

## PATTERNS OF RADIOGRAPHIC DAMAGE TO CERVICAL SPINE IN POLYARTICULAR JUVENILE IDIOPATHIC ARTHRITIS PATIENTS PRESENTING TO TERTIARY CARE HOSPITAL IN PAKISTAN

Ehtisham Khyzer, Tehreem Aftab\*, Hassan Junnaid\*\*

Pakistan Institute of Medical Sciences Islamabad Pakistan, \*Rawal Institute of Health Sciences Islamabad Pakistan, \*\*Combined Military Hospital Sialkot Pakistan

### ABSTRACT

**Objective:** To see the radiographic cervical spine damage in polyarticular juvenile idiopathic arthritis (PJIA) coming to a tertiary care hospital in Islamabad, Pakistan.

**Study Design:** Cross-sectional descriptive study.

**Place and Duration of Study:** The study was conducted in department of Rheumatology at Pakistan Institute of Medical Sciences from Jun 2013 to Dec 2013.

**Subjects and Methods:** A total of 50 patients of PJIA coming to Rheumatology Outpatient Department were recruited in the study after informed consent. Radiographs of cervical spine were performed for each patient in antero-posterior, lateral with flexion and extension and open-mouth views. Radiographs were reviewed for the following features: loss of cervical lordosis, odontoid process erosion, anterior atlantoaxial subluxation, C1-C2 arthritis, atlantoaxial impaction, inflammation of disc, apophyseal joint arthritis, anterior ankylosis, apophyseal joint ankylosis, anterior and posterior subaxial subluxation and growth disturbances. Data was analysed using SPSS version 18.

**Results:** Out of the total 50 patients, 28 (56%) were females while 22 (44%) were males. The mean duration of PJIA was  $5.54 \pm 3.28$  years. Radiological cervical spine involvement was seen in 52% patients. The most common structural lesions were anterior atlantoaxial subluxation (30%), C1-C2 arthritis (22%) erosion of the odontoid process (18%), and apophyseal joint arthritis (16%). Loss of cervical lordosis was found in 7(14%) patients. There was no growth disturbances observed in vertebra.

**Conclusion:** Cervical spine involvement is common in patients of PJIA. It is mostly asymptomatic, so routine cervical spine radiographs in all patients suffering from PJIA is recommended.

**Keywords:** Cervical spine, Polyarticular juvenile idiopathic arthritis, Radiographs.

### INTRODUCTION

Juvenile idiopathic arthritis (JIA) is a systemic inflammatory disease characterized by the onset of arthritis prior to the age of 16 years with symptoms that persist for more than 6 weeks<sup>1</sup>. JIA is heterogeneous group of diseases and polyarticular JIA (PJIA) has been identified as its important subtype<sup>2</sup>. It is difficult to diagnose due to the lack of definite biomarkers, wide range of presentation and overlap of symptoms with other diseases. Polyarticular joint disease has multiple etiologies and symptoms of arthritis are not certainly diagnostic of JIA. It may begin as a viral illness or can be post-infectious. So to accurately

diagnose polyarticular JIA, the most crucial step is to exclude arthritis with known etiologies<sup>3</sup>.

Radiographic cervical spine involvement has been reported in 5–80% of patients with PJIA<sup>4</sup>. JIA causes permanent changes in the joints of the extremities and spine. Extra-articular changes and interference with normal growth has also been observed in severe disease. The pathogenesis of cervical spine involvement in PJIA is synovial inflammation, hyperemia, and pannus formation at the occipitoatlantoaxial joints resulting in apophyseal joint ankylosis or fusion at C2–3 (vertebrae no. 2–3) level<sup>1</sup>. It is a characteristic cervical spine change in PJIA in 41–52% leading to growth disturbances especially in patients with early onset<sup>4</sup>.

Anterior atlantoaxial subluxation (aAAS), subaxial subluxation (SAS) at the second and seventh cervical vertebrae, and erosions of the

Correspondence: Dr Ehtisham Khyzer, Dept of Rheumatology, PIMS Islamabad, Pakistan

Email: doc\_ehtisham@yahoo.com

Received: 19 Feb 2015; revised received 24 Apr 2015;

accepted 29Apr 2015

odontoid peg resulting in an “apple core” deformity have been observed<sup>5</sup>.

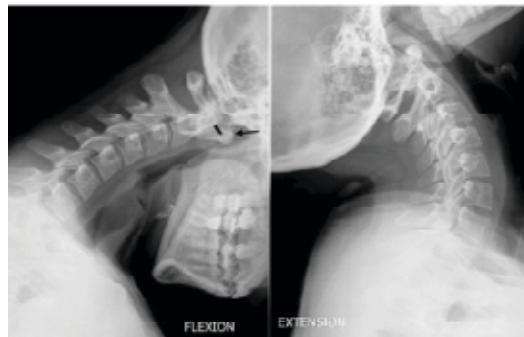
Involvement of cervical spine in PJIA patients is considered a feature of poor prognosis by the American College of Rheumatology (ACR)<sup>6</sup>. A high prevalence of radiographic damage to cervical spine in JIA has been reported previously. However, most of the studies were performed before the advent of modern biotherapies and included only the most severe patients<sup>7</sup>. Therefore, the occurrence and features of radiographic damage to cervical spine in PJIA in the modern world is not exactly known. Damage to cervical spine is an indication of severe PJIA resulting in ankylosis, erosions, narrowing of cranio-cervical junction so it has important therapeutic implications. The current study was, therefore, planned to observe the pattern of cervical spine damage in patients of PJIA using standard radiographs.

#### MATERIALS AND METHODS

We carried out a cross-sectional descriptive study. The study was conducted in the Department of Rheumatology at Pakistan Institute of Medical Sciences (PIMS), Islamabad. It was carried out from June, 2013 till December, 2013. Consecutive patients of PJIA of either gender coming to Rheumatology Outpatient Department were recruited in the study after taking their consent. A total of 50 PJIA patients were enrolled in the study. All of these patients were classified as having PJIA according to the International League of Associations for Rheumatology (ILAR) criteria. Patients having history of cervical surgery were excluded.

Demographic details, history of cervical pain or limitation of movement were recorded. Swollen and painful joint counts were done for each patient. Standard radiographs of cervical spine were performed for all patients. Radiographs were evaluated by two radiologists who were unaware of the diagnosis. Radiographic views observed were antero-posterior, lateral with flexion and extension and open-mouth views. The radiographs were evaluated for following characteristics: loss of cervical lordosis, odontoid process erosions, fracture of vertebrae

(indicated by a decline in height of vertebral body), aAAS, arthritis of 1<sup>st</sup> and 2<sup>nd</sup> cervical vertebrae (C1-C2), atlantoaxial impaction (AAI), inflammation of disc, apophyseal joint arthritis,



**Figure-1: Flexion and extension view of cervical spine showing anterior atlantoaxial subluxation.**

**Table-1: Clinical characteristics and cervical lesions observed in PJIA patients.**

Parameter	PJIA patient n=50
Age, years, mean $\pm$ SD	14.2 $\pm$ 4.83
Disease duration, years, mean $\pm$ SD	5.26 $\pm$ 3.28
RF positive	4 (8%)
Radiographic cervical lesion	26 (52%)
Number of lesion/patient	2
Odontoid process erosion	9 (18%)
aAAS	15 (30%)
C1-C2 arthritis	11 (22%)
AAI	0
Apophyseal joint arthritis	7 (14%)
Vertebral collapse	0
Anterior ankylosis	0
Apophyseal joint ankylosis	0
Anterior and posterior SAS and growthdisturbances	5 (10%)
Inflammatory discitis	1 (2%)
Loss of lordosis	5 (10%)

anterior ankylosis, apophyseal joint ankylosis (AJA), anterior and posterior subaxial subluxation (SAS) and disturbance in growth.

Loss of cervical lordosis was defined in cases of cervical kyphosis on lateral view. Odontoid process erosion was considered in case of changes in the typical outline of the odontoid process. aAAS was defined when the anterior atlantodental interval was  $>3\text{mm}$ <sup>8</sup>. C1-C2 arthritis was considered as narrowing of joint space between C1 and C2 without osteophyte. AAI was identified if tip of odontoid moved  $>4.5\text{ mm}$  above McGregor's line on lateral view<sup>11</sup>. Inflammation of disc was recorded if there was narrowing of disc space and endplate erosions without osteophytes. Anterior ankylosis and AJA were considered when vertebral bodies or apophyseal joints were fused. SAS was recorded when a vertebra was displaced  $>1\text{mm}$  as compared to the next lower vertebra. Disturbance in growth involved hypotrophy or hypertrophy of vertebrae in relation with neighbouring vertebrae.

Data was analyzed using SPSS version 18. Descriptive statistics were applied for both qualitative and quantitative variables. Means and standard deviation were calculated for the quantitative variables, while frequencies and percentages were calculated for the categorical variables.

## RESULTS

A total of 50 patients were enrolled in the study. The mean age of the patients was  $14.2 \pm 4.83$  years with a range of 3 to 20 years. Out of the total 50 patients, 28 (56%) were females while 22 (44%) were males. The mean duration of PJIA was  $5.26 \pm 3.28$  years with a range of 1 year to 15 years. Among the 50 patients, 46 (92%) were negative for rheumatoid factor (RF), while the remaining 4 (8%) patients were positive for RF. Out of the total 200 radiographs, 166 (83%) were taken at the Radiology Department of PIMS. Rest of 34 (17%) were done by patients from other facilities.

Radiological cervical spine involvement was seen in 26 (52%) patients. The prevalence of different structural lesion is shown in the table-1. Ten (38%) PJIA patients with cervical symptoms and all patients without clinical signs had radiographic cervical spine involvement. The most common cervical lesion

was aAAS (fig-1). The radiographic cervical involvement was not associated with age at onset.

## DISCUSSION

The present study was done on 50 patients of PJIA to observe radiographic involvement of cervical spine. The structural involvement was observed in 52% of patients with aAAS and C1-C2 arthritis being the most common lesion. The radiographic involvement of cervical spine in symptomatic patients related with a more grave disease.

Previous study has shown a high prevalence of structural cervical involvement of 65% in PJIA progressing into adult life<sup>10</sup>. The prevalence of cervical spine involvement in current study is less as compared to previous one. The disparity may be due to the fact that patients were evaluated at an earlier stage in current study. Mean disease duration and mean age was less as compared to previous study. Secondly our cohort has less severe disease as 92% patients had RF negative polyarthritis. It has been documented previously that RF-positive PJIA progressing into adult age is the most severe type of disease and has poor prognosis<sup>11</sup>.

The pattern of cervical spine involvement was aAAS, C1-C2 arthritis, odontoid process erosion and apophyseal joint arthritis. Similar pattern has been reported in previous study done on patients having PJIA persisting into adulthood. Clinical cervical involvement is usually considered common in JIA. Previous studies have shown a prevalence ranging between 20 and 70%<sup>12</sup>. In present study, only 10 (38%) patients were symptomatic. The shorter disease duration in current study may be attributed to the less prevalence of symptomatic cervical damage. The clinical cervical involvement, however, indicated a more severe disease, similar to the results of previous studies. Since cervical lesions are mostly seen in asymptomatic patients (62%), radiological assessment might be done at onset and then on regular basis during the course of the disease. It has been proposed that modern 2D and 3D imaging called EOS should be used to avoid the

risk of radiation exposure in young PJIA patients.

Our study has certain limitations. It was a cross-sectional study so natural history of disease cannot be studied. Second limitation was quality of radiographs. Majority (83%) of patients had digital X-rays done from our Radiology Department but the remaining 17% had nondigital ones which were done from other sources. Some of these radiographs had problems; mainly with the exposure. MRI was not done in this study as it was not included in the protocol. MRI is however considered helpful in assessing structural cervical spine involvement, particularly when neural structure compressions in the cervical spine is suspected<sup>13</sup>. MRI is also costly and time consuming. Therefore standard X-ray can be used to screen patients for cervical spine damage, whereas MRI might be utilized in cases of neurological manifestation or as a pre-operative examination.

## CONCLUSION

Radiographic cervical spine damage is a common presentation of PJIA. It is mostly asymptomatic and its presence indicates severe disease. Routine assessment of cervical spine through x-rays as part of initial assessment and follow-ups in patients of PJIA is recommended.

## CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

## REFERENCES

1. Enden K, Laiho K, Kautiainen H, Kautiainen MA, Belt E, Kauppi M. Subaxial cervical vertebrae in patients with juvenile idiopathic arthritis – Something special? *Joint Bone Spine* 2009; 76: 519-23.
2. 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; 31:390-2.
3. Singh S, Mehra S. Approach to polyarthritis. *Indian J Pediatr.* 2010; 77: 1005-10.
4. Espada G, Babini JC, Maldonado-Cocco JA, Garcá-Morteo O. Radiologic review: the cervical spine in juvenile rheumatoid arthritis. *Semin Arthritis Rheum* 1988; 17: 185-95.
5. Laiho K, Savolainen A, Kautiainen H, Kekki P, Kauppi M. The cervical spine in juvenile chronic arthritis. *Spine J* 2002; 2: 89-94.
6. Beukelman T, Patkar NM, Saag KG, Tolleson-Rinehart S, Cron RQ, DeWitt EM, et al. 2011 american college of rheumatology recommendations for the treatment of juvenile idiopathic arthritis: initiation and safety monitoring of therapeutic agents for the treatment of arthritis and systemic features. *Arthritis Care Res* 2011; 63: 465-82.
7. Minden K, Niewerth M, Listing J, Biedermann T, Bollow M, Schöntube M, et al. Long-term outcome in patients with juvenile idiopathic arthritis. *Arthritis Rheum* 2002; 46: 2392-401.
8. Kauppi M, Neva MH. Sensitivity of lateral view cervical spine radiographs taken in the neutral position in atlantoaxial subluxation in rheumatic diseases. *Clin Rheumatol* 1998; 17: 511-4.
9. Reiter MF, Boden SD. Inflammatory disorders of the cervical spine. *Spine* 1998; 23: 2755-66.
10. Elhai M, Wipff J, Bazeli R, Freire VR, Feydy A, Drape JL, et al. Radiological cervical spine involvement in young adults with polyarticular juvenile idiopathic arthritis. *Rheumatology* 2013; 52: 267-75.
11. Martini A, Lovell DJ. Juvenile idiopathic arthritis: state of the art and future perspectives. *Ann Rheum Dis* 2010; 69: 1260-3.
12. Hensinger RN, DeVito PD, Ragsdale CG. Changes in the cervical spine in juvenile rheumatoid arthritis. *J Bone Joint Surg Am* 1986; 68: 189-98.
13. Laiho K, Soini I, Kautiainen H, Kauppi M. Can we rely on MRI when evaluating unstable atlantoaxial subluxation? *Ann Rheum Dis* 2003; 62: 254-6.