



A Retrospective Evaluation of Children Diagnosed with Dermatomyositis: A Single-center Study

Dermatomiyozit Tanılı Çocuk Hastaların Retrospektif Değerlendirmesi: Tek Merkezli Bir Çalışma

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ABSTRACT

Objective: Juvenile idiopathic inflammatory myopathies are systemic autoimmune disorders that are characterized by chronic skeletal muscle inflammation, skin rashes and other systemic involvements. We analyzed the clinical findings, laboratory values at admission and treatment protocols and treatment responses of patients who were followed up with a diagnosis of juvenile dermatomyositis (JDM) in a department of pediatric neurology and rheumatology clinics.

Methods: Fifteen patients who were referred to the department of pediatric neurology and pediatric rheumatology clinics, diagnosed with JDM between 2010 and 2017 were evaluated retrospectively via their medical records.

Results: Of the study sample, 12 (80%) of the patients were female and 3 (20%) were male, and their mean age was 9.26 ± 3.21 years. The mean time between complaint and diagnosis was 7.8 ± 6 months, and the patients were followed up for 24.93 ± 15.28 months after their diagnosis. The mean creatine kinase levels of the patients were 1.354 ± 840 U/L. Fifteen (100%) of the patients had muscle weakness, 14 (93.3%) had Gottron's papules and 12 (80%) patients had a heliotrope rash. Ten (66.6%) underwent muscle biopsy, 9 (60%) underwent electromyography and 5 (33.3%) patients underwent muscle magnetic resonance imaging. All the patients were treated with corticosteroids and immunosuppressive agents.

Conclusion: JDM is a rare inflammatory myopathy observed during childhood. Better responses can be achieved by early diagnosis, intensive immunosuppressive therapy and effective physical therapy.

Keywords: Juvenile dermatomyositis, steroid, creatine kinase, muscle biopsy

ÖZ

Amaç: Juvenil dermatomiyoisit deri bulguları, kas tutulumu ve diğer sistemik tutulum ile seyreden enflamatuvar otoimmün bir hastalıktır. Bu çalışmada çocuk nörolojisi ve romatoloji kliniklerince juvenil dermatomiyoisit (JDM) tanısı ile izlenen hastaların klinik bulguları, laboratuvar değerleri, başvuru ve tedavi protokolleri ve tedavi yanıtlarını analiz etmeyi amaçladık.

Gereç ve Yöntem: 2010-2017 yılları arasında çocuk nöroloji ve çocuk romatoloji kliniğine sevk edilen ve JDM tanısı alan 15 hasta tıbbi kayıtlarla geriye dönük olarak değerlendirildi.

Bulgular: Çalışma örnekleminin 12'si (80%) kadın, 3'ü (%20) erkekti ve ortalama yaş $9,26 \pm 3,21$ yıldır. Şikayet ile tanı arasındaki ortalama süre $7,8 \pm 6$ aydı ve hastalar tanıdan sonra $24,93 \pm 15,28$ ay takip edildi. Hastaların ortalama kreatinin kinaz seviyeleri $1,354 \pm 840$ U/L idi. Hastaların tamamında kas güçsüzlüğü, 14'ünde (%93,3) Gottron papülleri ve 12'sinde (%80) heliotrope döküntüleri vardı. On (%66,6) hastaya kas biyopsisi, 9 (%60) hastaya elektromiyografi ve 5 (%33,3) hastaya kas manyetik rezonans görüntüleme uygulandı. Tüm hastalar kortikosteroidler ve immünoşüpresif ajanlarla tedavi edildi.

Sonuç: JDM, çocukluk çağında görülen nadir bir enflamatuvar miyopatidir. Erken teşhis, yoğun immünoşüpresif tedavi ve etkili fizik tedavi ile daha iyi yanıtlar elde edilebilir.

Anahtar Kelimeler: Juvenil dermatomiyoisit, steroid, kreatin kinaz, kas biyopsisi

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INTRODUCTION

Juvenile idiopathic inflammatory myopathies are systemic autoimmune disorders that are characterized by chronic skeletal muscle inflammation, skin rashes and other systemic involvement. The most common form of these disorders observed during childhood is juvenile dermatomyositis (JDM), while polymyositis is relatively more common in adults (1). The etiology of JDM is still unclear, although it has been associated with environmental triggers, immune dysfunction and specific tissue response in the muscles, skin and small vessel endothelium in genetically susceptible individuals (1,2). No remarkable familial transmission has been demonstrated. The mean age of onset is 7-9 years, and it is twice as common in girls as in boys. The incidence of JDM is estimated to be between 0.19 and 4.1 per million (3).

The disease is characterized by proximal muscle weakness and cutaneous findings such as heliotrope rash, Gottron's papules and calcinosis, and it can also affect such internal organs as the lungs and heart. The disease is still diagnosed and classified according to the criteria established by Bohan and Peter (4) in 1975. These criteria include symmetrical proximal muscle weakness and characteristic rashes, as well as the demonstration of elevated serum muscle enzyme levels, electromyographical changes and inflammatory myositis in muscle biopsy. In addition to the characteristic skin rashes, at least two of these findings should be present for diagnosis, although the pediatric suitability of these criteria is limited, as some pediatric patients may have normal electromyography (EMG) and muscle biopsy findings, despite the presence of typical rashes (5). These are referred to as amyopathic dermatomyositis cases (1). Amyopathic dermatomyositis manifests only with skin findings, with patients presenting no other finding of muscle involvement. However, a detailed investigation of muscle strength and muscle biopsy often show muscular involvement (4).

Corticosteroids are the gold standard for treating JDM, reducing both acute and long-term morbidity. Most patients respond well to therapy, although the presence of calcinosis, lipodystrophy and contractures, as treatment-resistant factors, can lead to poor prognostic (1,6). New advanced therapeutic options, such as biological agents and autologous stem cell transplantations, have recently been shown to be useful, particularly in refractory patients with poor prognosis (1).

In this study, we evaluated the demographic characteristics, clinical findings, laboratory values at admission, treatment

protocols and treatment responses of those patients who were followed up with a diagnosis of JDM in a department of pediatric neurology and pediatric rheumatology clinics.

METHODS

The medical records from the JDM registry of Inonu University Hospital in Turkey were retrospectively reviewed for the period between January 2010 and December 2017. A total of 20 patients were included in this study, but five patients were excluded due to missing data in their medical records. Therefore, 15 patients who were diagnosed with JDM and were followed up in pediatric neurology and pediatric rheumatology clinics, as the regional reference center in the eastern side of Turkey, between 2010 and 2017, were evaluated retrospectively. Their demographic characteristics, clinical findings, laboratory values at admission, treatment protocols and treatment responses were examined, and the findings of a detailed physical and neurological examination were reviewed. The creatine kinase (CK), aspartate transaminase (AST), alanine transaminase, lactate dehydrogenase, platelet (PLT), white blood cell, hemoglobin (HBG), C-reactive protein (CRP) levels and erythrocyte sedimentation rates (ESR) measured at the time of diagnosis were evaluated. The results of an antinuclear antibody (ANA) test, double-stranded DNA (dsDNA) tests, muscle biopsy, EMG and echocardiography performed for diagnostic and differential diagnostic purposes were reviewed. The treatments used during the acute phase of the disease and during maintenance treatments, as well as the responses of the patients to the treatment, were also evaluated. This study was approved by the institutional review board.

Statistical Analysis

All statistical analyses were performed using the SPSS statistics 22 software. All quantitative data are expressed as mean \pm standard deviation. All categorical variables are expressed as number and percentage (n, %).

Approval this study was obtained by the Inonu University Institutional Ethics Committee (decision no: 2018/9-7, date: 24.04.2018). Informed consent was obtained from the parents of all the patients. The study was conducted in accordance with the Declaration of Helsinki.

RESULTS

In total, 12 (80%) patients were female and three (20%) were male. The age range was 3 to 14 years and the mean age was 9.3 ± 3.2 years. The time between the onset of symptoms and diagnosis ranged from 1 to 24 months, with a mean

value of 7.8 ± 6 months. Patients were followed up for 10-60 months after being diagnosed, and the mean duration of follow-up was 24.9 ± 15.3 months (Tables 1, 2).

Among the most common findings of JDM are those related to the skin. In our study, Gottron's papules (Figure 1) and heliotrope rashes (Figure 2) were present in 14 (93.3%) and 12 (80%) patients, respectively, while three patients (20%) were seen to have alopecia, and hand calcinosis and hand ulcers were observed in three patients (20%) and a single patient (6.6%), respectively, in their follow-up process, although these were not present at the time of diagnosis. The musculoskeletal system is often affected by JDM, and in this study, six patients (40%) had contractures of various joints, seven patients (46.6%) had arthritis/arthritis, three patients (20%) had osteoporosis and a single patient (6.6%) had scoliosis. Furthermore, one (6.6%) patient had type 1 diabetes mellitus, two patients (13.3%) had repetitive pneumonia, a single patient (6.6%) had tuberculosis and a single patient (6.6%) died of sepsis. Gastrointestinal involvement is also common in patients with JDM. In this study, five patients (33.3%) were observed to have a gastrointestinal involvement, and of those, three had



Figure 1. 7th patient's Gottron's papules



Figure 2. 7th patient's Heliotrope rashes

dysphagia, one had hepatomegaly and one had gastritis. Raynaud phenomenon was also noted in three (20%) patients. No ocular complications developed in any patient (Table 3).

Patients with JDM almost always present with neurological involvements (5). The neurological examination findings of the patients at the time of diagnosis were reviewed. All the patients had varying degrees of muscular strength loss, particularly in the proximal muscles, eight (53.3%) had Gowers' sign and deep tendon reflexes were decreased in seven (46.6%) patients. During the follow-up period, a generalized tonic-clonic seizure was observed in a single patient (6.6%) and antiepileptic therapy was initiated, whereas two patients (13.3%) developed neuropathies. Chronic findings such as scapular winging and lumbar lordosis developed particularly in those patients with a protracted period between symptom onset and diagnosis (patients number 1, 4 and 13), and one patient (patient number 10) had no muscular involvement and was being followed up with a diagnosis of amyopathic JDM (Table 3).

Laboratory measurements obtained at the time of diagnosis are reviewed (Table 1). At the time of diagnosis, only 12 (80%) patients had an ESR higher than 20 mm and 11 (73.4%) patients had positive CRP measurements. Antinuclear antibodies were positive in eight (53.3%) patients; six (40%) patients were ANA positive, and two (13.3%) patients were positive for both ANA and anti-dsDNA (Tables 1, 2).

Of the 15 patients, 10 (66.6%) underwent muscle biopsy. Consistent with JDM, all biopsy procedures revealed mild diameter differences in muscles, atrophy in some perifascicular fibers and degenerated fibers with basophilic staining. Other tests were performed on patients who could not undergo such invasive procedures, such as muscle biopsies, to support their diagnosis. A standard magnetic resonance imaging (MRI) of the gluteal or gastrocnemius muscle was performed in five (33.3%) of the 15 patients. All patients had T2 and a flair intensity increase in the present muscle structures, consistent with the JDM diagnosis. Varying degrees of myofibril irritation (fibrillation potentials, repetitive complex discharges and sharp positive waves) were detected in eight of the nine patients who underwent EMG, which was interpreted in favor of myositis. An echocardiography was performed on eight (53.3%) patients, and no pathology was observed (Table 2).

During the acute phase, patients were administered pulse steroids and maintenance steroid therapy. Of all patients receiving steroid therapy, 12 (80%) responded during the acute phase, while three (20%) were nonresponsive. Following the acute phase, no recurrence was observed in

three (20%) of the 15 patients, and they were considered cured. Well-implemented physical therapy facilitates and supports the treatment process in JDM (5). All patients received physical therapy in this study, and of the total, six (40%) were therapy-resistant, and nine (60%) were treatment-resistant and experienced intermittent episodes of muscle weakness. These treatment-resistant patients were started on methotrexate following the steroid therapy. Patients who experienced recurrences despite methotrexate treatment were given other immunosuppressive therapies (cyclophosphamide, IVIG, hydroxychloroquine) (Table 2). Mortality is a rare finding of JDM. In this study, frequent pulmonary infections, ulcers and dysphagia developed in patient number 8, and in the 18th month of follow-up, this patient died from sepsis and intracranial bleeding (Table 3).

DISCUSSION

Although the incidence of JDM is low during the childhood period, it is still the most common inflammatory myopathy seen in children (5). The first clinical signs of JDM include skin and muscle involvement, while heart, lung, and gastrointestinal involvement are rare (7). The diagnostic criteria suggested by Bohan and Peter (4) in 1975 are still the standard means of diagnosis of JDM, although recently developed autoantibodies, biological agents and imaging

methods can also contribute, and calcinosis and findings of myositis in MRI have also emerged as diagnostic criteria (8,9).

JDM is more common among girls, with previous studies in the literature reporting F/M ratios of 2-3:1 in Europe and the United States (10,11). In the study by Barut et al. (12), the F/M ratio in the western regions of Turkey was reported to be 2.3:1, while this ratio was 4:1 in this study. This observed difference may be attributed to the small number of cases evaluated in this study, though regional or genetic factors may also be effective. The mean age of the patients included in this study was 9.2 years, the mean duration between the complaint and diagnosis was 7.8 months and the mean follow-up duration was 24.9 months. In the literature, the mean age of diagnosis was reported to be 7.5 years in the study by Malek et al. (13), 7.1 years in the study by Gowdie et al. (9) and 6.75 years in the study by Okong'o et al. (14). The mean time between symptom onset and diagnosis was reported to be between 4 and 6 months (12,13). The higher mean age of diagnosis in this study indicates a delay in the diagnosis. This was likely due to the location of our clinic in the eastern part of Turkey. The number of specialized physicians like pediatric neurologists and rheumatologists, is limited in this region. Patients with widespread systemic findings may be followed up by outpatient clinics having

Table 1. Patients' results of demographic findings, laboratory findings

Patient	n (%)
Gender	
Female	12 (80)
Male	3 (20)
Age (average \pm SD)	3-14 years (9.26 \pm 3.21 years)
Time interval between complaint and diagnosis (months)	1-24 month (7.8 \pm 6 months)
Length of follow-up (months)	10-60 month (24.93 \pm 15.28 month)
Laboratory tests (average \pm SD)	
CK	625-4,000 U/L (1,354 \pm 840 U/L)
AST	62-216 U/L (94 \pm 40 U/L)
ALT	44-163 U/L (84 \pm 32 U/L)
LDH	192-824 U/L (443 \pm 207 U/L)
WBC	12.3 \pm 1.2 g/dL
HGB	9.9 \pm 2.7 10 ⁹ /L
PLT	310 \pm 118 10 ⁹ /L
ESR	2-38 mm (25.4 \pm 10.1 mm)
CRP	0.3-3 mg/dL (1.58 \pm 1.03 mg/dL)

SD: Standard deviation, CK: Creatine kinase, AST: Aspartate transaminase, ALT: Alanine transaminase, LDH: Lactate dehydrogenase, PLT: Platelet, WBC: White blood cell, HGB: Hemoglobin, CRP: C-reactive protein, ESR: Erythrocyte sedimentation rate

Table 2. Patients' neurological findings at the first diagnosis, and systemic complications on follow-up

Findings	1. Patient	2. Patient	3. Patient	4. Patient	5. Patient	6. Patient	7. Patient
Cutaneous	Gottron Heliotrope	Gottron Calcinosis	Calcinosis Heliotrope	Gottron Heliotrope	Gottron Heliotrope	Gottron Heliotrope	Gottron Heliotrope
Muscle	Scoliosis	Contracture	Osteoporosis	Contracture Art/Art	-	Osteoporosis	Contracture
Endocrine	-	-	DM	-	-	-	-
Respiratory	Tuberculosis	-	-	-	-	Recurrent respiratory tract infection	-
GIS	Gastritis	-	-	HM	-	-	-
Eye	-	-	-	-	-	-	-
CV	-	-	-	-	-	-	Raynaud's phenomenon
Infection	Tuberculosis	-	-	-	-	-	-
Neurological examination (first diagnosis)							
Muscle force							
-Upper	2/5	5/5	4/5	3/5	4/5	3/5	5/5
-Lower	3/5	4/5	3/5	3/5	5/5	4/5	4/5
DTR							
-Upper	Hypoactive	N	Hypoactive	Hypoactive	N	Hypoactive	N
-Lower	Hypoactive	N	Hypoactive	Hypoactive	N	Hypoactive	N
Neuropathy	+	-	+	-	-	-	-
Seizure	-	-	-	-	-	-	-
Gowers' Sign	+	-	+	+	-	+	-
Others	Scapular winging Lumber lordosis	-	-	Scapular winging	-	-	-

CV: Cardiovascular, DTR: Deep tendon reflex, GIS: Gastrointestinal system, DM: Diabetes mellitus, HM: Hepatomegaly, N: Normal, IKH: Intracranial hemorrhage, ex: Exitus, GTC: Generalized tonic clonic, Art/Art: Arthritis/artralgia.

received a different diagnosis due to difficulties in accessing physicians, and this leads to diagnostic delay.

CK elevation is the most important laboratory parameter for a JDM diagnosis (12). The incidence of elevated CK is reported to be between 87% and 100% (12,13). In this study, all patients had elevated CK levels at the time of diagnosis, and the mean CK level was found to be $1,354 \pm 840$ U/L. Elevated CK levels may lead to confusion in the form of differential diagnoses, as some patients were being followed up by pediatricians with a preliminary diagnosis of muscular dystrophy (MD), based on their elevated CK. However, a detailed evaluation of skin findings, which are reported in almost all cases in different studies, may contribute to an accurate diagnosis. In the

presence of dermatomyositis, ESR and CRP elevations are significant findings in their indication of inflammation. In the study by Barut et al. (12), the mean ESR level was 35 ± 22.1 mm and CRP positivity was reported in 38% of the cases. In this study, the mean ESR value was 25.4 ± 10.1 mm and 80% of the patients were CRP positive. ANA positivity is important for JDM follow-up, while also being a marker of morbidity (13). In the literature, rates of ANA positivity of 68% and 60-70% have been reported, while in this study, 12 (53.3%) of the patients were found to be ANA positive, and better results were achieved in 10 of them during both the acute and long-term periods. ANA positivity can be considered a good predictive factor for treatment response (12,15).

8. Patient (ex)	9. Patient	10. Patient	11. Patient	12. Patient	13. Patient	14. Patient	15. Patient
Gottron Heliotrope Alopecia Calcinosis Ulcer	Gottron Heliotrope	Gottron Heliotrope	Gottron Heliotrope	Gottron	Gottron Heliotrope Alopecia	Gottron Heliotrope	Gottron Alopecia
Art/Art Contracture	Contracture Art/Art	-	Contracture Art/Art	-	Osteoporosis	Art/Art	-
-	-	-	-	-	-	-	-
Recurrent respiratory tract infection	-	-	-	-	-	-	-
Dysphagia	-	-	-	-	Dysphagia	-	Dysphagia
-	-	-	-	-	-	-	-
Raynaud's phenomenon	-	-	-	-	Raynaud's phenomenon	-	-
Sepsis (ex)	-	-	-	-	-	-	-
5/5 4/5	4/5 4/5	5/5 5/5	5/5 4/5	4/5 4/5	4/5 4/5	5/5 4/5	4/5 4/5
N N	Hypoactive Hypoactive	N N	N N	Hypoactive Hypoactive	N N	N N	Hypoactive Hypoactive
-	-	-	-	-	-	-	-
+ (GTC)	-	-	-	-	-	-	-
-	+	-	+	+	+	-	-
-	-	-	-	-	Lumber lordosis	-	-

Proximal muscle weakness and typical skin rashes are the pathognomonic clinical findings of JDM (16). In the study by Gowdie et al. (9), 95% of the patients had muscle weakness, 91% had Gottron's papules and 71% had a heliotrope rash. Barut et al. (12), on the other hand, reported a heliotrope rash in 100% of cases, Gottron's papules in 96% and muscle weakness in 90%. Concurring with the findings in the literature, muscle weakness, Gottron's papules and heliotrope rash were present in 100%, 93.3% and 80% of the patients in this study, respectively. Supporting this data, the presence of skin findings in almost all JDM cases admitted with muscle weakness would appear to be a valid aid to clinicians when making a definite diagnosis, although it should be kept in mind that amyopathic dermatomyositis

may also be observed, particularly in pediatric patients (1). Muscle findings are one of the most important clinical manifestations of the disease. Proximal muscle weakness, particularly at the hip and shoulder intersections, is the most important clinical finding. Affected children experience limited movement, may experience difficulties in climbing stairs and are frequently positive for Gower's sign (17). In this study, 14 of the patients had muscle involvement as the most common complaint at the time of admission, with lower and upper extremity involvement noted in 86.6% and 60% of the patients, respectively. One patient (patient number 10), on the other hand, had no muscular involvement, and this patient was followed up with a diagnosis of JDM. When presenting with CK elevation, this finding causes cases to be

followed erroneously with a diagnosis of MD, although AST levels may help differentiate such cases from MD, as such cases do not present with the elevated AST levels seen in MD cases, despite findings of myopathy. Our case only had a mild CK elevation. Duchenne muscular dystrophy (DMD) is the most common type of MD seen during childhood. While differentiating this condition from DMD, the dominance of the female sex in JDM should be considered, although symptomatic female DMD carriers should also be kept in mind.

While the leading means of diagnosis of JDM is standard muscle biopsy, an EMG and muscle MRI may also be helpful (18,19). In the study by Malek et al. (13), the findings of myositis in EMG was reported to be 96% and myositis in muscle biopsy was reported to be 93.7%. In the study by Okong'o et al. (14), a muscle MRI was performed in 36% of the patients, and the findings favored inflammation in 88.8% of the cases. In this study, muscle biopsy was carried out on the 66.6% of patients who agreed to the procedure, while a muscle MRI was performed on the remaining 33.4% of the patients. In both groups, 60% of the patients underwent an EMG. The findings pointed to myositis in 100% of the patients who underwent muscle biopsy, 88.8% of the patients who underwent an EMG and 100% of the patients who underwent an MRI. In our study, 33.4% of the patients underwent an MRI, and inflammation was noted in 100% of cases. Although muscle biopsy is the leading method of diagnosis, a muscle MRI can also be safely used as a non-invasive procedure.

Recent recommendations for treating JDM suggest the use of corticosteroids and methotrexate in combination (intravenous methylprednisolone 15-30 mg/kg/g and 1-2 mg/kg/g prednisolone+15-20 mg/m²/patient methotrexate for three consecutive days). A more conventional treatment approach is the use of intravenous methylprednisolone at 15-30 mg/kg/day and maintenance prednisolone at 1-2 mg/kg/g. Our treatment protocol was solely based on the use of steroids in the first stage, and treatment-resistant patients were administered methotrexate following steroid therapy. Other immunosuppressive therapies were given to patients who encounter episodes despite the methotrexate treatment, and well-implemented physical therapy facilitates and supports this treatment process (12). All the patients in this study underwent physical therapy and immunosuppressive therapy. A complete response to treatment was noted in 12 (80%) of the 15 patients during the acute phase, and in the long-term, six (40%) patients followed a treatment-resistant disease course and required multiple immunosuppressive therapies.

Table 3. Patients' positive examination findings of

Physical examination	n (%)
Skin	
Gottron's papules	14 (93.3)
Heliotrope rash	12 (80)
Calcinosis	3 (20)
Alopecia	3 (20)
Muscle-skeleton	
Contracture	6 (40)
Arthritis/arthralgia	5 (33.3)
Osteoporosis	3 (20)
Endocrine	
Diabetes mellitus	1 (6.6)
Respiratory	
Recurrent respiratory tract infection	2 (13.3)
Tuberculosis	1 (6.6)
Gastrointestinal	
Dysphagia	3 (20)
Eye	
	-
Cardiovascular	
Raynaud's phenomenon	3 (20)
Infection	
Sepsis	1 (6.6)
Neurological	
Lower extremity involvement	13 (86.6)
Upper extremity involvement	9 (60)
DTR hypoactive	7 (46.6)
Gowers' sign	8 (53.3)
Scapular winging	2 (13.3)
Lumber lordosis	2 (13.3)

DTR: Deep tendon reflex

Calcinosis is one of the most significant complications of JDM, being seen in 18-27.7% of children with JDM, as well as being one of the major complications affecting morbidity. It may already be present at disease onset or it may develop in later stages (11). In the study by Barut et al. (12), calcinosis was observed in 38% of the patients, while calcinosis was noted in only three (20%) patients in this study. During long-term follow-up, the involvement of the musculoskeletal or gastrointestinal systems can seriously affect the quality of life of patients with JDM (12,13). In our study, gastrointestinal involvement and musculoskeletal

system involvement was observed in five and eight patients, respectively. JDM is also associated with mortality; with one patient (6.6%) in our study dying from intracranial bleeding and sepsis, while in the study by Okong’o et al. (14), the mortality rate was reported to be 8%.

CONCLUSION

In conclusion, JDM is a rare inflammatory myopathy seen during childhood, and it has a high rate of morbidity and low mortality. Better responses can be achieved with early diagnosis, intensive immunosuppressive therapy and effective physical therapy.

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ETHICS

Ethics Committee Approval: The ethics committee decision regarding our study was taken from the Ethics Committee of İnönü University Faculty of Medicine with the decision reference number 2018/9-17 (date: 24.04.2018).

Informed Consent: We received informed consent from all the patients’ families.

Authorship Contributions

Surgical and Medical Practices: M.A., S.G., Y.T., B.Ö., S.K., Concept: S.G., Y.T., Design: M.A., S.G., Y.T., Data Collection or Processing: M.A., B.Ö., S.K., Analysis or Interpretation: M.A., S.G., Y.T., S.K., Literature Search: M.A., S.G., Y.T., B.Ö., S.K., Writing: M.A., B.Ö.

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