Congenital Defect of the Posterior Arch of Cervical Spine: Report of Three Cases and Review of the Current Literature

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Abnormalities of the posterior arch, including congenitally absent cervical pedicle and cervical spondylosis, are rare entities that are usually found incidentally on neck radiographs. It is important to recognize these characteristic radiological features because their radiographic appearance may cause them to be confused with more serious entities such as fractures, locked facets, and tumor-induced bony erosions. Also, it is important to distinguish these abnormalities from similar pathologies to prevent the use of inappropriate treatment. We report the relevant clinical and radiological findings seen in three cases of posterior arch defect after trauma with review of pertinent literature.

KEY WORDS: Absent cervical pedicle · Cervical spondylosis · Congenital abnormality.

INTRODUCTION

Abnormalities of the posterior arch including congenital absence of a cervical pedicle and cervical spondylosis are usually found incidentally after minor trauma and these are unusual disorders with a high potential for radiographic misinterpretation, especially in case of acute trauma.

Disconnected lesion of the cervical spine was first reported by Hadley in 1946 as a unilateral absent pedicle. In 1951, Perlman described a defect that allowed both forward migration of the superior vertebra. Patients with these anomalies tend to present with cervical pain after trauma. Initial evaluations with conventional radiography frequently lead to misinterpretation and misguided intervention; it may simulate jumped locked facet, unilateral facet fracture or nerve root tumor. Three dimensional computer tomographic (CT) studies have aided the understanding of the morphology of absent cervical spine pedicle and cervical spondylosis. CT scan should be performed in order to accurately evaluate the radiological diagnosis. Most reports have recommended a conservative nonsurgical management for a successful outcome. Neural compression or instability is uncommon with these anomalies, only in rare instances is surgical intervention appropriate.

We report three cases of posterior arch defects (two cases of congenitally absent cervical pedicles and one case of cervical spondylosis) and describe the presentation, diagnosis, and treatment of these patients. Also, review on literature on this clinical entity concerning the distinguishing features between absent cervical pedicle syndrome and spondylosis is presented.

CASE REPORT

Case 1
This 50-year-old woman presented with neck pain with paresthesia on bilateral upper and lower extremities during neck flexion after a motor vehicle accident. Physical examination showed no remarkable findings. Cervical spine X-ray showed an enlarged neural foramen extending from C4 to C5 on the left side, suggesting a unilateral C4-5 jumped locked facet or spinal tumor (Fig. 1A). CT scanning revealed the absence of the left pedicle of C4 (Fig. 1B, C). There were
no fractures. Flexion and extension films of his cervical spine without the collar revealed no instability and cervical spine MRI showed C3-4 disc bulging with mild cord compression. The patient was treated conservatively and eventually became asymptomatic after conservative trial of 3 weeks.

Case 2
A 11-year-old boy without significant past medical history presented with acute onset of severe neck pain after a fall from a bed. In the neurological examination, the patient was alert and oriented. Motor strength was 5/5 throughout, and sensation was without demonstrable deficits. Plain films of the cervical spine were obtained and thought to be consistent with a left, C5-C6, unilateral locked facet. Subsequent radiographic evaluation with 3-dimensional CT revealed no evidence of traumatic injury, and it did demonstrate absence of both C5 pedicle and spina bifida occulta at C5 (Fig. 2). The patient was treated conservatively with improvement of neck pain and no restrictions were placed on his activity.

Case 3
A 43-year-old man was referred with the diagnosis of unstable bipedicular fracture of C6 following a traffic accident. He presented with posterior neck and both arm pain. Neurological examination revealed normal findings. The standard X-rays (AP, lateral and oblique views) suggested a congenital or acquired anomaly with defect of the pars interarticularis and spina bifida occulta at C6 (Fig. 3A). Cervical CT scan revealed bilateral spondylosis of the C6 vertebra. Reconstruction CT confirmed the spondylosis of the pars interarticularis. The pars articularis defect and the C6 spina bifida were well visualized on the reformatted CT scan (Fig. 3B, C). Bone scan was normal, confirming the absence of recent bone injury. The patient was given symptomatic care. After resting for 2 weeks, the symptoms disappeared. Although spondylolisthesis (grade I) was observed, the disc degeneration was mild. Thus, we did not perform fusion for this patient. We are following this case further, and we will consider fusion surgery if further slippage and/or symptoms appear.

DISCUSSION
Posterior arch defects of the cervical spine, including congenitally absent cervical spine pedicle and cervical spon-
dyloysis, are complex of abnormalities in posterior arch development and occur rarely. Absence of a cervical spine pedicle is an unusual congenital anomaly with potentially confusing clinical and radiological manifestations. Fewer than 100 cases have been reported in the literature to date since it was first described by Hadley 60 years ago. 6,7,11,12,18

Cervical spondyloysis is much more rare than lumbar spondyloysis which is estimated 5% of the population. Perlman and Hawes10 first described cases of cervical spondyloysis and spondylolisthesis in 1951, and since then, only about 100 cases have been reported in the literature worldwide, mainly in adults. This anomaly is generally seen on C6 (74% of cases). The other cervical vertebrae (excepting C1 and C7) may also present this anomaly. They are similar to each other, but there are some distinctive differences between them.

The radiological criteria of congenitally absent cervical spine pedicle syndrome and cervical spondyloysis are now well established. Radiographic evaluation of these patients typically begins with conventional radiography but frequently includes CT for confirmation. As in three present cases, two/three-dimensional computed tomography allows clear confirmation of this specific diagnosis. Generally, MRI does not provide any further information due to the poor bone resolution, but it can be most helpful in cases with neurological involvement for differential diagnosis.

Wiener et al.10 described typical radiographic characteristics of a congenitally absent cervical pedicle and triads of findings are as follows: 1) an enlarged ipsilateral neural foramen because of the absent pedicle; 2) a dysplastic, dorsally displaced ipsilateral articular pillar and lamina; and 3) a dysplastic ipsilateral transverse process. Other osseous abnormalities are frequently associated with this anomaly including spina bifida occulta, vertebral body or arch fusion and additional hypoplastic pedicles.

Forsberg et al. proposed the following findings to be standard features of cervical spondyloysis: 1) a well-marginated cleft between the facets, 2) a triangular configuration of the pillar fragments on either side of the spondyloytic defect, 3) posterior displacement of the dorsal triangular pillar fragment, 4) hypoplasia of the ipsilateral pedicle, 5) spina bifida at the involved level, and 6) compensatory hyper- or hypoplasia of the ipsilateral articular pillars at the level above and/or below the defect.

It is important to recognize the features of cervical spondyloysis and distinguish it from fractures or absent pedicle syndrome. Congenital absence of the cervical pedicle is almost a unilateral lesion with ipsilateral articular dysplasia. In addition, the upper facet joint is hypoplastic or absent. There is no spondylolisthesis but spina bifida may be associated.

Bilateral aplasia of the pedicles has been reported in association with spondyloysis in one case but was more likely a CT acquisition or interpretation error. In case of recent stress fracture of the pars interarticularis, bone scan and MRI are helpful to distinguish it from congenital anomaly. For older fractures, the diagnosis is more difficult and relies on the notion of an old trauma and absence of associated dysplasia.

In general, congenital cervical pedicle defect and cervical spondyloysis are diagnosed as an incidental finding on routine radiologic examinations especially after trauma. The most common presenting symptom is neck pain, and is sometimes associated with the pain, numbness and/or paresthesia of an upper extremity. The majority of patients have normal findings from neurologic examination. It must be distinguished from similar pathologies to prevent the use of inappropriate treatment. Conservative therapy is recommended for patients with congenital absence of a cervical pedicle, as it was successful in most cases. Conservative therapy consists of rest, analgesics, muscle relaxants, and/or physical therapy and it is successful in alleviating symptoms. Surgery may not be necessary in most cases with this clinical entity. Surgical treatment is reserved for unstable and/or painful lesion. Cervical spine injuries, patients with intractable long standing neck pain or minor neurologic deficits after neck injury, who also have instability, should be considered surgical candidates.

Most patients with cervical spondyloysis have been effectively treated through conservative management by a temporary immobilization. But, patients with this condition who experience a traumatic event have a higher risk of developing neurologic deficit compared to patients with intact spines. Thus, early diagnosis and management of cervical spondyloysis is important. Surgical treatments should be reserved for cases in which conservative measures fail, or in the presence of instability, or fragility of the involved and adjacent segments. If patients have cervical instability or a painful lesion after trauma, surgical treatment (ex, fusion surgery) is necessary. If patients are asymptomatic without evidence of instability and spinal cord signal changes, patients should be managed conservatively. However, we treated two cases of congenital absent cervical pedicle and one case of cervical spondyloysis. All patients treated conservatively. We had a good results, because our patients had a single level lesion and showed no neurologic deficit or instability after trauma.

The embryological pathogenesis of congenitally absent pedicles is not conclusively known. Either failure of development of a vertebral chondrification center for the posterior arch of a particular sclerotome or failure of appropriate ossification can lead to the absence of a pedicle, the ventral
half of the lateral mass, and the dorsal part of the transverse process. Such a developmental anomaly probably develops at the gestational age of 7 to 9 weeks. The exact cause of cervical spondyloysis remains controversial. Several theories have been proposed, including congenital, developmental, microtrauma, and posttraumatic nonunion with pseudarthrosis. Failure of unification of the chondrification and ossification centers is another theory that may account for the spectrum of posterior arch defects, which includes cervical spondyloysis as well as an absent pedicle.

CONCLUSION

Congenitally absent cervical spine pedicle and spondyloysis involving the cervical spine are rare conditions. Careful inspection of cervical spine radiographs with knowledge of characteristic radiological features should allow proper diagnosis in all cases. Recognition of these features and differentiating it from acute cervical fracture or dislocation, which could result in inappropriate surgical intervention, is important in patients with recent trauma. CT scans, in particular two/three-dimensional reconstructions, coupled with plain films can be helpful in confirming the diagnosis and planning treatment. Appropriate knowledge and awareness of such lesions can prevent misdiagnosis and inappropriate therapy.

References