Butterfly Vertebra and other Congenital Vertebral Anomalies in a Child

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INTRODUCTION

Butterfly vertebra is a rare birth defect of the spine which is generally asymptomatic. Fewer cases of this defect are reported till now. Here we have reported a case of a 2 year old female baby, who presented with bilateral lower limb weakness and an involuntary passage of stools and urine since birth. Magnetic Resonance Imaging (MRI) confirmed the cause, which showed the presence of syrinx in the spinal cord extending from the vertebral level of T9-T11, butterfly vertebra at T11 level and spina bifida occulta. The neurological examination revealed decreased power in the bilateral lower limbs and bladder-bowel involvement.

Keywords: Butterfly Vertebra, Congenital Defect, Spine, MRI, Spina Bifida Occulta

ABSTRACT

Butterfly vertebra is a rare birth defect of the spine which is generally asymptomatic. Fewer cases of this defect are reported till now. Here we have reported a case of a 2 year old female baby, who presented with bilateral lower limb weakness and an involuntary passage of stools and urine since birth. Magnetic Resonance Imaging (MRI) confirmed the cause, which showed the presence of syrinx in the spinal cord extending from the vertebral level of T9-T11, butterfly vertebra at T11 level and spina bifida occulta. The neurological examination revealed decreased power in the bilateral lower limbs and bladder-bowel involvement.

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Introduction

Occurrence of Butterfly vertebra is a rare birth anomaly of the spine due to symmetric fusion defect during somatogenesis / embryogenesis between 3rd to 6th week of gestational period.

This anomaly is also known by several other names like cleft vertebra, sagittal cleft vertebra, anterior rachischisis, anterior somatoschisis, and anterior spina bifida. Its presentation is generally asymptomatic.

There is aplasia of anterior and median vertebra. The vertebral body has two chondrification centres, their failure of convergence during somatogenesis leads to this defect.

The vertebra with butterfly shape persisting completely or incompletely of associated ‘hemivertebrae’ halves, are separated by the sagittal cartilaginous septum. The peculiar shape of a butterfly is because of cleft and funnel shape ends in the involved vertebra.

Butterfly vertebra occurrence is very common in the lumbar region and only then in the thoracic and other parts of the spine.

Spinal canal narrowing may be found along with this entity because of protrusion of disk at adjacent level.

Multiple congenital syndromes are there in which this anomaly is a part like, Pfeiffer’s syndrome, Jarcho-Levins syndrome, Alagille syndrome, Diastematomyelia and Crouzon syndrome.

Many times it was seen that in search of other diagnosis, incidental finding of butterfly vertebra is there, which rarely creates any neurological problems. It may cause kyphotic deformity of the spine. Differential diagnosis includes the syndromes listed above, vertebral defects, infections and pathologic fractures.

Case Presentation

Parents of a 2 year old female child approached to the
outpatient department, of medicine with the chief complaint of bilateral lower limb weakness, inability of the child to exhibit control in passing stools and urine, a skin dimple at the middle of the vertebral column in the sacral area and an abnormal left foot since birth. There was frequent passage of small amounts of stool and urine. The problem was observed by the parents when the child started standing, walking and communicating. There was no history of trauma and any chronic disease. Developmental milestones of the baby were going normally. In the family, all other children including two elder sisters and one brother were normal. On general examination she was conscious and oriented with normal vitals. Physical examination showed left lower limb Congenital Vertical Talus (CVT) or “rocker bottom foot” deformity, which was present since birth (Figure 1). There was a skin dimple at sacral area indicating “spina bifida occulta” (Figure 2).

Detailed neurological examinations including motor, sensory and reflexes examination were done, which revealed decreased power up to 3/5 in both the lower limbs and 5/5 (normal power) in upper limbs. There was mild atrophy of distal and proximal leg muscles. Pain and temperature sensations were decreased below the lesion while light touch, vibration and position sense were intact (dissociated sensory loss).

Reflexes were decreased in both the lower limbs. Plantar reflex was flexor. An evaluation of the sphincter of the bladder and bowel/rectal sphincter reveals no volitional control. All laboratory parameters were within normal limits. A MRI (Magnetic Resonance Imaging) of the spine was done which revealed syringomyelia in dorsal cord at T9 level with butterfly vertebra anomaly at T11 (Figure 3).

Discussion

There are four subsets of the spine formation during somatogenesis which includes: mesenchymal, chondrification, primary and secondary ossifications. Generation and fusion of two lateral centres for chondrification occurs in 3rd and 6th week of pregnancy and forms the vertebral column. According to Kaplan, vertebral defects/ anomalies are divided into three types:
neural tube defects, defects of formation and defects of segmentation. A defect of formation was found in present case. Hence it is a partial anterior spina bifida found at single vertebra. Although, global occurrence of congenital vertebral malformation is as common as 0.5–1/1,000 of all live births, but butterfly vertebra malformation is very rare. CVT is associated with approximately 10% cases of spina bifida. Both conditions were found in this case. Only two cases in literature are reported for sacral involvement. Amalgamation of spinal anomalies like spina bifida, supernumerary lumbar vertebrae, intervertebral bars, kyphosis/ scoliosis or kyphoscoliosis, diastematomyelia may also be found with butterfly vertebra. This case is unique in the sense that it is very rare; the fourth reported case in thoracic vertebra after 1990 and diagnosed at the early age of 2 years. While previous case reports from 1990 to 2015, the cases were involved patients who were more than 10 years of age. This case is the second case reported after Patra, where the clinical presentation was bilateral lower limb weakness. Low back pain is most common presentation of butterfly vertebra, or it may be incidental finding/ asymptomatic, rarely may present with limb weakness. The etiology is undiagnosed in 85% of the cases of low backache while ligaments, joints, bones, and discs forms 97% source along with 1% source cause of vertebral anomalies. The clinical features of butterfly vertebra may be periodic, with a low back pain of long duration without any neurological deficits and of low clinical importance Cui reported L6 butterfly vertebra which was associated with neurologic deficit. The mode of treatment is conservative while surgery is preferred in cases presenting with low back pain, sciatica and/or motor deficiency including laminectomy (for root decompression) with or without spinal fusion and instrumentation.

**Learning Points**

1. Finding of Butterfly vertebra is very rare and this case is the fourth rare case report of thoracic vertebra involvement in a very little girl.
2. This case adds to the knowledge and awareness about an uncommon cause of low backache and vertebral dysplasia.

**Conflict of Interest:** None

**References**