.9%) were most frequent findings respectively. Cerebellar atrophy ( $n = 4$ , 14.3) and venous sinus dominance ( $n = 4$ , 14.3) were the rarest pathologies. A summary of the cranial and CNS findings is given in Table 1. (See Figs. 1–5.)

Facial asymmetry ( $n = 15$ , 53.6%) and frontal sinus hyperpneumatization ( $n = 15$ , 53.6%) were the most frequent findings in the ENT system and MF region. Cochlear bone asymmetry ( $n = 1$ , 3.6%) and lacrimal system asymmetry ( $n = 1$ , 3.6%) were most rarely detected. Summary of the findings in the ENT-MF system is given in Table 2.

Increased calvarial thickness showed a high positive correlation with facial asymmetry and frontal sinus hyperpneumatization ( $p = 0.002$ ,  $r = 0.571$  and  $0.024$ ,  $r = 0.424$ , respectively). The correlation statistics between cranial and ENT-MF findings are given in Table 3.

**Table 1** Summary of the cranial and CNS findings.

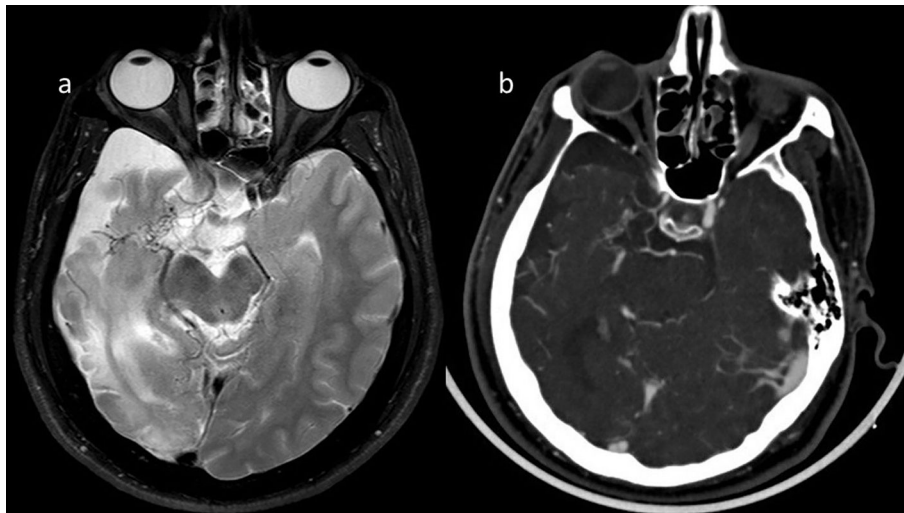
		n	%
Gender	Male	21	75.0
	Female	7	25.0
Involvement	Right	15	53.6
	Left	13	46.4
Cerebral atrophy		28	100.0
Increased Calvarial Thickness		17	60.7
Wallerian degeneration		18	64.3
Skull hyperpneumatization		15	53.6
Basal ganglia atrophy		15	53.6
Brainstem atrophy		7	25.0
Encephalomalacia		10	35.7
Corpus callosum damage		19	67.9
Cerebellar atrophy		4	14.3
Lateral ventricular dilatation		26	96.3

### Discussion

CT and MRI have an important role in the diagnosis and follow-up of DDMS. Radiologically distinctive findings are diffuse-focal cerebral hemiatrophy, encephalomalacia, or cerebral hypoplasia due to gliotic changes. In addition, ipsilateral sulcal enlargement, ventriculomegaly, compensatory calvarial hypertrophy, hyperaeration of paranasal sinuses, and hypoplasia in the thalamus, or lentiform nucleus, or caudate nucleus, or mesencephalon can be seen.<sup>3,5,6</sup> The most familiar manifestations of ENT-MF system are facial asymmetry, mastoid bone and frontal sinus hyperpneumatization.<sup>1,3</sup>

The number of studies involving large patient populations with DDMS is limited. Two large series in the literature, the mean age was detected as 6.8 years ( $n = 19$ ), and 11 years ( $n = 26$ ).<sup>3,7</sup> The mean age of the patients was found as  $23.6 \pm 10.6$  years (min. 1, max. 44) in this study. DDMS usually diagnosed in childhood. This finding, which differs from the literature, may be due to the fact that the patients included in this study were diagnosed later due to the low socioeconomic level of the study population. Solomon et al.<sup>8</sup> reported that imaging findings compatible with DDMS were evident only in children who had a brain injury before the age of 3 years. Therefore, cerebrovascular pathologies occurring during pregnancy up to 3 years of age will most likely have a tendency to result in DDMS. In this period, long-term ischemia may cause brain atrophy by reducing the production of neurotrophic factors derived from the brain.<sup>9</sup> So, related structures such as the calvarium and sinuses tend to grow compensatory. These results indicate that the diagnosis of patients in this study was delayed, or these patients may have been followed up with other diagnosis without imaging techniques with existing clinical findings.

Functionally, men have more hemispheric asymmetry than women and may cause differences in neuronal connectivity.<sup>10</sup> According to one hypothesis, the presence of circulating androgens can create a hyperplastic condition in the brain of a developing man, leading to wider neuronal remodeling after injury than the female brain.<sup>11</sup> In our study, there were 21 (75%) male patients, 3 times the number of women. Unal et al.<sup>7</sup> reported higher rates of DDMS in male patients with a rate of 73.5%. In other large



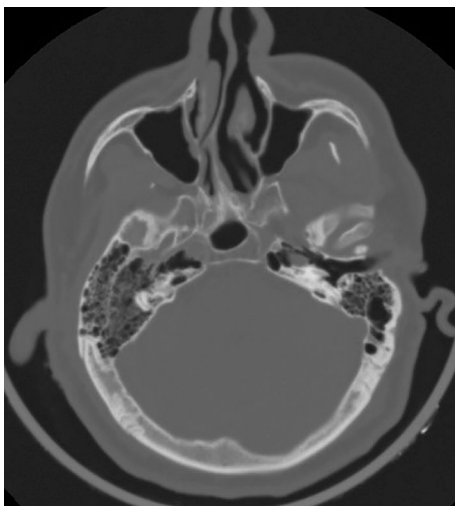
**Fig. 1** A 26-year-old male patient, vascular collateral structures (a) and parenchymal volume loss are present at the right MCA level on MRI (T2 weighted). CT angiography (b) indicates decreased right ICA calibration and vascular collateral structures at the MCA level. The findings are consistent with Moyamoya disease.

series, the proportion of male patients was found to be higher than women.<sup>3,12</sup> In this study, female-to-male distribution was compatible with the literature.

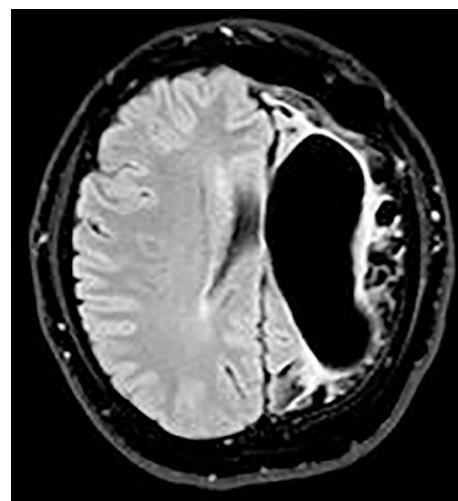
Cerebral hemiatrophy is one of the main components of DDMS. It has been reported that it results from intrauterine conditions in which calvarium maturation is not yet complete or brain damage in the first 3 years of life.<sup>13</sup> Although cerebral hemiatrophy completely involves the single hemisphere, it can be seen with encephalomalacia that causes more atrophy in certain lobes. It can occur due to congenital and acquired causes. Unal et al.<sup>7</sup> found left hemiatrophy in 69.2% of patients and left hemiatrophy was detected in 57.1% of patients by Ayas et al.<sup>4</sup> Some studies have suggested that the left hemisphere is more susceptible to cortical damage.<sup>14</sup> In the present study, 53.6% (n = 15) of cerebral hemiatrophy was observed on the right side.

Contrary to the literature, laterality was observed in favor of the right in DDMS in our study. The side of cerebral hemiatrophy is likely to be related to the side of the etiological cause. Moyamoya disease was found in two cases with cerebral hemiatrophy and polymicrogyria in one case.

Polymicrogyria is rarely seen in these patients and has been reported in two case reports including the pediatric population.<sup>15,16</sup> Increased calvarial thickness occurs as a result of compensatory hypertrophy of the affected cerebral hemisphere. Calvarial changes may or may not be present depending on the time of injury. Increased calvarial thickness has been reported in the literature between 57% and 100% in different studies.<sup>1,3,4,7,10,17</sup> Increased calvarial thickness was observed in 60.7% of cases in this study. The findings are consistent with the literature, but varying rates are likely due to sampling size.

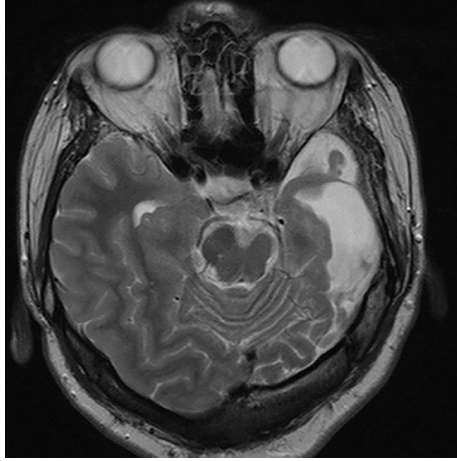


**Fig. 2** Right cerebral hemiatrophy, increased calvarial thickness on the right, and hyperaeration of the mastoid bone in a 28-year-old male patient (CT).



**Fig. 3** A 20-year-old female patient with left cerebral hemiatrophy and dilatation in compensatory lateral ventricles. Also, extensive encephalomalacia areas in the parenchyma and hyperaeration in the frontal sinuses (MRI Flair).





**Fig. 4** A 17-year-old female patient with cerebral hemiatrophy and parenchymal volume loss in the left half of the brainstem compared to the right (T2 weighted MRI).

Similarly, hyperpneumatization in the paranasal sinuses develops compensatory in the affected cerebral hemisphere. Paranasal sinus hyperpneumatization (HP) has been reported between 50% and 91% in studies with large samples in the literature.<sup>1,7,17</sup> In this study, HP was found in the frontal sinus in 53.5% of the cases, in the sphenoid sinus in 35.7%, and in the ethmoid sinus in 21.4%. The most important reasons for the difference in rates are the number of cases and the possible average age of the study population because it takes years for the paranasal sinuses to reach mature sizes. In addition, all patients with paranasal sinus HP had frontal sinus HP in this study. Also, 35.7% HP was observed in the mastoid bone in this study. Gökçe et al.<sup>1</sup> 75%, Taşdemir et al.<sup>12</sup> reported 100% mastoid HP. Reported rates of mastoid HP in the referenced studies is clearly



**Fig. 5** A 22-year-old patient with left cerebral hemiatrophy, ipsilateral atrophy of the basal ganglia (MRI Flair).

**Table 2** Summary of the findings in the ENT system and maxillofacial region.

	n	%
Facial asymmetry	15	53.6
Mastoid hyperpneumatization	10	35.7
Frontal sinus hyperpneumatization	15	53.6
Sphenoid sinus hyperpneumatization	10	35.7
Ethmoid sinus hyperpneumatization	6	21.4
Middle concha bullosa	4	14.3
Cochlear bone asymmetry	1	3.6
Lacrimal system asymmetry	1	3.6

different, so, mastoid HP may be caused by other causes such as eustachian canal dysfunction.

Ayas et al.<sup>4</sup> detected facial asymmetry in 6 of 7 (85.7%) patients but Unal et al.<sup>7</sup> found facial asymmetry in 8 of 26 (30.7%) patients with DDMS. In this study, facial asymmetry was there in 50.3% (n = 15) of patients. In this study, facial asymmetry was only significantly correlated with increased calvarial thickness. This finding may be important in facial plastic and maxillofacial surgery and in the differential diagnosis of a patient with ENT symptoms.

Unilateral fetal ventriculomegaly may develop DDMS in following years. It should be followed up with radiological imaging. Compensatory ventriculomegaly is observed in the part with cerebral hemiatrophy. In this study, dilatation was observed in the lateral ventricles in 96.4% of the cases. Gökçe et al.<sup>1</sup> detected lateral ventricular enlargement in all 12 patients. In these patients, ventriculomegaly occurs as a result of the depletion of ipsilateral brain tissue. The severity of ventriculomegaly is inversely proportional to the brain tissue. Porencephalic cysts can occur with severe reduction of brain tissue. In our study, encephalomalacia areas were observed in 35.7% of the cases. In the same study encephalomalacia was detected with a rate of 91.6%.<sup>1</sup> Encephalomalacia may develop focally or widespread. The corpus callosum may be affected by diffuse involvement. In this study, the damage was observed in the corpus callosum in 67.8% of the cases. Gökçe et al.<sup>1</sup> also found the corpus callosum damage in 75% of cases. The findings observed in this study are similar to the reference study.

In addition, 14.3% of our patients had cerebellar atrophy and one patient had crossed-cerebellar atrophy. Crossed-cerebellar atrophy is relatively less common and includes volume loss and signal changes in the contralateral cerebellar hemisphere. It occurs due to the interruption of cerebro-cerebellar pathways due to transneuronal cerebellar metabolic depression. Cerebellar atrophy can be focal or common. Atalar et al.<sup>3</sup> detected diffuse cerebellar atrophy in a patient with encephalomalacia in the left middle and posterior cerebral artery watershed areas and reported that it may develop as a result of insufficient recirculation. Demir et al.<sup>18</sup> reported left cerebellar atrophy in a patient with right cerebral hemiatrophy. Tasdemir et al.<sup>12</sup> observed cerebellar atrophy in 1 of 5 cases on the same side. In this study, basal ganglia atrophy was observed in 53.5% of patients, and brainstem atrophy was found in 25% of cases. Shubhakaran et al.<sup>19</sup> observed atrophy in the thalamic and lentiform nucleus in 39.2% of 28 patients. Ayas et al.<sup>1</sup> found 71.4% of atrophy in the basal ganglia and 66% in the brainstem.

**Table 3** The correlation table between cranial and ENT-MF system findings.

		Facial asymmetry	Frontal sinus HP	Mastoid HP	Sphenoid sinus HP
Lateral ventricular dilatation	r	0.204	−0.189	−0.256	−0.277
	p	0.309	0.345	0.198	0.161
Corpus callosum damage	r	−0.027	−0.181	0.034	−0.285
	p	0.890	0.357	0.863	0.142
Wallerian degeneration	r	0.203	0.053	−0.067	−0.067
	p	0.301	0.787	0.736	0.736
Basal ganglia atrophy	r	0.138	0.138	0.246	−0.053
	p	0.482	0.482	0.208	0.787
Increased calvarial thickness	r	0.571	0.424	0.294	0.294
	p	<b>0.002</b>	<b>0.024</b>	0.128	0.128
Skull hyperpneumatization	r	0.282	1.000	0.694	0.694
	p	0.146	<b>&lt;0.001</b>	<b>&lt;0.001</b>	<b>&lt;0.001</b>
Encephalomalacia	r	0.096	0.246	0.222	0.067
	p	0.627	0.208	0.256	0.736
Brainstem atrophy	r	0.041	−0.124	−0.086	−0.258
	p	0.835	0.529	0.663	0.185
Venous sinus dominance	r	−0.234	0.380	0.335	0.335
	p	0.231	<b>0.046</b>	0.082	0.082
Cerebellar atrophy	r	0.175	0.175	−0.091	0.335
	p	0.372	0.372	0.644	0.082

HP: hyperpneumatization. Significant p values are shown in bold.

Gökçe et al.<sup>1</sup> found atrophy in the basal ganglia at a rate of 66%. Wallerian degeneration was observed with a rate of 64.2% in this study. This entity has not been mentioned in the referenced studies with large samples. Therefore, it is not possible to decide on a generalizable ratio. In addition, venous sinus dominance was found at a rate of 14.3% in this study, and this finding was also not mentioned in the literature.

The differential diagnosis of DDMS includes Sturge–Weber syndrome, Rasmussen encephalitis, Silver–Russell syndrome, basal ganglia germinoma, Fishman syndrome (encephalocraniocutaneous lipomatosis), and linear nevus sebaceous syndrome. Cerebral hemispheric atrophy is observed in Rasmussen encephalitis but no calvarial changes are expected.<sup>20</sup> In Sturge–Weber syndrome, leptomeningeal angiomas lead to stasis, which causes ischemia and then laminar cortical necrosis and atrophy. Additionally, Tram-track signs in the brain due to calcification are a specific radiological finding for Sturge–Weber syndrome.<sup>21</sup> Silver–Russell syndrome has a classical facial phenotype (triangular face, small pointed chin, wide forehead, and thin wide mouth) and this syndrome is characterized by delayed bone age, growth retardation, clinodactyly, normal head circumference, hemihypertrophy, and normal intelligence.<sup>22</sup> These differential diagnoses of DDMS distinguished easily by performing a comprehensive clinical examination and neuroimaging.<sup>23,24</sup>

The main limitations of the study are its retrospective nature and independence from the patient's history and clinical information. The sample size of the study is superior to the reports in the literature. On the other hand, while reference studies usually include information near the time of diagnosis and usually in the pediatric population, this study analyzed the most recent radiological findings, including children and adults. In addition, although the relationship between cranial findings and ENT-MF system

findings was examined in this study, unlike the literature, no clinically significant evidence was found. But the sample size is very small for the cross-tables or correlation tests.

## Conclusion

DDMS is a rare syndrome diagnosed with clinical and radiological findings. It may occur with different radiological findings depending on the level of cerebral damage. DDMS tends to involve in the right cerebral hemisphere with compensatory calvarial thickening, causing HP in the paranasal sinuses, and more common in males. In addition, facial asymmetry is important in terms of diagnosis and treatment for maxillofacial surgery.

## Ethics approval

The institutional review board of Dicle University has approved the study protocol.

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## Declaration of competing interest

None.

## Acknowledgments

None.

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