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CASE REPORT

Bezold–Jarisch reflex causing bradycardia and hypotension in a case of severe dystrophic cervical kyphotic deformity: a case report and review of literature

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Abstract

Purpose A 17-year-old adolescent with neurofibromatosis and severe cervicothoracic deformity was identified to have thoracic inlet compression leading to bradycardia and hypotension, only during prone positioning, and we discuss its successful management.

Methods Preoperative halo-gravity traction reduced the deformity from 126° to 91°. During prone positioning, sudden onset bradycardia was followed by asystole, which disappeared immediately on turning over to supine position. Surgery was called off after two additional failed attempts of prone positioning.

Results A retrospective analysis of CT and MRI showed severe narrowing of the thoracic inlet. In this patient, the right thoracic inlet was severely narrow, and prone positioning caused a further dynamic compromise stimulating right vagal nerve. The right vagus supplies the sinoatrial node, which is the natural pacemaker of the heart, and its stimulation causes sympathetic inhibition. Bezold–Jarisch reflex is a cardio-inhibitory reflex occurring due to vagal stimulation resulting in sudden bradycardia, asystole, and hypotension. To facilitate prone positioning, the medial end of the clavicles, along with limited manubrium excision, was performed relieving the vagal compression. C2–T4 instrumented decompression followed by anterior reconstruction and cervical plating was performed. The postoperative period was uneventful, and the final deformity was 45°.

Conclusion Bezold–Jarisch Reflex as a result of narrow thoracic inlet caused by cervical kyphosis and compensatory hyperlordosis of the upper thoracic spine has never been reported. This case highlights the need to introspect into thoracic inlet morphology in severe cervicothoracic deformities. Thoracic inlet decompression is an efficient way of addressing this unique complication.

Keywords Neurofibromatosis · Cervical kyphosis · Narrow thoracic inlet · Bradycardia · Bezold–Jarisch reflex

Case report

A 17-year-old adolescent boy with neurofibromatosis and a gradually progressive cervical kyphotic deformity for ten years presented to us with the sudden progression of deformity associated with recent-onset clumsiness of hands, loss of dexterity, gait instability and difficulty in performing day-to-day activities over past six months. On examination, he had multiple Cafe au lait spots over the trunk with a severe rigid cervical kyphotic deformity and compensatory thoracic hyperlordosis. Neurologically, weakness of left C6 and C7 roots (MRC grade −4/5) and exaggerated deep tendon reflexes were noted. The plain radiographic image revealed a severe dystrophic cervical kyphotic deformity of 126° (Fig. 1c). CT showed C5–C6 and C6–C7 facet joint subluxation with buckling collapse of the cervical spine with the apex at the C5–C6 level (Fig. 2). MRI demonstrated stretching of the cord with a thinned-out caliber over the acutely deformed cervical spine. No intraspinal mass lesions were detected. The plan was to do a single-stage 540-degree approach correction comprising anterior release (corpectomy), posterior column shortening, and instrumentation

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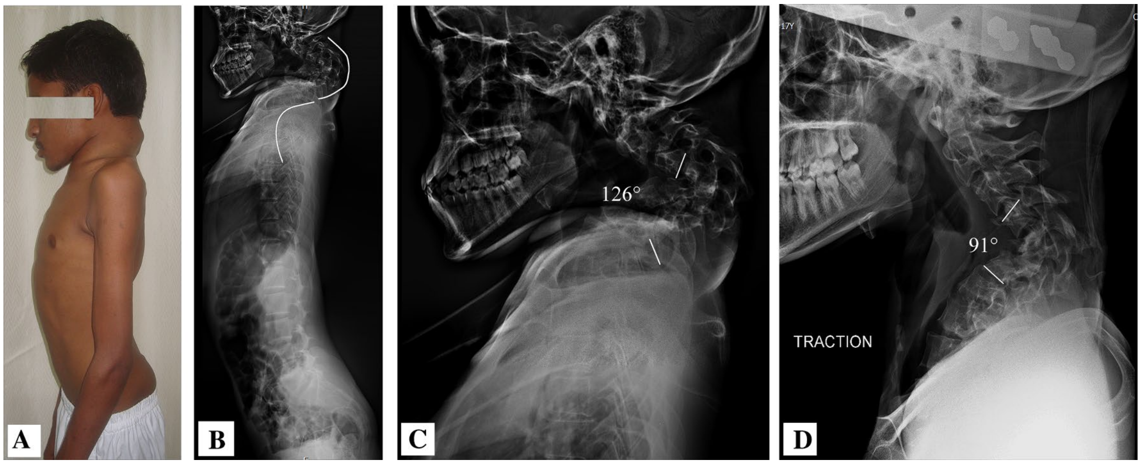

Fig. 1 Preoperative clinical and radiograph images. **a** Clinical image and **b** whole spine standing lateral image of the 17-year-old adolescent with NF showing severe cervical kyphotic deformity and compensatory thoracic hyperlordosis, **c** and **d** focused films of the cervical spine showing a deformity of 126° getting corrected to 91° following four weeks of halo-gravity traction

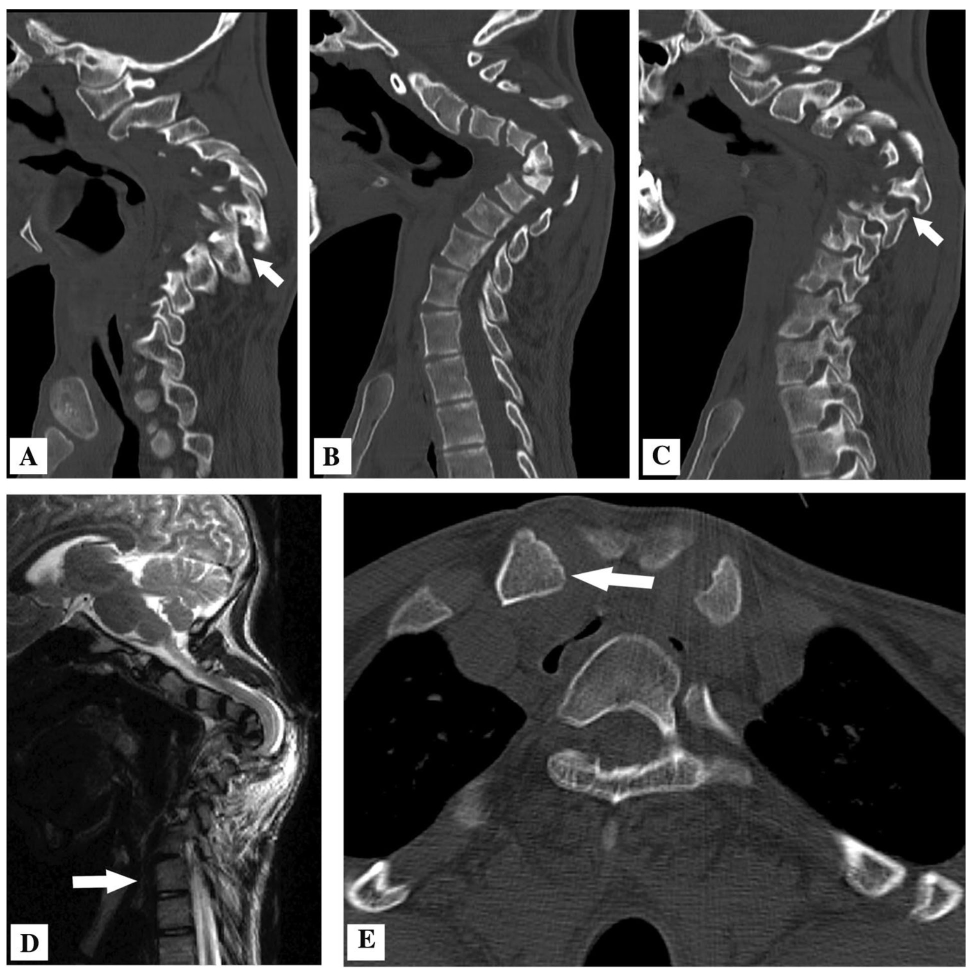

Fig. 2 Preoperative MRI and CT images. Sagittal reformatted **a** right parasagittal, **b** midsagittal, and **c** left parasagittal CT images showing C5–C6 and C6–C7 facet joint subluxation with buckling collapse of the cervical spine with the apex at the C5–C6 level. **d** MRI showing severe narrowing at the thoracic inlet (white arrow), causing soft tissue compression. **e** Axial CT images demonstrate the deviation of the trachea to the right side due to the anteriorly pushed vertebral body secondary to compensatory hyperlordosis, and the arrow mark points to the right sternal end of clavicle penetrating into the narrow thoracic inlet, which on prone positioning stimulates the vagus nerve causing Bezold–Jarisch reflex

followed by anterior reconstruction. Preoperative ambulant halo-gravity traction (HGT) reduced the deformity to 91° with a 35° curve correction (Fig. 1d). After C5, C6, and C7 corpectomy, during prone positioning, sudden onset bradycardia and hypotension were followed by asystole, which disappeared immediately on turning over to supine position.

With no detectable scientific reasons behind this phenomenon, surgery was called off after two additional failed attempts of prone positioning. A retrospective analysis of CT and MRI showed severe narrowing of the thoracic inlet (Fig.2d, e), which led to the diagnosis of dynamic thoracic inlet compromise 48 h following the initial surgery.

Discussion

Neurofibromatosis (NF) is associated with severe acute and rigid dystrophic deformities of the spine and can have serious anesthetic complications [1]. Thoracic inlet or outlet is an opening at the apex of the thoracic cavity bounded by T1 vertebra posteriorly, manubrium anteriorly, and the first rib laterally. The clavicle forms the anterior border of thoracic inlet, and the vital structures which pass through it include trachea, esophagus, thoracic ducts, apices of the lungs, nerves (phrenic, vagus, recurrent laryngeal nerve and sympathetic trunks, vessels (carotid and subclavian arteries, jugular and brachiocephalic veins) [2]. Pitcher et al. realized that in severe pediatric chest wall deformity narrow thoracic inlets cause severe tracheal obstruction and proposed new thoracic inlet index values based on morphometry of thoracic inlet and suggested that a value of > 10 was highly predictive of airway compromise [3]. While narrow thoracic inlets can cause many clinical issues depending on the structure getting compromised, in our case, right vagal nerve got compressed as it passes on the right side of trachea and exactly lies behind the sternoclavicular joint. Fig.2e depicts the narrowed thoracic inlet with sternal end of clavicle protruding inward, just anterior to the space between trachea and esophagus, which might get more compromised during prone positioning. Randall Brenn and his colleagues observed a similar phenomenon in a case of Pompes disease, and they managed it with sympathetic drugs as they initially thought it could be due to bronchospasm [4]. They later concluded that it could be due to tracheal stenosis which, however, cannot explain the cardiac arrest in an intubated patient. The response to epinephrine in their case clearly shows that it was probably due to cardiac inhibition rather than tracheal stenosis.

Bezold–Jarisch reflex is a cardio-inhibitory reflex occurring due to vagal stimulation and results in sudden bradycardia, asystole, and hypotension [5]. In this patient, the right thoracic inlet was severely narrow, and prone positioning caused a further dynamic compromise stimulating right vagal nerve. The right vagus supplies the sinoatrial node, which is the natural pacemaker of the heart, and vagal stimulation causes sympathetic inhibition resulting in sudden parasympathetic overtone. A review article discussing the association of hypotension and bradycardia with Bezold–Jarisch reflex in shoulder arthroscopic surgeries could not establish evidence to support venous pooling as a potential cause as there are equal numbers of contradicting studies toward the hypothesis of venous pooling [6]. It could be possible that hyperabduction in shoulder arthroscopic surgeries may as well stimulate the vagus nerve, causing a similar phenomenon, especially in patients with a narrow thoracic inlet, which needs to be investigated.

This phenomenon has been well established in an experimental animal study where the stimulation of right vagus nerve resulted in bradycardia, slowing of atrioventricular conduction and reduction of atrial contractility. More importantly, the study analyzing the effect of both direct and indirect electrical stimulation of right vagus nerve revealed that maximal bradycardia and decrease in systolic pressure occurred at the level of thoracic inlet [7]. Kim et al. reported dynamic compromise of thoracic inlet during prone positioning in a case of scoliosis resulting in inter-arm arterial pressure difference, which worsened on application of compressive force to the surgical field and got reversed during supine positioning [8]. The end result of this dynamic compromise occurring in prone positioning depends on the anatomical structure being compressed and may vary from patient to patient, and it was right subclavian artery being compressed in their report. In another report, Abcejo et al. observed profound obstructive reversible hypotension due to prone positioning in scoliosis secondary to left atrium and biventricular collapse but reported it secondary to direct chest compression [9]. In patients with chest wall deformities and coexisting scoliosis, similar episodes of hypotension have occurred due to prone positioning and the need for procedures to address the cause of such cardiovascular compromise prior to scoliosis correction has been described [9–11]. Alternatively, a lateral prone position has been used to complete surgical correction of scoliosis, which in our case was not a viable option [12].

After a multidisciplinary team meeting involving anesthetists, otolaryngologists, spine surgeons, and cardiothoracic surgeons, it was decided to attempt surgical decompression of thoracic inlet if prone positioning was not possible during the return to the operating room 72 h later. As expected, the same phenomenon occurred, and the patient had to be turned immediately to the supine position. The medial end of the clavicles, along with limited manubrium excision (Fig. 3a, b), relieved the vagal compression and allowed prone positioning. C2–T4 instrumented decompression followed by anterior reconstruction and cervical plating was performed. The postoperative period was uneventful, and the final Cobbs angle was only 45° (Fig.3e). The patient has recovered completely from his preexisting neurological deficits and has completed 29 months of follow-up without additional complications. This report exemplifies dynamic thoracic inlet compression during prone positioning. It brings out the importance

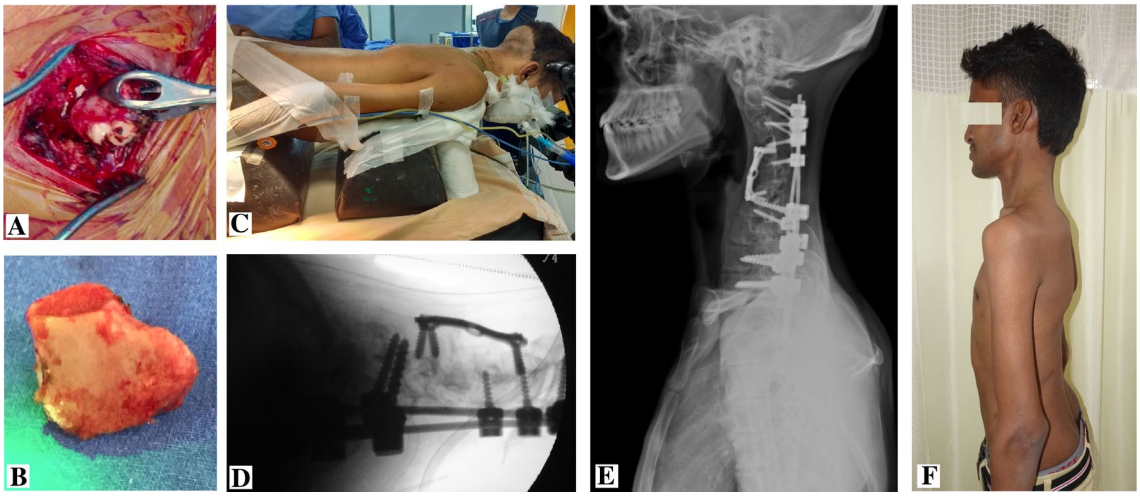

Fig. 3 Intraoperative and postoperative images. **a** The prominent medial end of clavicle which worsens the thoracic inlet compromise being exposed **b** resected medial end of clavicle for thoracic inlet decompression, facilitating **c** prone positioning and completion of 540 approach deformity correction as seen in the **d** intraoperative fluoroscopic image, **e** final postoperative radiograph showing significant deformity correction from 126° to 45° and **f** clinical image taken at 3-year follow-up

of preoperative analysis of thoracic inlet and possible complications in patients with severe cervicothoracic and chest wall deformities, which the spine surgeons and anesthetists need to be aware of and manage appropriately.

Conclusion

Bezold–Jarisch reflex as a result of narrow thoracic inlet caused by rigid cervical kyphosis and compensatory hyperlordosis of the upper thoracic spine has never been reported. Preoperative recognition of narrow thoracic inlet might help surgeons and anesthetists to foresee this rare entity. Thoracic inlet decompression is an efficient way of addressing this unique complication.

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Availability of data and material All details pertaining to the reported case are available with the corresponding author.

Compliance with ethical standards

Conflict of interest All authors declare that there are no conflicts of disclosure.

Consent for publication Appropriate consent has been obtained from the patient concerned.

Ethical approval The study was performed only after approval of the IRB committee.

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