Romanian Neurosurgery | Volume XXXI | Number 1 | 2017 | January - March

Article

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ROMANIA

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DOI: 10.1515/romneu-2017-0012

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Butterfly vertebra: a case report

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Abstract: *Background and importance*: Butterfly vertebra is an uncommon congenital spinal anomaly, which can be easily mistaken for vertebral fracture, infection or tumor. *Clinical presentation*: We report the case of a 20-year-old male patient with a 3-year history of intermittent mid-thoracic spinal pain. Local examination showed a mild thoracic kyphoscoliosis. Neurological examination was normal. Computer tomography revealed the presence of a T6 butterfly vertebra associated with morphological anomalies in adjacent vertebral bodies and T6-T9 Schmorl's nodes. MRI scan confirmed the diagnosis and additionally showed a mild spinal stenosis caused by apparent posterior epidural fat hypertrophy. The patient was treated with painkillers and physical therapy. *Conclusion*: Butterfly vertebra is a benign condition. Once diagnosed, additional diagnostic procedures are not necessary. Neurosurgeons must be aware of this congenital anomaly that should not be confused with a vertebral fracture.
Key words: butterfly vertebra, congenital vertebral anomaly

Background and importance

Back pain is a common complaint, mainly caused by degenerative spinal disease. Thoracic spine pain is less frequently encountered in comparison with the lumbar or cervical regions. Congenital spinal anomaly at this level is a rare cause pain.

Butterfly vertebra, which consists of a sagittal cleft in the vertebral body, is a congenital malformation that gives vertebrae the particular appearance of butterfly wings on anterior-posterior view of imaging studies.

We report the case of a patient diagnosed with this rare spinal anomaly and perform a literature review concerning this condition.

Clinical presentation

A 20-year old male patient was admitted to our Neurosurgical Department, complaining of intermittent mid-thoracic spinal pain, particularly while maintaining a sitting position for prolonged periods of time. Onset of symptoms occurred 3 years before presentation, with progressive worsening and alleviation after bed rest and occasional over-the-counter pain medication.

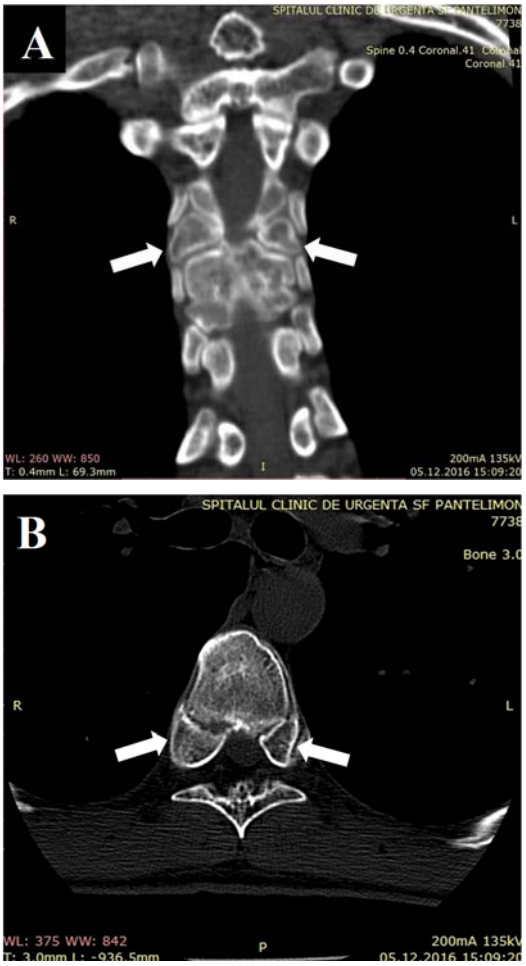
Medical history was unremarkable. The patient mentioned a pulmonary condition at birth and a restrictive pulmonary syndrome diagnosed in his teens, but the he could not supply any medical documents for confirmation. Taking into consideration his particularly considerable height (208 cm), he had previously undergone basic endocrinological testing which was normal. He denied any history of trauma and no other probable cause of symptoms could be identified.

Local examination revealed a mild thoracic kyphoscoliosis. No abnormalities were detected after a thorough neurological examination. Chest X-ray and routine blood tests were normal.

CT scan of the thoracic spine revealed a congenital anomaly of the T6 vertebral body with a butterfly vertebra appearance (Figure 1.A). The vertebra appeared incompletely developed with a hypoplastic aspect, only the posterior third of the vertebral body being visible, which consisted of two unjoined halves (Figure 1.B); the posterior vertebral arch was normal. The thoracic kyphosis was augmented at this level. Other encountered anomalies included morphological changes of adjacent vertebral bodies with a T5-T7 incomplete vertebral block tendency (Figure 1.C) and Schmorl nodes at T7-T9 levels.

MRI of the thoracic spine confirmed the diagnosis of T6 butterfly vertebra (Figures 2.A, 2.B, 2.C). Additionally, it showed a mild spinal stenosis at this level caused by the apparent hypertrophy of posterior epidural fat tissue with no intramedullary signal changes. No thoracic disc protrusions or herniations were observed (Figure 2D).

The patient was recommended pain medication and physical therapy with good outcome at follow-up.



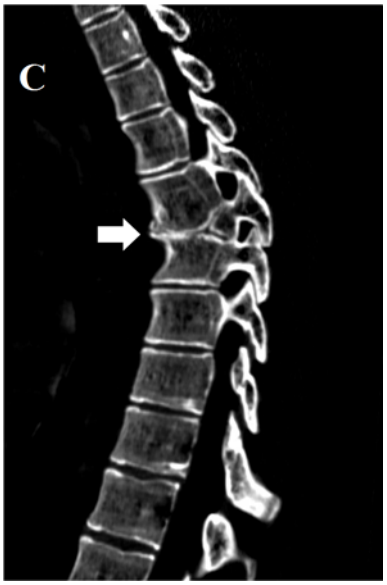
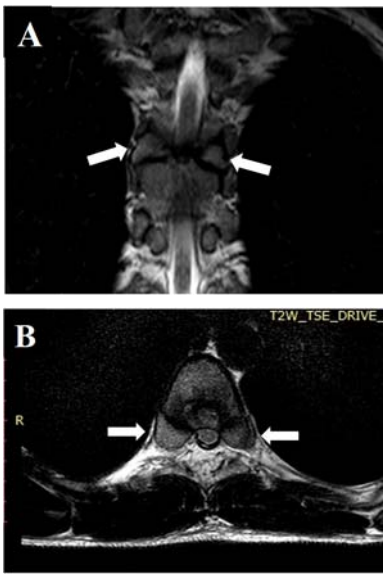


Figure 1 - CT scan of thoracic spine revealing T6 butterfly vertebra
(A) coronal view showing the characteristic two-winged aspect (white arrows) of butterfly vertebra. (B): axial slice at T6 level displaying the two unjoined hemivertebrae (white arrows). (C): sagittal view showing the augmented kyphosis at T6 level and T5-T7 incomplete vertebral block aspect (white arrow)



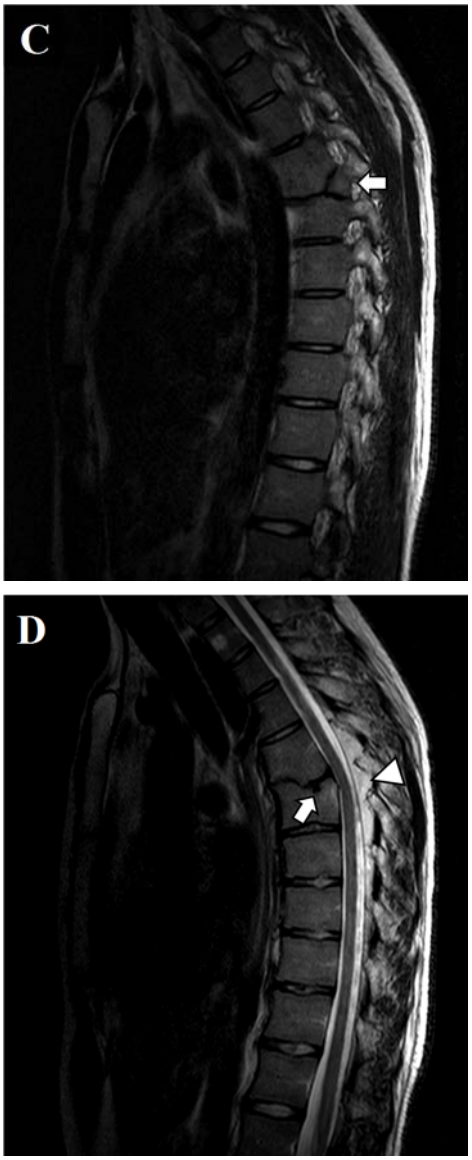


Figure 2 - T2-weighted MRI scan of thoracic spine with T6 butterfly vertebra
Coronal (A), axial (B) and sagittal (C) T6-level centered images, confirming specific butterfly vertebra aspect. (D): Sagittal slice revealing mild spinal canal stenosis at T6 level caused by posterior epidural fat hypertrophy (arrow-head); multiple Schmorl nodes can be seen (the largest at the superior T7 endplate - arrow)

Discussion

Rokitansky first described butterfly vertebra in 1844, located at the 12th thoracic vertebra1. This is a congenital anomaly of the spine, which has received various names: sagittal cleft vertebra, anterior rachischisis, anterior spina bifida and anterior somatoschisis. Since its first description, very few cases of this malformation have been reported.

Butterfly vertebra develops between the 3rd and 6th gestational week, when the two chondrification centers fail to fuse while giving rise to the vertebral body. These centers remain separated by notochordal tissue, which normally regresses from the level of the vertebral body and develops into the nucleus pulposus of the intervertebral disc. Thus, the vertebral body is comprised of two hemivertebrae and the notochordal derived tissue between them. This explains the anteroposterior X-ray appearance of two hemivertebrae separated by an empty space or cleft, more or less large.

This malformation can be associated with other spinal anomalies, such as lumbosacral transitional vertebrae, spina bifida, diastematomyelia and several congenital syndromes: Allagille syndrome, Jarcho-Levin syndrome, Klippel-Feil syndrome or with gastrointestinal and genitourinary anomalies (2, 3).

The etiology is supposed to be genetic (deletions of chromosome 20) (4) or a congenital deficiency of vascularization (5).

Butterfly vertebra is usually asymptomatic. Patients with this malformation are diagnosed incidentally or may present atypical chronic pain.

This condition can be diagnosed prenatally with 3D transabdominal ultrasound images6 or after birth with plain X-ray or bone window computer tomography. On plain radiography, butterfly vertebra appears wedge-shaped in lateral view and can be easily misdiagnosed as a compression fracture. Anteroposterior spine X-ray reveals the vertebral body split in two halves. MRI shows the particular butterfly vertebra aspect on sagittal, axial and coronal images, other possibly associated congenital anomalies or intervertebral disc prolapses.

The butterfly vertebra malformation has been described not only in clinical cases, but also in dry bone specimens from archeological excavations of necropolises (7, 8).

The differential diagnosis of butterfly vertebra can be made with a compression fracture of the vertebral body, either osteoporotic or pathologic in nature. This is the reason why a DEXA test or laboratory investigations are sometimes necessary in order to rule out osteoporosis, neoplasia or infection.

There have been reports of butterfly vertebra cases associated with disc protrusion or herniation of nucleus pulposus at the abnormal vertebral body level (5, 9–11).

Patients with Alagille syndrome present atresia/hypoplasia of the intrahepatic biliary tree, chronic cholestasis, peculiar facies (hypertelorism, sunken eyes, straight nose, pointed chin, abnormal dental implantation), pulmonary stenosis and skeletal abnormalities (butterfly vertebra, phalangeal or ulnar

shortening, pelvic abnormalities, osteoporosis) (2).

Clinicopathological spectrum of Jarcho-Levin syndrome is composed of a short neck and trunk, constricted thorax, rib and vertebral anomalies (butterfly vertebrae, hemivertebrae, fused or hypoplastic vertebrae) (3).

When a butterfly vertebra is detected prenatally in association with other anomalies, it can usually lead to the termination of pregnancy6.In the case of postnatal diagnosis in children, only follow-up is necessary in order to exclude potentially associated rare syndromes.

This condition is a benign spinal anomaly which must be differentiated from other pathological conditions. In adult patients with butterfly vertebra, physical therapy is recommended to achieve unrestricted and pain-free movements. Once the diagnosis of butterfly vertebra has been established, patients must be informed that it represents a benign anomaly and further investigation procedures can be avoided.

Conclusion

Butterfly vertebra is a rare, benign condition. Once diagnosed, additional diagnostic procedures are not necessary. Neurosurgeons must be aware of this congenital anomaly, that should not be confused with a vertebral fracture.

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