INTRODUCTION
Spranger, et al. first described progressive pseudorheumatoid chondrodysplasia (PPRC) as a progressive connective tissue disease, which combined the radiological features of Scheuermann’s disease, with radiographic features of juvenile rheumatoid arthritis. The disorder is classified as an autosomal recessive chondrodysplasia, with absence of inflammatory parameters.

Os odontoideum (OO) is an uncommon anomaly in which the odontoid process (OP) is separated from its caudal or basilar part by a transverse gap located at or slightly above the level of the superior articulating facets of the axis. Two main types of OO are recognised; a congenital type and an acquired type, most often due to trauma. In both the types, the OO may be orthotopic or dystopic. In the orthotopic type, the OO is located in the normal position of the OP. In the dystopic type, the OO is at the base of the skull and may articulate or may be fused with the clivus. Os odontoideum can lead to atlantoaxial instability, which can be either reducible or a fixed dislocation.

The clinical history and course, negative rheumatoid tests and the radiographic examinations in the presently reported patient were consistent with PPRC.

CASE REPORT
A 7-year-old girl, born of consanguineous family was referred to our department because of persistent arthralgia along the weight bearing joints. She had been under strict observation at the department of rheumatology since she was 3 years old. She had no neurological deficit except for a recent development of transient episodes of suboccipital pain associated with headache.

Clinical examination showed a girl with no associated facial dysmorphism and normal stature. Prominence of the weight bearing joints was evident. Mild limitation of neck movements was noticed. It was associated with occasional suboccipital pain. Both knees were swollen and felt firm. There was limited flexion and extension of the knees and painful hips. Neurological examination was normal as were the vision, hearing and mental development. Movements of the hip, knee, and ankle joints were restricted and associated with pain. Both knees were swollen and felt firm. Skeletal survey was conducted. The thoraco-lumbar spine were platyspondylic associated with multiple osteophytes and irregular end plates (Figure 1). Broad iliac bodies and femoral necks and irregular acetabular roofs, associated with irregular epiphyses and osteoarthrosis of both hip joints (Figure 2). Sagittal CT scan showed the detached and fragmented odontoid that had become attached to the clivus (dystopic type) of OO associated with sclerotic changes (Figure 3). The odontoid abnormality was still in the silent phase (no radiographic evidence of significant instability), therefore, medical treatment including...

**ABSTRACT**
We report the case of a girl-child who manifested the clinicoradiographic features of pseudorheumatoid arthritis. 3D-CT scan of the craniocervical junction showed distinctive features of dystopic type of os odontoideum. The report highlights the necessity to explore the craniocervical junction in patients with progressive pseudorheumatoid arthropathy.

cervical traction, occasional firm collar use, and anti-inflammatory medications were the initially employed measures. Follow-up would include assessment of cerebellar and brain stem function, gait assessment, and Romberg testing.

If pain was transformed from being transient to persistent and or disabling, she might be referred to stabilization.

**DISCUSSION**

Progressive pseudorheumatoid arthropathy (PPRC) can be confused with rheumatoid arthritis. The distinguishing features are the presence of platyspondyly, a generalised bone dysplasia, and the absence of destructive bone changes. It is an autosomal recessive disorder in which patients exhibit platyspondyly and progressive arthropathy. Pain, swelling and stiffness of the joints are also characteristics, resembling the findings in rheumatoid arthritis, although without synovitis. Confusion with the disorder as presented in this patient is unlikely as Juvenile rheumatoid arthritis is associated with systemic symptoms such as fever, skin rash, lymphadenopathy, hepatosplenomegaly and positive rheumatoid tests.

There have been a number of reports concerning the extent of the bony abnormalities in association with (PPRC). None of the above mentioned reports described the craniocervical abnormalities in patients with (PPRC). Al Kaisi, et al. described a 13-year-old boy who manifested the clinicoradiographic features of PPRC and manifested os odontoideum. A divided odontoid process, in which the cephalad part was detached from its base, and an os odontoideum was present.

Os odontoideum is a condition in which a smoothly corticated ossicle exists, dorsal to the anterior arch of the atlas, taking the place of the rostral dens, but with no bony connections to the body of the axis. In this patient, there was a malformation of the posterior arch of the atlas, and a detached cephalad part of the odontoid process (dystopic os odontoideum), both likely to be a result of a congenital failure or fusion of ossification centres. The disease may be silent that is without any neurological deficits. The development of neurological manifestations of spasticity, hyperreflexia, clonus and proprioceptive loss are red flags signify atlantoaxial instability.

Computed tomography scans provide excellent delineation of osseous deformity patterns in children with syndromic association. These investigations have important implications in understanding the mechanism of injury, judging potential instability, evaluating deformity, and planning surgical procedures.

**REFERENCES**


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**Figure 3:** Ultrasound image of right renal mass.