IJN
Iranian Journal of Neonatology
http://ijn.mums.ac.ir

Open Access
Case Report

Juvenile Idiopathic Arthritis Onset in a Neonate: A Rare Case Report

Abdolreza Malek[1], Mohammad-Saeed Sasan[1], Narges Afzali[2], Somayeh Ghahremani[3], Sara Ghahremani[1]*

1. Department of Pediatrics, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran
2. Department of Radiology, Mashhad Branch, Islamic Azad University, Mashhad, Iran
3. Nuclear Medicine Research Center, Mashhad University of Medical Sciences, Mashhad, Iran

ABSTRACT

***Background*:** A common type of chronic arthritis in children and adolescents is juvenile idiopathic arthritis (JIA). According to the International League of Associations for Rheumatology (ILAR) classification, JIA diagnostic criteria include age under 16 years and disease duration of six-weeks. Based on the number of involved joints in the first six-months of disease onset, JIA is categorized into oligoarticular or polyarticular subtypes. Age is a characteristic factor in the diagnosis of disease subsets; it is worth mentioning that cases younger than six months of age are seldom found in any of the subtypes.

Case report: In this report, we present a rare case of JIA in an infant, presenting at 20 days of age. Effusion of the right hip joint was one of the primary manifestations of the disease. During hospitalization, she went through sepsis work-up and a four-week antibiotic therapy for management of lower limb pseudoparalysis. In spite of antibiotic therapy, she developed effusion of a second joint. According to the course and duration of symptoms and ILAR classification for JIA, oligoarticular JIA was diagnosed and treated.

Conclusion: In this case, infectious diseases, such as tuberculosis and brucellosis, and malignancies were ruled out as a cause of inflammation through bone marrow aspiration, culture, and tests; ultrasound and magnetic resonance imaging showed no lytic and sclerotic lesions or a fracture. Our experience showed a rare case of JIA and suggested that JIA must be considered in children with joint inflammation at any age.

Keywords: Chronic arthritis, Juvenile idiopathic arthritis, Neonate

Introduction

A common chronic musculoskeletal disease in children and adolescents is arthritis that brings about short- to long-term disabilities and need to care (1, 2). There are several types of arthritis, the most common of which is juvenile idiopathic arthritis (JIA) (3). The International League of Associations for Rheumatology (ILAR) has developed a classification for the pediatric types of JIA in order to homogenize diagnosis and treatment modalities with respect to a single standardized classification. The recent classification also modified what formerly was introduced by American College of Rheumatology and European League against Rheumatism (4).

The new classification of JIA introduced oligoarticular, polyarticular (five or greater than five involved joins, respectively) and systemic subtypes. In this classification, the age of the child should be less than 16 years and disease duration more than six weeks. While the disease diagnosis is clinical, there is some evidence regarding the higher applicability of ultrasound or magnetic resonance imaging (MRI) in detecting the involved joints compared with physical examination and history taking (5, 6).

The involved joints show good response to disease modifying anti-rheumatic drugs (DMARDs), particularly methotrexate (MTX), which are well-tolerated and commonly used. Since 1970, it has been proven that JIA

* *Corresponding author*: Sara Ghahremani, Mashhad University of Medical Sciences, Imam Reza Hospital, Mashhad, Iran. Tel: +985138593045; Email: Gahremanis@mums.ac.ir

Please cite this paper as:
Malek A, Sasan MS, Afzali N, Ghahremani S, Ghahremani S. Juvenile Idiopathic Arthritis Onset in a Neonate: A Rare Case Report. Iranian Journal of Neonatology. 2017 Jun: 8(2). DOI: 10.22038/ijn.2017.22119.1261

demonstrates satisfactory response to DMARD treatment, especially MTX (7).

The disease does not seem to be rare, but its prevalence remains unknown. It has a global distribution, however, the incidence of the disease is widely different among various regions. The literature, mostly originated from the western countries and Australia, reported a prevalence of about 1.7-8.4 million for chronic arthritis. Nonetheless, this rate cannot be accurate as it is commonly missed or misdiagnosed as general joint inflammation (8, 9). About half of the patients with JIA suffer from the oligoarthritis (1).

Studies on JIA in the Asian population revealed polyarticular and systemic subtypes to be the most prevalent forms of the disease in India and China, respectively (10, 11). In Iranian children, polyarticular subtype was reported to be the most prevalent (1).

Although the incidence rate of IJA varies depending on the type of involvement, it is reported to be two times higher in girls than boys in the western countries and Australia. In contrast, in Turkey, Africa, India, and Iran gender discrepancy is not present or at times boys are the more commonly involved ones (12, 13). JIA is present in the different countries based on the biological variations and/or case ascertainment bias. Age of onset of JIA is key to identifying the disease subtype, and cases younger than six months are seldom diagnosed with this disease.

In this report, we present a rare case of JIA in an infant, in whom the disease manifestation appeared earlier than 20 days of age. Our experience highlights the need to consider JIA in newborns.

Case report

In the current report, a female nine-month-old infant was admitted to Pediatric Ward of Imam Reza Hospital at the age of 40 days with stiffness of the left lower limb and restlessness for two weeks. The disease manifestations began at 20 days of age.

She was afebrile and laboratory data were normal as presented in tables 1 and 2. Ultrasound imaging of the hip showed effusion with maximum size of 2 mm, asymmetric bulging of the articular capsule in the left hip joint, and synovial hypertrophy. X-ray showed no lytic or sclerotic lesions and no fracture. In the magnetic resonance imaging (MRI) coronal section, head, neck, and shaft of femur were bilaterally normal.

TC99m bone scan was performed and reflected elevated tracer activity in the left hip joint suggesting inflammatory process (arthritis and synovitis; Figure 1).

She underwent sepsis work-up and antibiotic therapy for the management of lower limb pseudoparalysis for four weeks. In spite of antibiotic therapy, the patient developed effusion of the left hip joint, as well. Considering the course and duration of symptoms and ILAR classification for JIA, oligoarticular JIA was diagnosed and treated. Ultrasound imaging demonstrated no effusion in the left hip and ankle joints. At one-year follow up, she could creep, crawl, and stand with support, her length of lower limbs was symmetric.

Table 1. The laboratory data of juvenile idiopathic arthritis case

Test	Result	Test	Result
CBC diff		ANA* (U/ml)	0.2
White blood cells × (10^3/µl)	8.0	Urea (mg/dl)	35
Neutrophils (%)	20.0	Creatinine (mg/dl)	0.4
Lymphocyte (%)	75.0	Erythrocyte sedimentation rate (1 h)	64
Mixed (%)	5.0	C-reactive protein (mg/dl)	8.4
Red blood cell × (10^6/µl)	3.5	Wright	Neg.
Haemoglobin (g/dl)	10.5	2ME	Neg.
Platelets × (10^3/µl)	442	Anti CCP**	1.1
		RF*** (IU/ml)	6

*Antinuclear antibody: less than 0.8 is interpreted as negative.
**Anti-cyclic citrullinated Peptide antibody: less than 2 is interpreted as negative.
***Rheumatoid factor: less than 10 is interpreted as negative.

Table 2. Diagnostic procedures of juvenile idiopathic arthritis case

Test	Synovial fluid smear and culture	Bone marrow aspiration	PPD*
Result	Neg.	Active marrow	Neg.

*A purified protein derivative skin test.

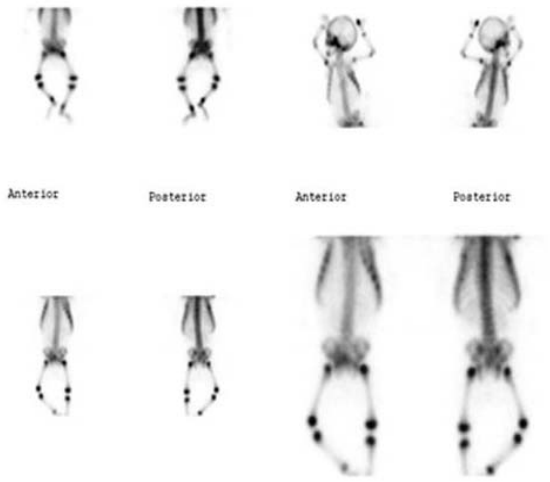

Figure 1. Tc99m-MDP Bone scan

Discussion

JIA is a globally common chronic rheumatoid disease in the pediatric population that leads to hospitalization (14). In definition, the joint inflammation and arthritis duration should be six weeks in patients younger than 16 years. Besides, the other differential diagnoses of arthritis and injuries should be ruled out and the clinical manifestations remain for at least six months.

In this study, we presented a rare case of JIA in an infant, whose clinical manifestations appeared during the neonatal period (20 days of age). In this case, infectious diseases like tuberculosis and brucellosis and malignancies were excluded as a cause of inflammation through bone marrow aspiration, culture, and tests; ultrasound and MRI imaging showed no lytic and sclerotic lesions neither a fracture.

The patient developed effusion of the right ankle joint during the four weeks of hospitalization in spite of sepsis work up and antibiotic therapy, confirming the 2-joint oligoarticular JIA diagnosis.

In this case, the patient was afebrile and involvement of two large joints (the left hip and ankle) suggested the oligoarticular subtype of JIA. Based on this diagnosis, treatment with nonsteroidal anti-inflammatory drugs (ibuprophen) and disease modifying anti-rheumatic drug started. The patient successfully improved and was discharged from hospital. Frequent follow up for nine months showed response to our treatment. She received low doses of ibuprophen and MTX; treatment with MTX was planned for one year.

According to ILAR classification, there are seven subtypes for JIA; the most common subtype is oligoarticular, defined based on the number of joints with inflammation (less than five) (15). However, studies on JIA by Fujikawa and Moe performed in Asian populations showed polyarticular and systemic subtypes to be the most prevalent forms of disease in India and China, respectively (10, 11). In Iranian children, polyarticular subtype was reported to be the most prevalent subtype (1).

In our previous study (1) and the study by Kahn (3), children suffering from JIA presented multiple symptoms consisting of fever, anorexia, weight loss, sleep disorder, impaired growth, uveitis, and joint stiffness; these symptoms are shown less clearly in oligoarticular subtype (1). Prince et al. reported that large joints are more frequently involved in oligo- and polyarticular subtypes (15). These studies confirm our diagnosis based on patient's symptoms and signs.

The disease involves girls more than boys, but there are some exceptions, as well (13). While age is a criterion for classification of the disease, it is rarely observed in the neonates younger than six months (1).

Conclusion

Our experience showed a rare case of JIA in our population and JIA must be considered in children with joint inflammation and systemic symptoms at any age. Generally, choosing the appropriate and safe medications is essential for each patient; thus, further studies are required to find a curative treatment for JIA.

Acknowledgments

The authors appreciate the cooperation of Nooshin Abdollahpour, who provided technical help.

Conflicts of interests

None.

References

1. Malek A, Aghighi Y, Parvisy BP, Raeeskarami SR, Ziaee V. Epidemiologic investigation of juvenile rheumatoid arthritis: a ten-year study. Tehran Univ Med J. 2014; 72(3):187-93 (Persian).
2. Sherry DD, Jung LK. Juvenile idiopathic artritis. Tehran: Medscape; 2016.
3. Kahn PJ. Juvenile idiopathic arthritis - what the clinician needs to know. Bull Hosp Jt Dis. 2013; 3(71):194-9.
4. Petty RE, Southwood TR, Manners P, Baum J, Glass DN, Goldenberg J, et al. International League of Associations for Rheumatology classification of juvenile idiopathic arthritis: second revision, Edmonton, 2001. J Rheumatol. 2004; 31(2):390-2.

5. Parentin F, Matarazzo L, Lepore L, Pensiero S, Ventura A. Early onset bilateral anterior uveitis preceding a late manifestation of juvenile idiopathic arthitis. Ocul Immunol Inflamm. 2015; 23(1):102-5.
6. Al-Hemairi MH, Albokhari SM, Muzaffer MA. The pattern of juvenile idiopathic artritis in a single tertiary center in Saudi Arabia. Int J Inflamat. 2016; 2016:1-8.
7. Funk RS, Becker ML. Dissease modifying anti-rheumatic drugs in juvenile idiopathic artritis: striving for individualized therapy. Exp Rev Precision Med Drug Dev. 2016; 1(1):53-68.
8. Thierry S, Fautrel B, Lemelle I, Guillemin F. Prevalence and incidence of juvenile idiopathic arthritis: a systematic review. Joint Bone Spine. 2014; 81(2):112-7.
9. Danner S, Sordet C, Terzic J, Donato L, Velten M, Fischbach M, et al. Epidemiology of juvenile idiopathic arthritis in Alsace, France. J Rheumatol. 2006; 33(7):1377-81.
10. Fujikawa S, Okuni M. A Nationwide surveillance study of rheumatic diseases among Japanese children. Actas Paediatr Jpn. 1997; 39(2):242-4.
11. Moe N, Rygg M. Epidemiology of juvenile idiopathic arthritis in northern Norway. Clin Exp Rheumatol. 1998; 16(1):99-101.
12. Weakley K, Esser M, Scott C. Juvenile idiopathic arthritis in the Western Cape, South Africa. Pediatr Rheumatol Online J. 2012; 10(1):35.
13. Yilmaz M, Kendirli SG, Altintas DU, Karakoc GB, Inal A, Kilic M. Juvenile idiopathic arthritis profile in Turkish children. Pediatr Int. 2008; 50(2):154-8.
14. Huang H, Qian X, Yu H, Li J, Zhang Y. Clinical analysis in 202 children with juvenile idiopathic arthritis. Clin Rheumatol. 2013; 7(32):1021-7.
15. Prince FH, Otten MH, van Suijlekom-Smit LW. Diagnosis and management of juvenile idiopathic arthritis. BMJ. 2010; 341:c6434.