

Butterfly vertebra. A case report and a short review of the literature

S. Kapetanakis, E. Giannopoulou, E. Nastoulis, T. Demetriou

Department of Anatomy, Medical School of Alexandroupolis, Democritus University of Thrace, Alexandroupolis, Greece

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A butterfly vertebra is a rare congenital anomaly, encountered as isolated finding or as part of syndromic diseases. We report a case of a 40-year-old female presenting with low back pain and sciatica due to 'butterfly' dysplasia of the first sacral vertebra. This novel case includes posterolateral displacement of the completely separated hemivertebrae, causing left lateral recess stenosis and compression of S1 nerve root. Additionally, we conducted a short review of the literature. Few cases are reported in literature. Only one refers to a sacral vertebra. There is no previous case of a butterfly vertebra that accounts for narrowing of the lateral recess and associated radiculopathy. (Folia Morphol 2016; 75, 1: 117–121)

Key words: congenital sacral anomalies, sagittal cleft vertebra, sciatica, low back pain

INTRODUCTION

Butterfly vertebra or sagittal cleft vertebra is a deformity characterised by the congenital failure of convergence of the chondrification centres of the vertebral body [17]. The deformed vertebra consists of two partially or completely separated 'hemivertebrae' [7], divided by a persistent sagittal cartilaginous septum. The osseous parts are more frequently of equal size and non-displaced from their normal position [7]. Predominant region is the lumbar spine, followed by the thoracic region [4]. The defect is usually asymptomatic and incidentally detected or it may cause episodes of low back pain. In some cases there is co-existence with spinal canal narrowing due to adjacent level disk protrusion.

Few cases have been reported in literature and only one is located at S1 level [1]. We report a rare congenital dysplasia of the type of 'butterfly' vertebra located at S1 vertebral body accompanied by displacement of the bony structures and stenosis of the left lateral recess at the level of S1 nerve roots. Our report is accompanied by a short review of the

existing bibliographic references from 1990, according to which a butterfly vertebra causing sciatica by compromising the lateral recess has not been previously described.

CASE REPORT

Female, 40 years old, presented with complaints of severe low back pain and left leg pain. The onset was gradual 2 weeks before. No other symptoms (gastrointestinal, gynaecological) were present. Neurologic examination revealed sciatica without motor defect. Systematic clinical examination was negative. Her previous medical history includes episodes of low back pain since early adolescence, which resolved by taking non-steroidal anti-inflammatory drugs (NSAIDs) and analgesic therapy. No trauma or previous problems in the back are mentioned. In the past, she had two uneventful pregnancies. Her family history is unremarkable.

Lateral and anteroposterior (AP) radiographs of pelvis were ordered. On the AP view, an osseous defect was recognised, as a radiolucent cleft 'splitting'



Figure 1. Axial computed tomography at the level of the first sacral vertebra (S1). Figure demonstrates an osseous defect sagittally traversing the vertebral body, combined with a coronal deficit mainly on the left. The hemivertebrae are posteriorly dislocated into the sacral canal.

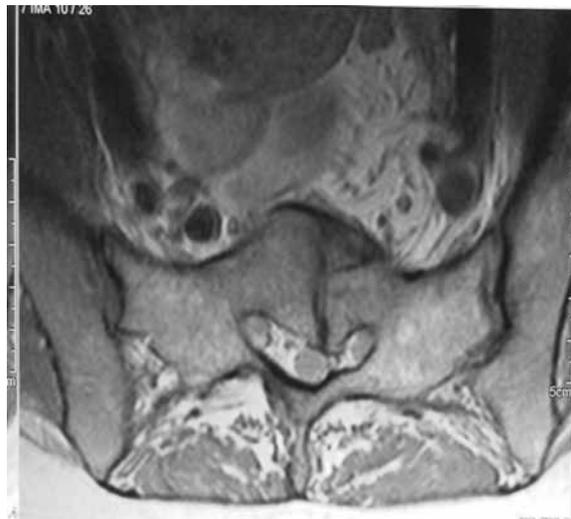


Figure 2. Axial magnetic resonance (T1-weighted) image at S1. The defect is fulfilled with low signal material. Anterior epidural fat is diminished. Dural sac and left S1 nerve root are compressed. Absence of bone or soft tissue oedema.

the first sacral vertebral body approximately at mid-line. Computed tomography (CT) and magnetic resonance images (MRI) of sacral spine were performed in order to further describe the defect.

CT scan revealed an isolated dysplasia of the first sacral vertebra, described as a sagittal cleft of few millimetres in the vertebral body, splitting it into two completely separated 'bony remnants' or hemivertebrae. The cleft followed an AP, slightly left direction and traversed the vertebra at its whole height and length. An additional anterior, coronal cleft was recognised on the left side, as seen in Figure 1. This morphology classifies our case into the rare 'double wedge' type or type 2 according to Cave [2], rather than the 'double D' or type 1 without coronal deficits. The margins of the bony remnants were corticated, sign characteristic of chronicity. Laminae and pedicles were intact. Absence of acute injury was in favour of a 'butterfly' or 'sagittal' cleft vertebra of S1. The hemivertebrae showed posterior displacement, compromising the cross sectional area of the left lateral recess (Fig. 1). Moreover, an incidental finding of a cystic formation was detected in the lower sacral region probably representing an arachnoid cyst.

MRI was also obtained (Fig. 2). The bone gap was fulfilled with material of low and intermediate signal intensity on both T1 and T2 sequences, signs suggestive of fibrous or cartilaginous tissue. Compression of the left S1 nerve root due to lateral

recess stenosis on the left was evident. The fat of the anterior epidural space was almost completely eliminated. No signs of meningeal or epidural fat elements were detected in the bone gap. Oedema of the bone or the para-vertebral soft tissues was absent. Differential diagnosis included compression or pathologic fractures, neoplasm and infection. MRI helped in further characterisation of the cystic formation. The signal was higher from cerebrospinal fluid on T2-weighted and low on T1-weighted sequences. The limits were slightly lobular without septa with dimensions of $5.3 \times 3.5 \times 1.8$ cm (maximum height \times traverse diameter \times maximum sagittal diameter in centimetres). The cyst was diagnosed as intrasacral epidural arachnoid cyst which caused enlargement of the sacral canal and remodelling of the sacral bone due to chronic compression. No bone marrow signal change was present. CT myelography is considered to be redundant, as MRI in most cases is diagnostic as a method of first choice. Cervical, thoracic and lumbar spine appeared free of pathologic findings. An incidental lesion of 1.2 cm diameter at L5 level was consistent with haemangioma.

The patient after the final diagnosis followed a conservative treatment. Analgesic therapy (NSAIDs and cortizone) was prescribed and used according to the course of the symptoms. Complementary physiotherapy (McKenzie Method) was consulted. The patient showed good compliance and the episodes of

low back pain and sciatica decreased in frequency and extensity one year after final diagnosis. Decompression surgery was not indicated at the moment of diagnosis due to the untried effectiveness of conservative therapy and to absence of muscular weakness.

DISCUSSION

Embryologic formation of the spine is divided in 4 overlapping stages: mesenchymal, chondrification, primary and secondary ossification. Formation of the neural plate and the notochord during mesenchymal stage is the onset event that stimulates mesoderm to divide into paraxial, intermediate and lateral on both sides. Paraxial mesoderm condensates to form 42 to 44 pairs of somites at the end of the 3rd week of gestation, which will further differentiate into sclerotomes and dermatomyotomes. Somite formation follows craniocaudal direction and sacrum elements are the last to appear at about 31st day of intra-uterine life [17]. The 31st sclerotome corresponds to L1–L2 level, considering that every vertebra forms from the fusion of the caudal half of the superior sclerotome with the cephalad half of the inferior sclerotome [3]. Progress of embryogenesis results in the appearance of 2 lateral chondrification centres at each level, which are considered to fuse between 3rd and 6th week, forming the cartilaginous vertebral column. Simultaneously, the notochord degenerates at the level of vertebral bodies and remains only at the intervertebral level to form the nucleus pulposus [4].

Kaplan et al. [9] classified vertebral anomalies into 3 categories: neural tube defects, defects of formation and defects of segmentation. Our case falls into defects of formation. Butterfly vertebra is attributed to failure of convergence of the symmetrical chondrification centres at midline during the 3rd to 6th week of gestation and, therefore, is a type of partial anterior spina bifida localised at 1 vertebral level. The term 'sagittal cleft vertebra' is also used as indicative of the anteroposterior direction of the bony defect [18]. This pattern of defect has been embryologically attributed to non-degeneration of the notochord or the perichordal sheath during the chondrification period or to failure of notochord to separate from the endodermal or ectodermal tissues during somitogenesis. Persistence of notochord explains the existence of normal disc material in the cleft, while persistence of the perichordal sheath — the absence of it. Other embryogenetic mechanisms include vascular supply defects and disordered somitogenesis [4]. Posterior

displacement of the unfused segments is attributed to biomechanical instability and chronic, abnormal force transmission at the lumbosacral junction. However, displacement of the halves is a rare finding [7].

Although, vertebral anomalies are relatively frequent with a global incidence of 0.5–1/1,000 live births [18], a butterfly vertebra is uncommon. The most frequent location is the lumbar spine, followed by the thoracic region [4]. As far as the authors are concerned, a sacral butterfly vertebra has been previously described only once. Boulet et al. [1] described a novel case of S1 sagittal cleft vertebra, accompanied by space narrowing at L5–S1 level. Butterfly formation of vertebra can be part of syndromic diseases such as Pfeiffer, Jarcho-Levin, Crouzon or Alagille [1, 16] or associated by additional spinal anomalies such as intervertebral bars, supernumerary lumbar vertebrae, spina bifida, diastematomyelia, kyphosis/scoliosis or kyphoscoliosis [5].

We reviewed the literature for cases of isolated butterfly vertebra. Inclusion criteria were date of publication from 1990 until now, absence of diagnosed syndromic disease and full access to article's content. Cases of prenatal diagnosis were excluded. We describe 12 cases of reported 'butterfly vertebrae' with reference to location, clinical presentation, gender and age of diagnosis. The results are presented at Table 1. No previous study comments on the total number of reported cases of butterfly vertebrae, added in the literature in the last two decades. Finally, all of the articles that met the inclusion criteria are published no sooner than 2001 with the great majority being added after 2011. Raising knowledge on spine biomechanics has refreshed the interest on rare anatomic variants and their clinical relevance. Our case differs due to its extremely rare location at S1. Moreover, this is the first case report, which radiologically describes a posterior displacement of the halves of a butterfly vertebra and associates ipsilateral sciatica to this disordered vertebral morphology.

Our review states that despite its rarity, a butterfly vertebra is clinically presented by low back pain, a very common clinical entity, especially among people of 30 to 50 years of age. Low back pain is considered to be an episodic disease due to its high rate of recurrence and is the main cause of work loss. Underlying pathology is most frequently (85%) not detected. Mechanical aetiologic factors from bones, discs, ligaments, joints are the source for 97% of low back pain. Congenital anomalies account for another 1% [14].

Table 1. Cases of butterfly vertebra reported in the literature during the period 1990 to 2015, in chronological order

First author	Reference No.	Date	Level	Clinical presentation	Gender	Age
Sonel	[16]	2001	L3	Low back pain, sciatica	Male	37
Satpathy	[13]	2004	L1	Incidental	Female	13
Patinharayil	[11]	2008	T6	Low back pain	Female	46
Ekin	[6]	2010	L2	Low back pain	Male	32
Boulet	[1]	2011	S1	Low back pain	Male	35
Cho	[4]	2011	L4	Sciatica, motor weakness	Female	56
Cui	[5]	2011	L6	Radiculopathy	Female	13
Patra	[12]	2013	C2	Spastic quadriparesis	Female	10
Zuo	[18]	2013	L4	Low back pain	Female	24
Frost	[8]	2014	T8	Mid thoracic pain	Male	54
Sifuentes Giraldo	[15]	2015	T10	Investigation of osteoporotic fracture	Female	62
Kapetanakis		2015	S1	Low back pain	Female	40

T — thoracic; L — lumbar; S — sacral

Isolated dysplasia of the first sacral segment is rare. Cadaveric dissections performed by Larmon [10] proved that congenital malformations of S1 can be an independent cause of low back pain and sciatica by compromising the L5 or S1 nerve roots at the intervertebral foramen. He described 2 variants of S1. The first characterised by projections emerging from the posterior margin of vertebral body, causing impingement on the nerve root; the second characterised by a deep groove on the limit between the body and the transverse processes of S1, causing sharp bending and entrapment of L5 nerve root due to ligamentous fibres [10].

Butterfly vertebra can remain asymptomatic for a long period and diagnosis is rarely established. Clinical manifestations usually include chronic, periodic low back pain, usually of many years duration before diagnosis, without neurological deficits [1, 4, 5, 16, 18]. It was considered to be of low clinical importance. However, raised knowledge on spinal biomechanics established the belief that malformations of the lumbosacral junction interfere with force transmission from spine to pelvis and compromise spine stability, accelerating intervertebral disc or/and apophyseal joints degeneration [1, 4, 16, 18] and causing non-specific low back pain. Neurologic symptomatology can occur in cases of co-existence with adjacent level disc herniation. Herniated disc may be located at inferior [16], superior [1] or correspondent level protruding throughout the sagittal cleft [4]. Cui et al. [5] described a novel case of L6 butterfly vertebra

associated with scoliosis and spondylolisthesis at L5–L6 and L6–S1 levels, where altered lumbosacral anatomy due to butterfly vertebra caused entrapment of L5, L6 roots and neurologic deficit [5]. Moreover, alteration in vascular supply to the anterior area of vertebral body is a potential aetiologic factor for anterior hypoplasia. Subsequent ventral sedimentation and kyphosis can compress the spinal cord, especially in thoracic region, where the spinal canal diameter is smaller [16]. Treatment is symptomatic and in most cases conservative.

We describe a novel case of S1 butterfly vertebra with dorsal projection of the hemivertebrae within the sacral canal, causing stenosis of the left lateral recess and impingement on left S1 nerve root. S1 dysplasia was recognised itself as the cause of chronic low back pain and sciatica in our patient and the finding of the intrasacral, epidural arachnoid cyst considered incidental. Conservative therapy was followed and episodes of low back pain and sciatica gradually recessed in one year. In our patient, aetiology of low back considered to be spinal instability of the L5–S1 junction due to altered biomechanics and of radicular pain compression of the nerve root in the compromised lateral recess. Therefore, if the symptoms (low back pain, sciatica and/or motor deficiency) rebound, surgical intervention should be considered. Surgical approach includes laminectomy (for root decompression) with or without spinal fusion and instrumentation (for mechanical stability) [19, 20]. The patient is free of symptoms until now, 1 year after the conservative intervention.

CONCLUSIONS

Butterfly vertebra is an uncommon vertebral anomaly. Bibliographic references are limited. Our case is the second to describe a sacral butterfly vertebra and the first to comment on its aetiologic, anatomic relation with stenosis of lateral recess. Spine surgeons, orthopaedists and anaesthesiologists should be aware of isolated abnormalities of the first sacral segment, insofar as they compromise the success of surgical procedures in sacral spine and the safety of caudal epidural block and may require specific surgical treatment. This clinical presentation adds to our knowledge of the clinical evaluation of isolated, incidental vertebral dysplasia and underlines the significance of a correct diagnosis.

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