Spine Deformity (2020) 8:819–827
https://doi.org/10.1007/s43390-020-00050-3

CASE REPORT

Thoracolumbar kyphoscoliotic deformity with neurological impairment secondary to a butterfly vertebra in an adult

Anouar Bourghli[1] · Salim M. Abduljawad[1] · Louis Boissiere[2] · Ibrahim Obeid[2]

Received: 20 July 2019 / Accepted: 22 December 2019 / Published online: 5 February 2020

Abstract

Study design Case report.

Objective To describe a rare case of thoracolumbar kyphoscoliosis secondary to a butterfly vertebra in an adult, and its surgical correction technique.

Background Kyphoscoliosis secondary to an isolated butterfly vertebra is rare and its management can be very challenging.

Methods We report the case of a 39-year-old male, complaining of chronic middle and low back pain with unsteady gait and altered sensation of lower extremities. Full spine anteroposterior and lateral X-rays revealed a thoracolumbar kyphosis with an angulation of 60° between T10 and T12, with a short thoracolumbar scoliosis of 32 degrees. CT scan confirmed the presence of a butterfly vertebra at the level of T11 with posterior arch fusion between T10 and T12. MRI showed cord compression at the apex of the kyphosis associated to syringomyelia.

Results The patient underwent a posterior resection of the T11 butterfly vertebra with instrumentation from T8 to L2, and use of a one-sided domino on the convex side and a mesh cage on the concave side for asymmetrical correction and vertebral height preservation. Thoracolumbar kyphosis was corrected to 10°. Scoliosis was corrected to 6°. He could walk on day 2 with a satisfactory clinical and radiological result at 2 years.

Conclusion Literature is sparse on the management of thoracolumbar kyphoscoliosis secondary to butterfly vertebra in the context of neurological impairment. The current case described a surgical treatment strategy to correct both deformity planes simultaneously by a vertebral resection performed through a posterior only approach.

Keywords Butterfly vertebra · Congenital kyphosis · Thoracolumbar · Kyphoscoliosis · Vertebral column resection

Introduction

Butterfly vertebra is a rare congenital anomaly, presenting as a sagittal cleft in the vertebral body due to failure of fusion of the two chondrification centers. It has been mainly described as an isolated finding in the literature, but it can be associated with various syndromes, such as Alagille, Jarcho-Levin, Crouzon and Pfeiffer syndrome [1]. Usually the abnormality occurs in the thoracic spine, rarely in the lumbar or cervical spine [2]. It is usually asymptomatic, but it could lead to chronic back pain due to alteration of spine biomechanics, disc herniation [3] and rarely deformity such as kyphosis or kyphoscoliosis with or without neurological symptoms [4]. Most reported cases on butterfly vertebra focused mainly on the imaging findings [5–8] and only few papers discussed a treatment strategy in case surgery was indicated [9, 10]. We present the case of a male patient, almost reaching his 4th decade, who has been complaining of chronic back pain and legs pain caused by thoracolumbar kyphoscoliosis in relation to a butterfly vertebra at the level of T11.

Case report

A 39-years old male has been complaining of middle and low back pain for over 10 years with progressive onset of bilateral radicular pain with legs heaviness, forcing him to use a case during walking. His past history revealed the use

✉ Anouar Bourghli
Anouar.bourghli@gmail.com

[1] Orthopedic and Spinal Surgery Department, Kingdom Hospital, P.O.Box 84400, Riyadh 11671, Saudi Arabia

[2] Orthopedic Spinal Surgery Unit 1, Bordeaux Pellegrin Hospital, Bordeaux, France

of a brace during childhood for a congenital spinal condition that he could not specify.

Physical examination revealed back pain at the thoracolumbar level with a significant gibbosity noticed clinically, associated to a low back pain probably due to a facet joint syndrome as hyperlordosis could be noted. Neurological assessment showed an unsteady gait with bilateral quadriceps weakness (4/5), altered sensation below the belly button (T11 level) mainly on both thighs, and enhanced lower extremities reflexes with positive Babinski sign and ankle clonus, but normal bowel and bladder functions.

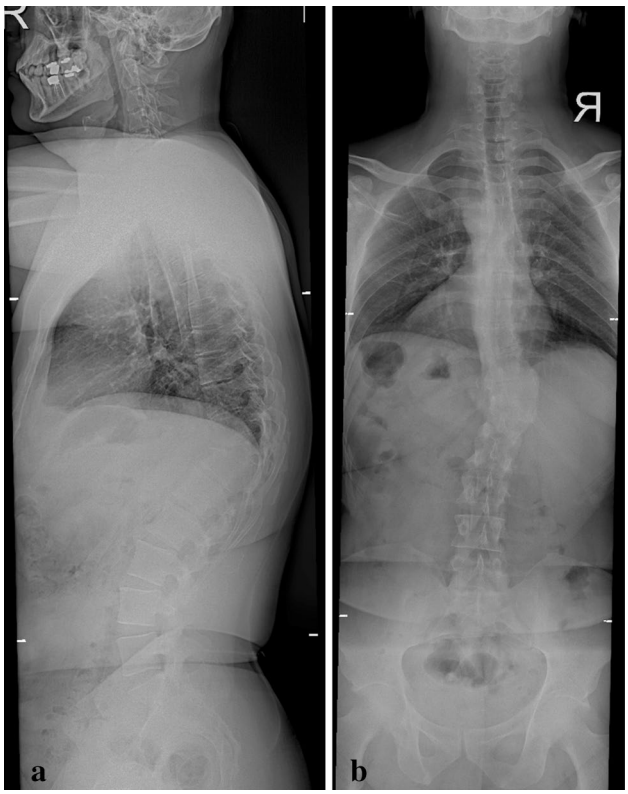

Fig. 1 Full spine anteroposterior and lateral X-rays at the time of presentation revealing a thoracolumbar kyphosis with an angulation of 60° between T10 and T12, with compensatory hyperlordosis of 71°, thoracic hypokyphosis of 20 degrees and coronal thoracolumbar deformity of 32 degrees (**a** and **b**)

Full spine anteroposterior and lateral X-rays at the time of presentation revealed a pelvic incidence of 41°, a sacral slope of 31°, a thoracolumbar kyphosis with an angulation of 60° between T10 and T12, with compensatory lumbar hyperlordosis of 71° (including an L4S1 angle of 34°), a thoracic hypokyphosis of 20°, and a global tilt angle [11] of 10°, also a short thoracolumbar scoliosis of 32 degrees was seen (Fig. 1a, b). Preoperative Global Alignment and Proportion (GAP) score [12] was calculated based on the previously mentioned spinopelvic parameters, it resulted in a total score of 4 which corresponds to a moderately disproportioned sagittal spinopelvic state (Table 1). CT scan confirmed the deformity (Fig. 2a, b) and revealed its congenital nature with the presence of a butterfly vertebra at the level of T11, where failure of fusion of the two chondrification centers of the vertebral body with anterior and median aplasia could be seen (Figs. 2c, 3a, b), with posterior arch fusion between T10 and T12 (Fig. 3c). MRI showed cord compression at the apex of the kyphosis associated to syringomyelia (Fig. 4a, b) which was an incidental finding rather than the primary pathology, and no Chiari malformation could be detected.

Given the presented deformity with neurological impairment, the patient underwent a posterior resection of the butterfly vertebra (T11) with instrumentation from T8 to L2.

Surgical technique

The patient was installed in a prone position, on four cushions (Fig. 5a). During the surgery, we used transcranial motor evoked potentials, somatosensory evoked potentials, and free running electromyography (EMG) of the lower extremities as well as evoked EMGs with pedicle screw stimulation.

Table 1 Global alignment and proportion (GAP) score based on the spinopelvic sagittal parameters

GAP score				
Target correction goals	Ideal sacral slope 33°	Ideal lumbar lordosis 54°	L4S1/L1S1 50–80%	Ideal global tilt 5°
	Preoperative score		Postoperative score	
Relative pelvic version	0		0	
Relative lumbar lordosis	3		0	
Lordosis distribution index	1		0	
Relative spinopelvic alignment	0		0	
Age factor	0		0	
Total	4		0	
Category	Moderately disproportioned		Proportioned	

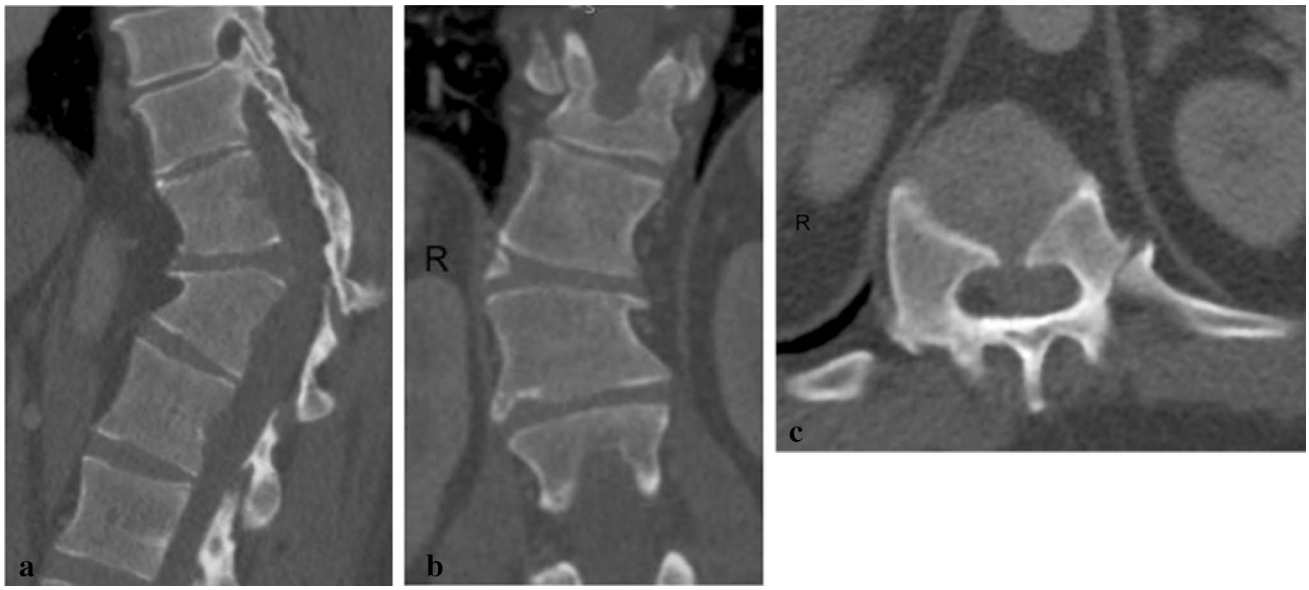

Fig. 2 CT scan confirming the sagittal and coronal deformity (**a** and **b**), and revealing a typical aspect of a butterfly vertebra of T11 on the axial view (**c**)

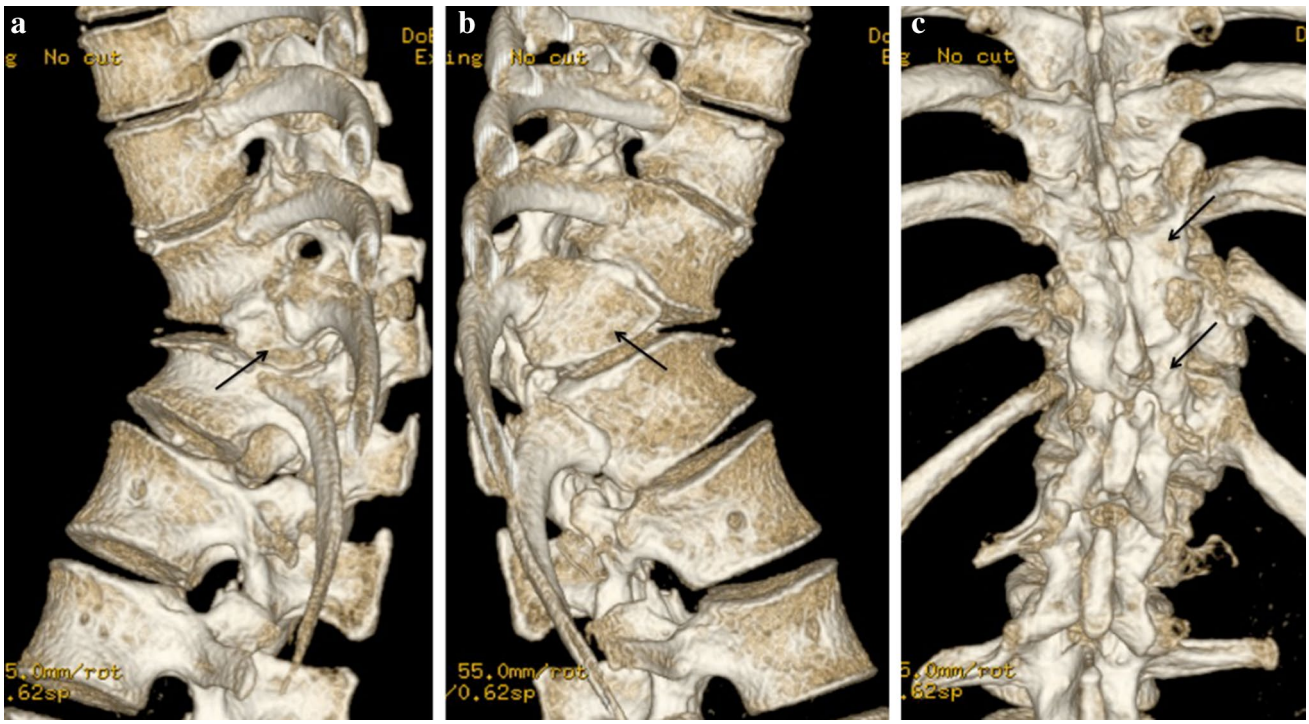

Fig. 3 3D reconstruction detailing the left and right parts of the T11 butterfly vertebra (**a** and **b**, black arrows), and showing a posterior fusion mass between T10 and T12 (**c**, black arrow)

The operative field was exposed from T8 to L2; a posterior cutaneous midline incision was made. The spine was exposed subperiosteally, going laterally to the costotransverse junction. Resection of the inferior articular processes at all levels was performed bilaterally to provide maximum flexibility to the spine. We next used the free-hand technique to place the pedicle screws from T8 to T10 and from T12 to L2, but left T12 pedicle was congenitally absent; therefore, no screw was inserted (Fig. 5b), two sublaminar offset hooks were put at the distal level of the construct to increase the

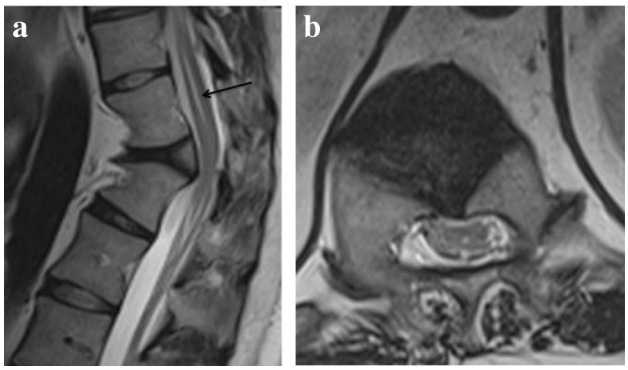

Fig. 4 MRI T2-weighted images revealing cord compression at the apex of the kyphosis associated to syringomyelia (**a**, black arrow and **b**)

resistance to pull out forces. To expose the lateral wall of T11, the transverse processes were removed with a rongeur, and the proximal 3 cm of the right and left ribs (including the rib head) were removed. A cobb elevator was than placed on the lateral wall of the vertebra, bilaterally, and moved anteriorly to the anterolateral quadrant, to retract all the lateral soft tissues, a complete surgicel was left in place to maintain a safe distance between the bone and the soft tissues. Two complete foraminotomies both cephalad and caudad to the T11 pedicles on both sides were made, completed by complete laminectomy of the concerned level, with partial laminectomies of the levels just above and just below, this enabled surrounding of the pedicles. Both pedicles were than removed exposing the posterior walls of each part of the butterfly vertebra, and the nerve roots above and below were identified. Both bodies of the butterfly vertebra were then removed with the use of osteotoms and pituitary rongeurs, including the discs above and below, and no retraction of the dural sac as it is prohibited in the thoracic region (Fig. 5c). A small mesh cage filled with bone graft was inserted anteriorly on the left side, between T10 and T12 vertebral edges, to act as a hinge to enable asymmetrical correction in the coronal plane, and maintain vertebral height to avoid spine over-shortening during correction (Fig. 5d). For the correction technique, at first, cantilevering of the spine with two prebended titanium alloy rod connected by a domino on the right side was performed, and then, further closure of the resection site by compression with the use of the domino was applied, the rod was than completely locked (Fig. 5e). The bone on bone contact on the right side was checked, the spinal cord was carefully controlled as kinking could occur in case of important reduction, and the cage showed a strong primary stability. Contralateral rod was placed and secured. The fixation was completed by the placement of one cross-link connector between the two rods. The prepared autologous bone grafts were placed to cover the maximum surface (Fig. 5f). Postoperative improvement of the gibbosity could be seen (Fig. 5g). Operative time was 320 min, and total blood loss was 800 mL.

The patient could walk on day 2, with a Thoracic Lumbar Orthosis, to be kept for 3 months, and with assistance from a physical therapist. He was discharged on Day 10. Postoperative X-ray and regular X-rays later on showed a stable correction of the thoracolumbar kyphosis with angulation of 10° between T10 and T12 with correction of the compensatory hyperlordosis to 48° (including an L4S1 angle of 32°) and hypokyphosis to 44 degrees. Sacral slope was 32° and global tilt 10°. In addition, scoliosis was corrected to 6°. Postoperative GAP score was 0, which corresponds to a sagittally proportioned spinopelvic state (Table 1). He could walk without a cane at 3 months and showed satisfactory clinical and radiological results at 2 years (Figs. 6a, b, 7a, b).

Discussion

Butterfly vertebra is considered a benign spinal anomaly and its discovery is usually coincidental in patients evaluated for routine back ailments. It is frequently asymptomatic, but it may be the cause of back pain due to alteration of spine biomechanics. Sciatica may also be part of its presentation when disc herniation from the sagittal cleft occurs [3].

This congenital condition can be confused with a pathological fracture in the lateral view due to wedging [13], it may also be confused with infection or metastases [1], but MRI would rule out these differential diagnoses, as it does not show any altered signal intensity or soft tissue enhancement, which usually occur in these pathologies. Computerized tomography scan, with the help of 3D reconstruction, typically shows the split vertebra with two lateral halves.

Butterfly vertebra may be associated with an anterior column defect of its two lateral halves, this anterior aplasia leads to kyphotic deformity, and depending on the asymmetry between the 2 halves, in terms of size, scoliotic deformity component may also come into play [4]. As described in the literature, congenital kyphosis (or kyphoscoliosis) is a much less common deformity than pure congenital scoliosis [14], and previous efforts to control the former by physiotherapy and bracing have been ineffective [15]. Therefore, surgical management is recommended, though challenging, consisting traditionally of a combined anterior and posterior approaches in one or two stage procedures [16], but during the past decade, the use of a single-stage posterior approach in combination with vertebral osteotomies became very popular, avoiding the morbidity of an additional anterior approach, with satisfying correction results [17].

Few reports in the literature investigated the surgical outcome of congenital thoracolumbar kyphosis, focusing mainly on the pediatric and adolescent population. In a series of 10 patients treated by a single posterior approach

Fig. 5 Intra-operative images showing the thoracolumbar hump (**a**), spinal exposure and screws insertion from T8 to L2 but sparing T11 and left T12 (**b**), resection of the T11 butterfly vertebra (**c**, black arrows), insertion of the cage on the left side (**d**, black arrow), reduction of the kyphosis with placement of a domino on the right side (**e**), placement of autologous bone graft (**f**), clinical aspect at the end of the procedure (**g**)

and using either pedicle subtraction osteotomy or vertebral column resection as a correction technique, Spiro [17] showed an improvement of the thoracolumbar kyphosis from 60° preoperatively to 17.5° postoperatively with no neurological complications, which is similar to the results of our case report. In another case series by Shi [18], 38 congenital thoracolumbar kyphosis patients were all surgically managed by SRS-Schwab grade 4 osteotomy, kyphosis improved from 49.5° to 6.8°, mean operative time was 242 min and mean blood loss was 634 mL.

In addition, only few case reports describing congenital bony spinal anomalies treated in adulthood could be retrieved. Polly [19] reported two adult patients (24 and 42-years old) with thoracic kyphoscoliosis due to fully segmented thoracic hemivertebra, they were surgically treated by posterior transpedicular lateral extracavitary excision and spinal instrumentation with satisfactory results and no neurological complications. Another case report by Ansari [20] described a 31-years old patient presenting with a waddling gait, legs numbness and urinary incontinence in relation to

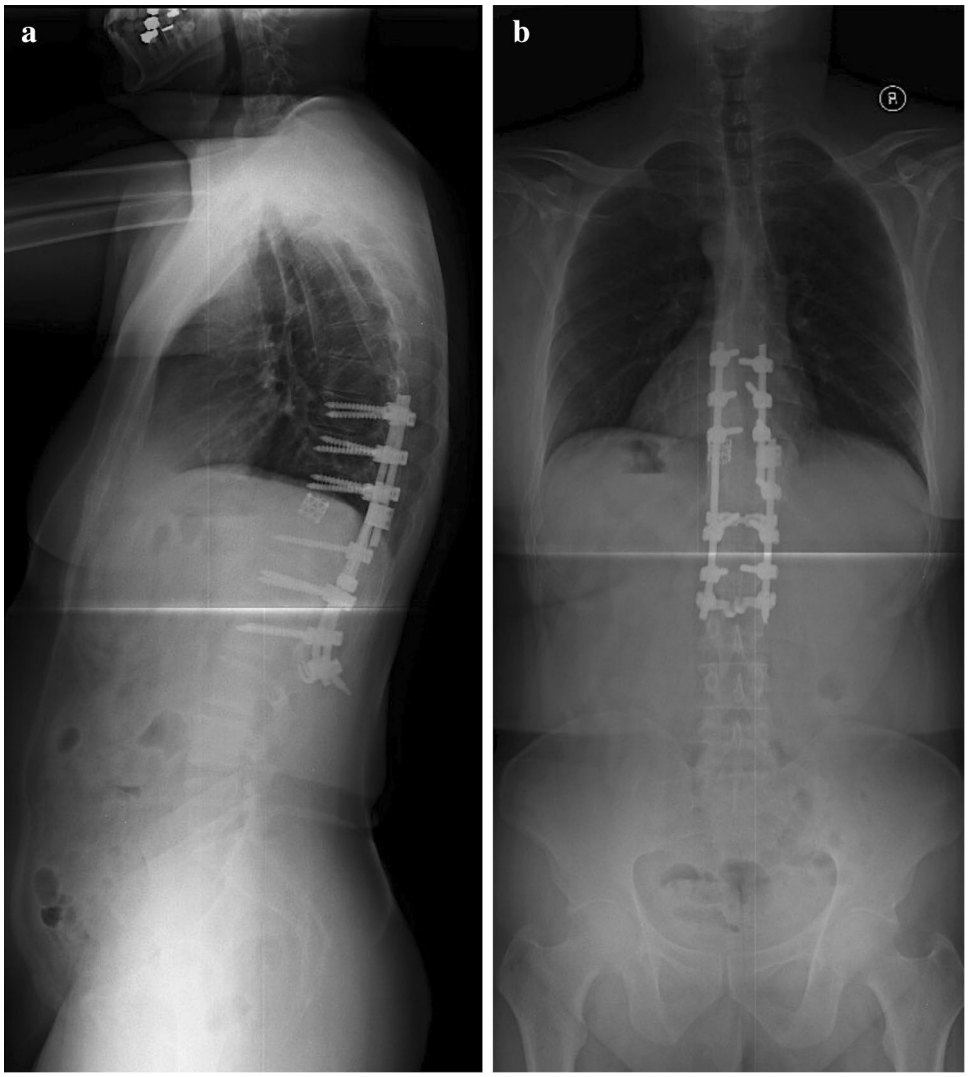

Fig. 6 Two-year full spine X-rays showing stable correction of the thoracolumbar kyphosis with angulation of 10° between T10 and T12, correction of the compensatory hyperlordosis to 48° and hypokyphosis to 44°. In addition, coronal deformity was corrected to 6°

a dorsal midline hemivertebra causing kyphosis at the lumbosacral junction, he underwent resection of the hemivertebra with posterolateral instrumented fusion from L2 to the pelvis with no deformity correction but significant clinical improvement. In addition, Ruf [21] described a 42-years old patient presenting with head coronal malalignment who underwent C2 hemivertebra resection by combined anterior (transoral) and posterior approaches with good clinical and radiological outcome.

A case in which an isolated butterfly vertebra caused symptomatic kyphoscoliosis at the thoracolumbar junction with neurological impairment has, to our knowledge, never been reported. In fact, few cases reported the association of butterfly vertebra with spinal deformity and discussed its surgical management. Cui et al. [9] suggested, in a case of lumbosacral butterfly vertebra causing lumbar scoliosis and spondylolisthesis, a single-stage partial vertebra resection, interpedicular graft with a cage, and instrumentation via a posterior approach, to preserve the two distal concave nerve roots and correct the deformity. In addition, in a previously operated patient for L2 butterfly vertebra with secondary kyphosis, Zhan et al. [10] described the use of a pedicle subtraction osteotomy at the level of the butterfly vertebra combined to multiple levels Ponte osteotomies, to correct the sagittal deformity. The aforementioned cases are summarized in Table 2.

The current report described a case of thoracolumbar kyphoscoliosis secondary to a butterfly vertebra of T11 causing progressive neurological impairment. Complete resection of the congenital anomaly, given its sharp angulation, with an asymmetrical coronal correction by the insertion of a mesh cage on the concave side, and a domino on the convex side was performed. It was decided to go 3 levels down, as the sagittal stable vertebra concept [22] was used to prevent distal junctional kyphosis; therefore, the most proximal lumbar vertebra body touched by the vertical line from the posterior—superior corner of the sacrum, which was L2, was included. In addition, because of the sharp and

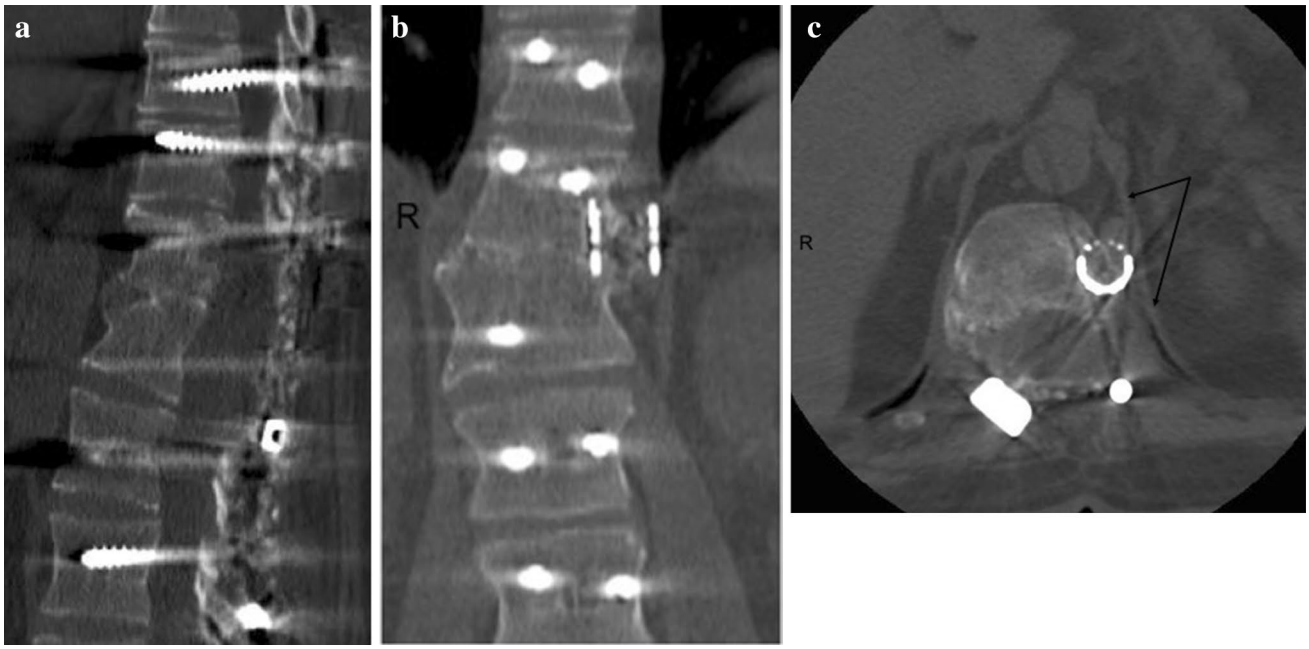

Fig. 7 Two-year postoperative CT scan confirming the complete fusion and a stable correction in both planes at the level of the resection (**a** and **b**). In addition, the axial image (**c**) shows how the lateral part of the cage lies in the fatty area between the vertebral body and the left diaphragmatic crus (postero-medially to the latter, black arrows), with no proximity to any vascular or neurological structures

Table 2 Previous reports of butterfly vertebra associated to spinal deformity with surgical management

References	Age (years)	Gender	Level	Previous surgery	Symptoms	Deformity	Cobb angle	Type of surgery
[9]	13	Female	L6	No	Left sciatica	Scoliosis Spondylolisthesis	30°	Ponte osteotomies concave side Partial resection of the butterfly vertebra Interpedicular cage L5S1
[10]	34	Male	L2	L1L3 fixation at age 19	Back pain Leg pain	kyphosis	57°	Pedicle subtraction osteotomy of the butterfly vertebra
Current study	39	Male	T11	No	Back pain Bilateral radicular pain Unsteady gait	Kyphosis Scoliosis	60° 32°	Complete resection of the butterfly vertebra Concave interbody cage T10T12

No postoperative complications were reported at final follow-up for any of the cases

stiff deformity with vertebral resection, and the long lever arm required for correction, fixation was extended 3 levels up. Two sublaminar offset hooks were put at the distal level of the construct as biomechanical studies showed that a pedicle screw combined with an infra-laminar hook offers greater fixation strength (resistance to loosening tests) and a stiffer construct, when compared with fixation by pedicle screws alone [23]. In addition, supplemental offset sublaminar hooks significantly increase the construct stiffness without sacrificing an additional level distally, and adsorb some part of the construct strain, thereby reducing pedicle screw bending moments [24]. The main disadvantage of sublaminar hooks is the canal intrusion on both sides, nevertheless in our case, the canal was wide at the L2 level which enabled smooth insertion of the hooks with no neurological compromise. Another disadvantage is the possibility of hook migration into the canal, which generally occurs in case there is not a tight fit of the hook throat to the lamina;

such complication can be avoided by selecting a proper hook size and by keeping a hook holder on the hook during manipulation such as rod insertion [25]. The titanium mesh cage was intended to be placed on the edges of the vertebral endplates of T10 and T12 to be as asymmetrical as possible for the coronal correction, nevertheless the inserted cage was 15 mm in diameter and the ideal width would have been 10 mm which was unfortunately not an available size at the time of the surgery, this is why it overstepped the edges laterally by 5 mm; nonetheless, axial CT image (Fig. 7c) showed that its lateral part lies in the fatty area between the vertebral body and the left diaphragmatic crus (posteromedially to the latter), with no proximity to any vascular or neurological structures.

In summary, complete resection of the butterfly vertebra in combination to asymmetrical coronal correction, was effective for the treatment of this rare pathology in both planes, with a favorable long-term outcome.

Author contributions Anouar Bourghli conception, drafting, and final approval; Saleem Abduljawwad data analysis, drafting and final approval; Louis Boissière interpretation of data, critical revision, and final approval; Ibrahim Obeid design, critical revision, and final approval.

Funding No funds were received for the current study.

Compliance with ethical standards

Conflict of interest AB (none), SA (none), LB (none), and IO (other from Alphatec, outside the submitted work).

Informed consent Permission has been granted from the patient to report the results of this case.

References

1. Patinharayil G, Han CW, Marthya A, Meethall KC, Surendran S, Rudrappa GH (2008) Butterfly vertebra: an uncommon congenital spinal anomaly. Spine 33:E926–E928. https://doi.org/10.1097/brs.0b013e31818ad3e1 **(Phila Pa 1976)**
2. Katsuura Y, Kim HJ (2019) Butterfly vertebrae: a systematic review of the literature and analysis. Glob Spine J 9:666–679. https://doi.org/10.1177/2192568218801016
3. Cho HL, Kim JS, Paeng SS, Lee SH (2011) Butterfly vertebra with lumbar intervertebral disc herniation. J Neurosurg Spine 15:567–570. https://doi.org/10.3171/2011.6.SPINE1178
4. McMaster MJ, Singh H (1999) Natural history of congenital kyphosis and kyphoscoliosis. A study of one hundred and twelve patients. J Bone Jt Surg Am 81:1367–1383. https://doi.org/10.2106/00004623-199910000-00002
5. Ekim A (2010) Butterfly vertebra anomaly: a case report. J Back Musculoskelet Rehabil 23:161–164. https://doi.org/10.3233/BMR-2010-0262
6. Kapetanakis S, Giovannopoulou E, Nastoulis E, Demetriou T (2016) Butterfly vertebra. A case report and a short review of the literature. Folia Morphol (Warsz) 75:117–121. https://doi.org/10.5603/FM.a2015.0066
7. Sonel B, Yalcin P, Ozturk EA, Bokesoy I (2001) Butterfly vertebra: a case report. Clin Imaging 25:206–208
8. Slouma M, Cheour E, Sahli H, Elleuch M (2016) Congenital butterfly vertebra. Acta Med Port 29:491. https://doi.org/10.20344/amp.6634
9. Cui G, Watanabe K, Ishii K, Toyama Y, Chiba K, Matsumoto M (2011) Interpedicular graft using a titanium mesh cage in a patient with lumbar scoliosis associated with a congenital butterfly vertebra. J Neurosurg Spine 14:215–218. https://doi.org/10.3171/2010.10.SPINE10191
10. Zhan J, Wang Y, Chen B, Lin D (2018) Revision surgery after instrumental fixation in patient with butterfly vertebra: a case report. Acta Neurol Belg. https://doi.org/10.1007/s13760-018-1021-x
11. Obeid I, Boissiere L, Yilgor C, Larrieu D, Pellise F, Alanay A, Acaroglu E, Perez-Grueso FJ, Kleinstuck F, Vital JM, Bourghli A, European Spine Study Group E (2016) Global tilt: a single parameter incorporating spinal and pelvic sagittal parameters and least affected by patient positioning. Eur Spine J 25:3644–3649. https://doi.org/10.1007/s00586-016-4649-3
12. Yilgor C, Sogunmez N, Boissiere L, Yavuz Y, Obeid I, Kleinstuck F, Perez-Grueso FJS, Acaroglu E, Haddad S, Mannion AF, Pellise F, Alanay A, European Spine Study G (2017) Global alignment and proportion (GAP) score: development and validation of a new method of analyzing spinopelvic alignment to predict mechanical complications after adult spinal deformity surgery. J Bone Jt Surg Am 99:1661–1672. https://doi.org/10.2106/JBJS.16.01594
13. Karargyris O, Lampropoulou-Adamidou K, Morassi LG, Stathopoulos IP, Chatziioannou SN, Pneumaticos SG (2015) Differentiating between traumatic pathology and congenital variant: a case report of butterfly vertebra. Clin Orthop Surg 7:406–409. https://doi.org/10.4055/cios.2015.7.3.406
14. Marks DS, Qaimkhani SA (2009) The natural history of congenital scoliosis and kyphosis. Spine 34:1751–1755. https://doi.org/10.1097/brs.0b013e3181af1caf **(Phila Pa 1976)**
15. Winter RB, Moe JH, Wang JF (1973) Congenital kyphosis. Its natural history and treatment as observed in a study of one hundred and thirty patients. J Bone Jt Surg Am 55:223–256
16. Leatherman KD, Dickson RA (1979) Two-stage corrective surgery for congenital deformities of the spine. J Bone Jt Surg Br 61-B:324–328
17. Spiro AS, Rupprecht M, Stenger P, Hoffman M, Kunkel P, Kolb JP, Rueger JM, Stuecker R (2013) Surgical treatment of severe congenital thoracolumbar kyphosis through a single posterior approach. Bone Jt J 95-B:1527–1532. https://doi.org/10.1302/0301-620x.95b11.31376
18. Shi B, Zhao Q, Xu L, Liu Z, Sun X, Zhu Z, Qiu Y (2018) SRS-schwab grade 4 osteotomy for congenital thoracolumbar kyphosis: a minimum of 2 years follow-up study. Spine J 18:2059–2064. https://doi.org/10.1016/j.spinee.2018.04.011
19. Polly DW Jr, Rosner MK, Monacci W, Moquin RR (2003) Thoracic hemivertebra excision in adults via a posterior-only approach. Report of two cases. Neurosurg Focus 14:e9
20. Ansari SF, Rodgers RB, Fulkerson DH (2015) Dorsal midline hemivertebra at the lumbosacral junction: report of 2 cases. J Neurosurg Spine 22:84–89. https://doi.org/10.3171/2014.9.SPINE1411
21. Ruf M, Welk T, Merk HR, Smiszek FG, Pitzen T (2015) Resection of a hemivertebra within the craniocervical junction. Spine 40:E1191–E1194. https://doi.org/10.1097/brs.0000000000001030 **(Phila Pa 1976)**
22. Cho KJ, Lenke LG, Bridwell KH, Kamiya M, Sides B (2009) Selection of the optimal distal fusion level in posterior instrumentation and fusion for thoracic hyperkyphosis: the sagittal stable

vertebra concept. Spine 34:765–770. https://doi.org/10.1097/brs.0b013e31819e28ed **(Phila Pa 1976)**
23. Murakami H, Tsai KJ, Attallah-Wasif ES, Yamazaki K, Shimamura T, Hutton WC (2006) A biomechanical assessment of infralaminar hooks as an alternative to supra-laminar hooks in thoracolumbar fixation. Spine 31:967–971. https://doi.org/10.1097/01.brs.0000214932.49027.6a **(Phila Pa 1976)**
24. Chiba M, McLain RF, Yerby SA, Moseley TA, Smith TS, Benson DR (1996) Short-segment pedicle instrumentation. Biomechanical analysis of supplemental hook fixation. Spine 21:288–294. https://doi.org/10.1097/00007632-199602010-00006 **(Phila Pa 1976)**
25. Ondra SLMTJ (2006) Posterior thoracic and lumbar hook-rod instrumentation. In: Dickman CA (ed) Spinal cord and spinal column tumors: principles and practice. Thieme, New York