


CASE REPORT

Torticollis, Facial Asymmetry, Local Pain, and Barré-Liéou Syndrome in Connection with One-Sided Ponticulus Posticus: A Case Report and Review of the Literature

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Abstract

Background: Ponticulus posticus (PP) occurs frequently and may cause symptom series, including vertebrobasilar insufficiency, migraine, hearing loss, and Barré-Liéou syndrome. However, few studies to date have described surgical treatment of PP. We report a rare case of a patient who suffered from torticollis, facial asymmetry, localized pain, and Barré-Liéou syndrome in connection with PP. We also review the pertinent literature, focusing on surgical treatment for symptoms due to PP.

Case presentation: A 23-year-old male presented with the chief complaint of continuous significant dizziness to the point of losing consciousness while rotating his head to the right. Plain radiographs and computed tomography (CT) scans of the cervical spine showed a C1 anomaly with the formation of complete PP on the left (dominant) side, with acute-angled, C-shaped kinking of the vertebral artery. Resection of PP via the posterior midline was performed successfully. The patient had satisfactory postoperative relief from localized pain and Barré-Liéou syndrome, but there were no obvious changes in the torticollis and facial asymmetry observed during the 3-month follow-up period.

Conclusions: This case is a rare presentation of torticollis, facial asymmetry, localized pain, and Barré-Liéou syndrome in connection with one-sided complete PP. This tetrad indicates that PP may affect the patient earlier than expected. In such situations, early diagnosis and timely surgical treatment may significantly improve patients' quality of life and avoid the development of torticollis and face asymmetry.

Key words: Barré-Liéou syndrome; case report; ponticulus posticus; tetrad

Background

Ponticulus posticus (PP) is described as an anomalous malformed bony bridge (complete or incomplete) between the superoposterior lateral mass and posterior arch of the atlas^{1,2}. Other synonyms for this bony bridge are the arcuate foramen, Kimmerle anomaly, ponticulus posterior of the atlas, canalis vertebralis, and retroarticular vertebral artery ring²⁻⁴. PP occurs at rates of 1.1%–37% according to different studies^{2,3}. This bony canal usually includes the V3 segment of the vertebral artery (VA) and may also include

the accompanying vein, sympathetic nerves, posterior C1 spinal nerve, and posterior branch of the C1 spinal nerve⁵. Therefore, compression or irritation of these structures by PP with spinal movement may lead to the clinical signs^{5,6}.

The majority of the population with PP have no symptoms; clinical signs (including symptoms of vertebrobasilar insufficiency, migraine, cervicogenic headache, neck and shoulder pain, hearing loss, and Barré-Liéou syndrome) have been reported in only 5.5%–7% of PP patients^{2,7-9}. Until now, few studies have described the surgical treatment of PP,

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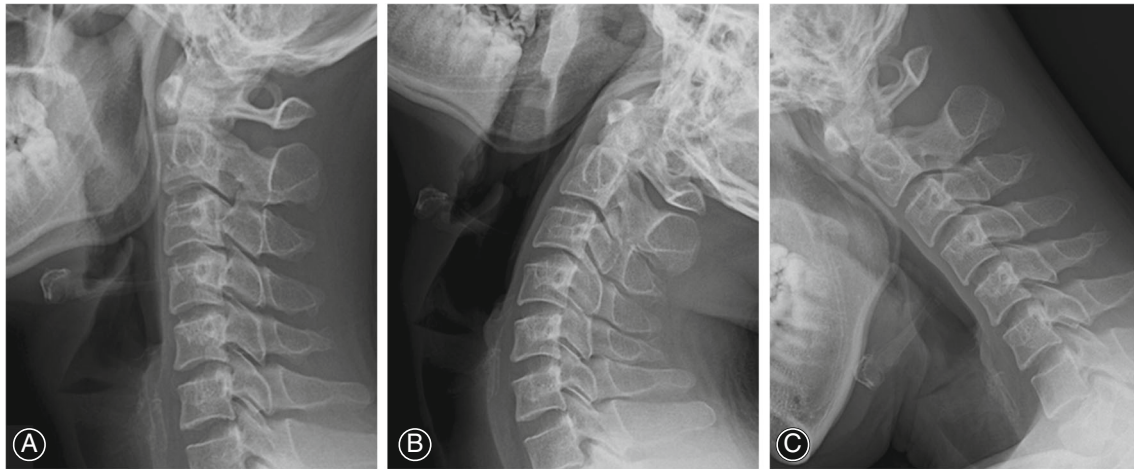


Fig. 1 Plain radiographs and lateral dynamic views of the cervical spine. (A) Lateral plain radiograph of the cervical spine showed the formation of a complete PP; (B) Lateral dynamic radiograph (extension) revealed no apparent occipitocervical or atlantoaxial instability and the sagittal alignment of the cervical spine was a normal lordosis; (C) Lateral dynamic radiograph (flexion) revealed no apparent occipitocervical or atlantoaxial instability and the sagittal alignment of the cervical spine was a normal lordosis

and the reported symptoms of these patients were mainly vertebrobasilar insufficiency symptoms and/or localized pain^{3,5,9-11}.

Here, we report a rare case of a patient who suffered from torticollis, facial asymmetry, localized pain, and Barré-Liéou syndrome in connection with one-sided PP. We also review the pertinent literature, focusing on the surgical treatment for symptoms due to PP.

Case Presentation

Data and Examination

A 23-year-old Chinese male presented with the chief complaint of continuous significant dizziness to the point of losing consciousness while rotating his head to the right for a period of 18 months. The patient also complained that manual or house work exacerbated the dizziness, which was accompanied by headache, pain in the occipitocervical and retro-orbital regions, and recurrent disturbances of vision, leading to his unemployment 12 months ago. The patient reported that a photographer discovered his facial asymmetry 3 years ago when he was “asymptomatic,” and thus, he did not consult his doctor. There was no history of trauma.

Physical examination revealed facial asymmetry and persistent torticollis without pain; neck torsion and a mild lateral inclination of the head to the left shoulder was observed. The facial asymmetry was characterized by the left hemiface being wider than the right hemiface, with the chin deviated to the right¹². Evaluation of facial asymmetry using Economou’s methods revealed respective measured differences between the height and width of the superior palpebrale between the left and right sides of 0.2 cm and 0.1 cm, and differences between the height and width of the cheilion between the left and right sides of 1.1 cm and 0.1

cm, respectively¹³. Meanwhile, there was also a right side depression of the malar prominence, with moderate downward displacement of the ear, eye, and mouth¹⁴. Neither a shortening nor spasm of the sternomastoid muscle on either side was found¹⁵. Barré-Liéou syndrome (manifesting as dizziness, headache, retro-orbital pain, and recurrent disturbances) was produced by turning the head in an extended position to the right side and exerting pressure with the thumb on the region between the external occipital protuberance and the level of the lateral masses of the atlas⁹. Occipitocervical-region pain (VAS 6) was produced by this pressure compression in the region of the left external occipital protuberance and posterior C1 lateral mass. The strength and sensation of the extremities were normal, as were the tendon reflexes of both upper and lower extremities. No pathological signs were elicited. No pigmented skin lesions nor endocrinopathy on the patient’s body were noted.

Plain radiographs of the cervical spine showed a C1 anomaly with complete PP formation (Figure 1A). Lateral dynamic views showed no apparent occipitocervical or atlantoaxial instability (Figure 1B and C). Computed tomography (CT) of the cervical spine showed the formation of complete PP on the left side with acute-angled, C-shaped kinking of the VA (Figure 2). We considered the left VA to be dominant based on the average diameters of the V3 segment of the VA on the left and right sides measuring 3.6 mm and 1.5 mm, respectively. A coronal CT image showed a deviation of the shape and height of C1 lateral masses on different sides; the heights of the left and right lateral masses were 1.83 cm and 1.74 cm, respectively (Figure 3). Magnetic resonance imaging (MRI) of the cervical spine did not reveal compression or irritation of the VA and spinal cord in the subaxial level.

Fig. 2 Computed tomography (CT) of the cervical spine. (A) Parasagittal CT image of the cervical spine revealed the formation of complete PP on the left side. (B) Three-dimensional CT revealed formation of complete PP on the left side with acute-angled, C-shaped kinking of the VA (dominant)

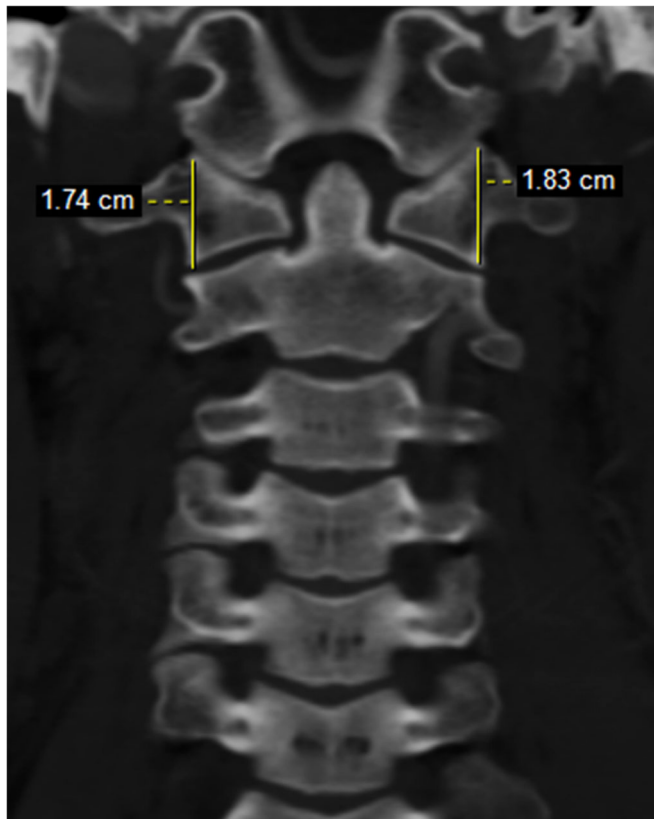
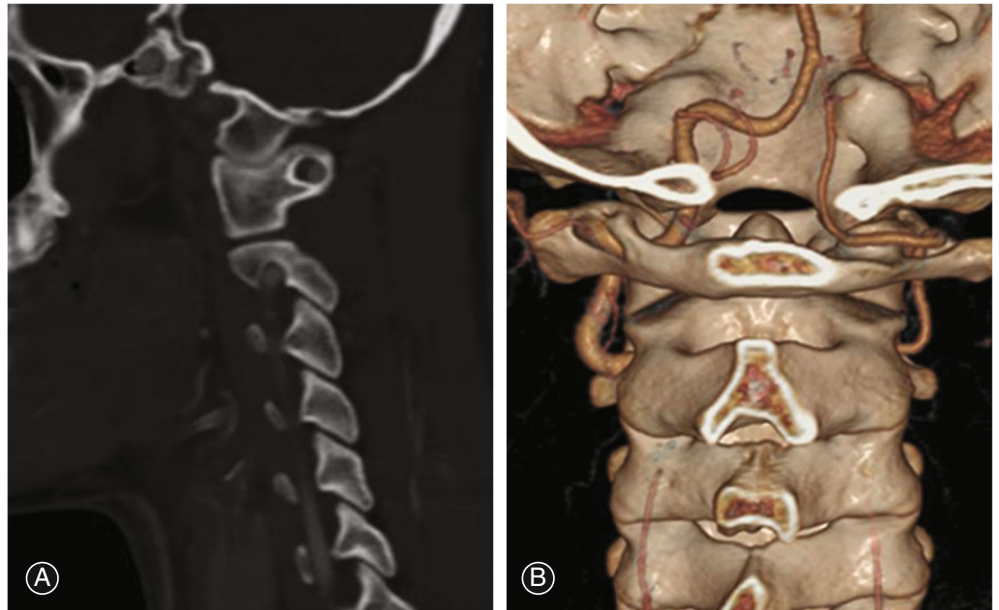


Fig. 3 Coronal CT image showed a deviation of the shape and height of C1 lateral masses on different sides. The heights of the left and right lateral masses were 1.83 cm and 1.74 cm, respectively

Surgery

Because of the ineffectiveness of various conservative methods attempted for 12 months prior, a resection of PP *via* the posterior midline was performed. The possibility of a combination diagnosis with other neurological diseases was excluded before operative treatment. The formation of complete PP on the left side was confirmed after the soft tissue and muscles were dissected. Two nerve dissectors were used to protect the VA within the bony bridge, and the bony bridge was accurately skeletonized from the external to the internal cortical edges using Kerrison rongeurs, a curet, and an ultrasonic bone cutter. When the internal cortical edge of the bony bridge was the only remaining residue, it was resected using a nerve dissector and Kerrison rongeur. At this point, venous bleeding was meticulously coagulated by bipolar coagulation and temporarily tamped with an absorbable gelatin sponge. The remaining bony bridge was then meticulously resected. After removal of the bony fragments, a slight compression of the VA remained owing to a thin layer of fibrous tissues under the removed bony bridge. When the fibrous tissues were removed using Kerrison rongeurs and blunt-pointed microscissors, the pulse of the VA became gradually enhanced.

Postoperative Period and Follow-Up

The patient was extubated immediately after surgery and reported significantly alleviated symptoms of localized pain (VAS 2) and Barré-Liéou syndrome 1 day post-operation. A postoperative plain radiograph (Figure 4) and CT (Figure 5) showed the complete resection of PP and sufficient decompression of the VA. Three months after surgery, the patient



Fig. 4 Postoperative plain radiograph showed complete resection of PP

reported a significant improvement in quality of life and returned to his previous manual work. However, there were no obvious changes in the torticollis and facial asymmetry.

Discussion

PP is a prevalent condition, with many studies demonstrating associated symptom series including vertigo, dizziness, vertebrobasilar insufficiency, diplopia, migraine, cervicogenic headache, and neck pain^{2,3,5,10,16-19}. However, recent studies about PP seem to focus on its association with headache and its implications during the placement of lateral mass screws in the atlas^{17,20,21}. Until now, only six studies have described the surgical treatment of PP (Table 1)^{3,5,8-11}.

We reviewed all the information of the six published studies reporting surgical treatment from 1979 to 2018, consisting of three case reports and three retrospective studies (Table 1)^{3,5,8-11}. Out of a total of 75 patients, information on patient age was provided for 68 patients (23 to 58 years old); most of these patients (60/68 cases) were aged 23 to 37 years. Information about the population, gender, type, location of PP, and symptoms are shown in Table 1. Vertebrobasilar insufficiency symptoms accounted for the most common conditions (100%, 75/75 cases), usually manifesting as Barré-Liéou or Bowhunter's syndrome^{9,10,22}. Localized pain was

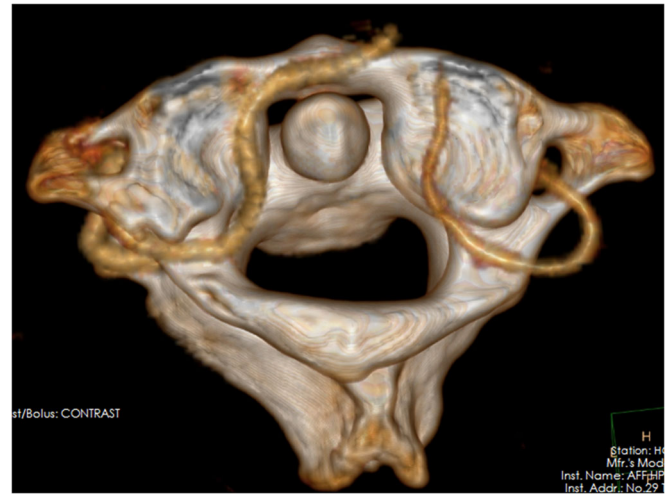


Fig. 5 Postoperative three-dimensional CT showed complete resection of PP

found in 13% of patients (10/75 cases). Reported approaches include resection of PP through the posterior midline (71 patients) and a minimally invasive lateral approach (four patients). After operation, 71 patients (94.7%) reported to have very good, good, or fair results according to the classification for evaluating clinical outcome, and only four patients (5.3%) reported poor results⁹.

The patient we report herein suffered from torticollis and facial asymmetry in addition to localized pain and Barré-Liéou syndrome. There are no studies reporting the association between PP and such a tetrad. Our patient's torticollis should be considered compensatory, based on the following: (i) the patient did not show torticollis in his childhood; (ii) the deviation of the shape and height of the C1 lateral masses between the two sides was not significant, which could not be congenital, since a severe deviation of the C1 lateral masses might develop in patients with a congenital bony anomaly; and (iii) imaging and physical examination did not show muscular torticollis or other causes for torticollis. Therefore, the presentation of torticollis could have been a compensatory posture to spontaneously alleviate the compression or irritation of the structures within PP, similar to the compensatory scoliosis in patients with lumbar disc herniation. The compensatory torticollis might have contributed to the development of deviation of the shape and height of the C1 lateral masses, revealing that the influence of PP-associated physiopathology had started earlier than expected. Similarly, the long-term compensatory posture likely contributed to the development of facial asymmetry.

The patient we described in this study reminds us that the early diagnosis and timely treatment of PP is important to improve patients' quality of life and avoid potential deformities, such as torticollis and facial asymmetry. The patient had satisfactory relief of localized pain and Barré-Liéou

TABLE 1 Literature review findings of the surgical treatment of PP

Authors/type of report	year	Population	Number of patient	Age (years)/sex	Complete/incomplete	Left/right/both	Symptom	Other findings	Treatment	Response to operation ^a	FU
Tedeschi G ¹¹ / case report	1979	Italy	1	NA	NA	NA	Vertebro-basilar insufficiency	None	Posterior midline approach	Good	NA
Limousin CA ⁹ / retrospective study	1980	South America	30 30 PP cases with cervical spondylosis as control group	23/NA 37/NA	NA NA	NA NA	Barre-Lieou syndrome Barre-Lieou syndrome	Anxiety Probable symptoms of cervical spondylosis	Posterior midline approach Posterior midline approach	21 very good 5 good 3 fair 1 bad 19 very good	NA NA
Sun JY et al. ¹⁰ / retrospective study	1990	Chinese	6	NA/ NA	2 complete/ 3 incomplete/ 1 osteophytic incomplete	2 right/ 1 left/ 1 both	Dizziness, Barre-Lieou syndrome, local pain	None	Posterior midline approach	Very good	1-3 years
Taylor et al. ⁸ / case report	2012	North America	1	41/M	Incomplete	Right	Bowhunter syndrome	None	Removal of PP	Very good	NA
Lvov et al. ⁵ / retrospective study	2017	Russia	3 cases 3 cases with minimally invasive approach	30-58/F 20-58/M (2),F (1)	NA NA	NA NA	Dizziness, Barre-Lieou syndrome, local pain	None None	Posterior midline approach Minimally invasive lateral approach	Good Good	1 year
Lukianchikov et al. ⁷ / case report	2018	Russia	1	34/F	Complete	Right	Bowhunter syndrome, local pain	ponticulus lateralis	Minimally invasive lateral approach	Very good	6 months
Present case/ case report	2021	Chinese	1	23/M	Complete	left	Barre-Lieou syndrome	torticollis, facial asymmetry	Posterior midline approach	Good	3 months

Abbreviations: PP, ponticulus posticus; FU, follow-up.; ^a Response to operative excision of PP were classified as “very good,” “good,” “fair,” and “bad” according to the description of Limousin CA in 1980⁹. “Very good” means patients remain asymptomatic following surgical treatment; “Good” means patients experience only occasional episodes of neck pain and vertebrobasilar insufficiency; “Fair” means patients still have episodic symptoms of vertebrobasilar insufficiency, which is usually of short duration; “Bad” means patients still have the original symptoms with the same or less severity.

syndrome after surgery, but there were no obvious changes in the torticollis and facial asymmetry. Improvement of torticollis and facial asymmetry would likely be greater if surgery had been performed early. We believe that the deviation of the shape and height of the C1 lateral masses on different sides was the structural factor causing our patient's persistent torticollis; understandably, it was less likely to satisfactorily correct torticollis and facial asymmetry in this mature patient owing to the structural factor and loss of remodeling opportunity. However, there are a paucity of studies about early diagnosis and timely surgical treatment³. Although approximately 5.5%–7% of the population with PP has symptom series, it is necessary to pay attention to this situation because of the high prevalence of PP in the general population. Conservative treatment with physiotherapy and injection might relieve symptoms for most patients with PP. The criteria for surgical treatment are the failure of conservative treatments and the increase of clinical signs severely impacting patient quality of life. Resection of PP is recommended in such cases^{3,9,10}.

In summary, the significant aspect of the presented clinical case was the tetrad of torticollis, facial asymmetry, localized pain, and Barré-Liéou syndrome caused by complete one-sided PP. Although this PP anomaly was seemingly “asymptomatic” for a long time, the presentation of torticollis and facial asymmetry reveals the early influence of PP in this patient might have started early¹⁹. The patient had

satisfactory relief of localized pain and Barré-Liéou syndrome after surgery, while the torticollis and facial asymmetry predictably did not show obvious changes. This case study serves to remind spine surgeons to pay greater attention to various clinical signs with PP besides localized pain and vertebral insufficiency.

This case is a rare presentation of torticollis, facial asymmetry, localized pain, and Barré-Liéou syndrome associated with one-sided complete PP. Active surgical strategies should be considered if the patient is recalcitrant to conservative therapy. Early diagnosis and timely treatment could significantly improve patients' quality of life, likely avoiding residual deformity such as torticollis and facial asymmetry.

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Conflict of Interest

None of the authors have any potential conflicts of interest.

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