Early Symptomatic Cervical Spine Involvement in an Infant with Polyarticular Juvenile Idiopathic Arthritis

Satyabrata Roychowdhury¹, Satarupa Mukherjee¹, Kalpana Datta²

Introduction

Poly-articular juvenile idiopathic arthritis (JIA) is defined as an involvement of 5 or more joints exhibiting symptoms of pain, redness, swelling or restriction of movement in children less than 16 years of age lasting for more than 6 weeks [1]. The peak age of disease onset is usually around 5 years with a female predisposition. Spine involvement, particularly at disease onset, is quite rare in JIA. Here, we report a case of severe symptomatic cervical spine involvement at disease onset in an infant with JIA, the first case ever reported in the literature.

Case Report

A one-year old male baby was admitted with symptoms of an inability to move his neck, occasional fever and swollen wrist, elbow and ankle joints for the last 4 months. The irritable baby was not turning his head, but instead turned his entire body to look at objects around the room and cried on minimal handling (Picture A). He was born at term, appropriate for the gestational age, and achieved the age-appropriate developmental milestones until 8 months of age. Age-appropriate language and social milestones were reached. The

Abstract

Juvenile idiopathic arthritis (JIA), a spectrum of chronic arthritis in children, may involve the cervical spine, although it is not as commonly found as in adults with rheumatoid arthritis. Many children may remain asymptomatic. An MRI should be performed, even if the patient is asymptomatic, to detect early changes, followed by the immediate initiation of aggressive treatment. Early symptomatic cervical spine involvement is quite rare in children with JIA.

Key words: Poly-articular juvenile idiopathic arthritis, cervical spine
The perinatal period was uneventful. The last milestone achieved was sitting without support at 7 months; however, since then there have been no signs of standing or walking. The presenting fever was low grade (37.3-38.0 degrees Celsius), spiking twice daily, with the temperature returning to baseline. The bilateral elbow and wrist, and small joints of the hand, knee, and ankle were swollen starting from 9 months of age and gradually increased in severity. The joints were tender with restricted overall movement. There was no history of any loss of appetite, weight loss, behavioral disturbances, skin rash or abnormal bleeding. Systemic examination revealed no abnormalities regarding organomegaly and lymphadenopathy. Diagnostic tests showed a normocytic and normochromic anemia, raised erythrocyte sedimentation rate (ESR = 57 mm in the first hour) and C-reactive protein (CRP) levels of 8 mg/dl. Tests for anti-nuclear antibody (ANA) and rheumatoid factor (RF) were negative. A bone marrow examination was within normal limits showing normal haemopoiesis and erythroid hyperplasia. An X-ray of the joints revealed soft tissue swelling (Picture C). An X-ray of the cervical region of the neck showed retro-pharyngeal soft tissue swelling (Picture D). A USG of the knee joint revealed effusion in the bilateral joint space with a thickened synovium. An MRI of the cervical spine showed erosion of the odontoid process, C1-C2 arthritis and apophyseal joint arthritis. Screening for HIV (human immunodeficiency virus) infection was non-reactive, as well as Venereal Disease Research Laboratory Test (VDRL) for syphilis for both the mother and her baby. A Mantoux test was negative and there was no history of any contact. An ELISA for brucellosis was negative. Urine analysis for glycosaminoglycans was negative, ruling out mucopolysaccharidosis. A chest X-ray was normal and there was no evidence of any fluid accumulation in any other body cavity. Therefore, a diagnosis of polyarticular juvenile rheumatoid arthritis with symptomatic cervical spine involvement was made. The baby was started on NSAID, naproxen for 2 months, with no apparent improvement. He was thereafter started on oral methotrexate with a bridging period of an oral steroid. Rehabilitative physiotherapy continued. The joint swellings started decreasing with subjective improvement within 3 months of methotrexate therapy. He is at present under follow-up and is on the 9th month of methotrexate treatment.

Discussion

Although early symptomatic cervical spine involvement in JIA has been previously reported in the literature, it deserves special mention due to its rarity, severity and the need for aggressive treatment. As with adult rheumatoid arthritis where cervical spine involvement is a recognized feature, it is not a recognized feature in JIA. A review of the literature [2] suggests that the median age of onset of JIA is approximately 6 years, with symptomatic cervical spine involvement occurring at approximately 7 years of age, and many patients remaining asymptomatic. While the most common symptom was limited range of motion, pain was absent in many cases and the majority of patients were entirely asymptomatic [2]. Our case involved an infant that presented with severe pain and a re-
striction of movement. Investigators also found that cervical spine arthritis is a severe and persistent manifestation of JIA with severe skeletal sequelae [2]. Although X-ray remains the most widely used imaging technique of choice for diagnosis, it may miss the early signs of cervical spine involvement of joint effusion and bone-marrow changes, thereby possibly delaying the immediate start of aggressive treatment. An MRI should be performed initially to avoid missing important findings and hence delaying treatment. However, Kjellberg et al. demonstrated that lateral cephalometric radiographs can detect early changes and should be performed when applicable [3]. Children with cervical spine involvement often require aggressive treatment with biologics [2] to prevent impairment and disability, which hampers their quality of life. Hospach et al. treated almost all their patients suffering with cervical spine involvement with biologics, prompting the need of aggressive treatment [2]. Semolic et al. found a prevalence of cervical spine involvement of 3% in children, with the majority of them showing good response to anti-TNFα agents [4]. Investigators have found that JIA is not as benign as was previously thought, as many patients may experience persistent inflammation and disability in adulthood [5].

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**References**


