Egyptian Journal of Neurosurgery

CASE REPORT Open Access

Congenital agenesis of lumbo-sacral pedicles with associated anomalies: case report with an emphasis on the use of O-arm, navigation in the management with literature review

Bharat Dave, Devanand Degulmadi*, Ajay Krishnan, Shivanand Mayi, RaviRanjan Rai, Aditya Raj and Umesh Meena

Abstract

Background: Congenital spinal pedicle agenesis is rare. The majority of cases are asymptomatic. Few present with either low back pain or radiculopathy. Pedicle screw insertion may pose difficulties in view of abnormal anatomy.

Case presentation: Between Jan 2005 to March 2021, all the data was retrospectively reviewed for the cases operated for congenital anomalies. Three such cases were operated. Case 1 had bilateral agenesis of L5 vertebra with an ectopic kidney in right iliac fossa, absent uterus, absent ovaries and vaginal atresia. Case 2 had absent S1 pedicle with conjoint left L5 nerve root. Case 3 had hypoplastic left L5 pedicle, contained disc herniation, facetal arthropathy. Pedicle screws were placed using O-arm and navigation.

Conclusion: Pedicle agenesis increases the difficulty in pedicle screw fixation and fusion. Anomalous facet and conjoint nerve root need to be looked for intra-operatively. Use of O-arm and CT navigation is of great help in getting safe and accurate anchor points for screw placement avoiding neurological injury.

Keywords: Pedicle agenesis, Lumbo-sacral, Nerve root anomaly, O-arm and navigation

Background

Congenital hypoplasia and agenesis of the spinal pedicle is an uncommon entity. Cervical and thoracic spine are more commonly reported than lumbar or sacral pedicles [1–6]. Pedicle agenesis has specific radiographic features: a false appearance of an enlarged neural foramen; a dysplastic, dorsally displaced ipsilateral articular pillar and lamina; and a dysplastic ipsilateral transverse process [5]. The majority of cases with an absence or hypoplasia of the lumbosacral pedicles are asymptomatic [4, 7]. Few of them can present with either low back pain or radiculopathy [7, 8]. Computed tomography (CT) is preferred diagnostic modality [5, 9]. Pedicle screw insertion may pose difficulties. We present 3 cases of pedicle agenesis in lumbo-sacral spine with associated anomalies. We also discuss the intra-operative challenges encountered and the role of O-arm and navigation in surgical management of these cases.

Case presentation

Between Jan 2005 to March 2021, all the data was retrospectively reviewed for the cases operated for congenital anomalies. Data was retrieved for cases with agenesis of lumbo-sacral pedicles from the records and presented here.

*Correspondence: drdanand@yahoo.co.in
Stavya Spine Hospital & Research Institute, Ahmedabad, Gujarat, India

Case 1

A 44 year old female presented to us with symptoms of low back pain since few years. She had an electric shock injury 2 months back followed by which she sustained an injury to her back and had severe back pain. Inspite of taking 2 months of medical treatment and bed rest, she did not get relief and her mechanical back pain worsened. Sleep was disturbed. On examination she had tenderness over upper and lower lumbar spine. Neurology and distal vascularity was normal. Plain radiography showed wedge compression fracture of L2, bilateral absent pedicles of L5 with L5-S1 anterolisthesis (Fig. 1a). MRI further confirmed the findings. An ectopic kidney was seen in right iliac fossa (Fig. 1b). CT showed L5-S1 anterolisthesis, bilateral absent L5 pedicles, hypoplastic transverse process, absent posterior elements (Fig. 2 a, b, d). Further

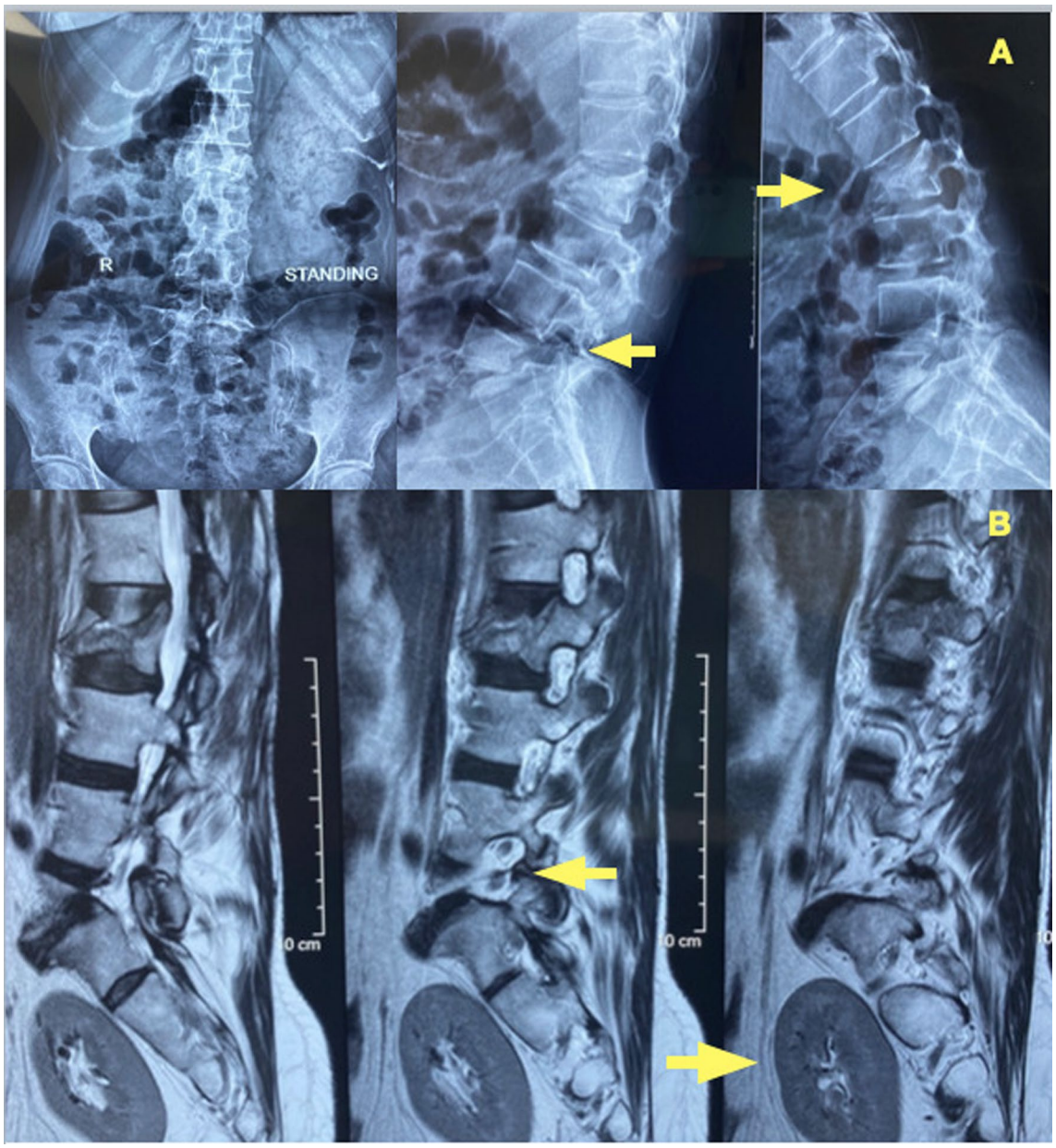

Fig. 1 **a** Plain radiographs of case 1 depicting wedge compression of L2 vertebra and dysplastic anterolisthesis of L5 over S1. The pedicles of L5 vertebra are absent bilaterally. There is some opening up of L2 fracture as visualized on flexion–extension radiographs. **b** MRI of the same patient showing corroborative findings. Two nerve roots can be seen passing through the neural foramen. Also noted is ectopic location of kidney in the pelvis

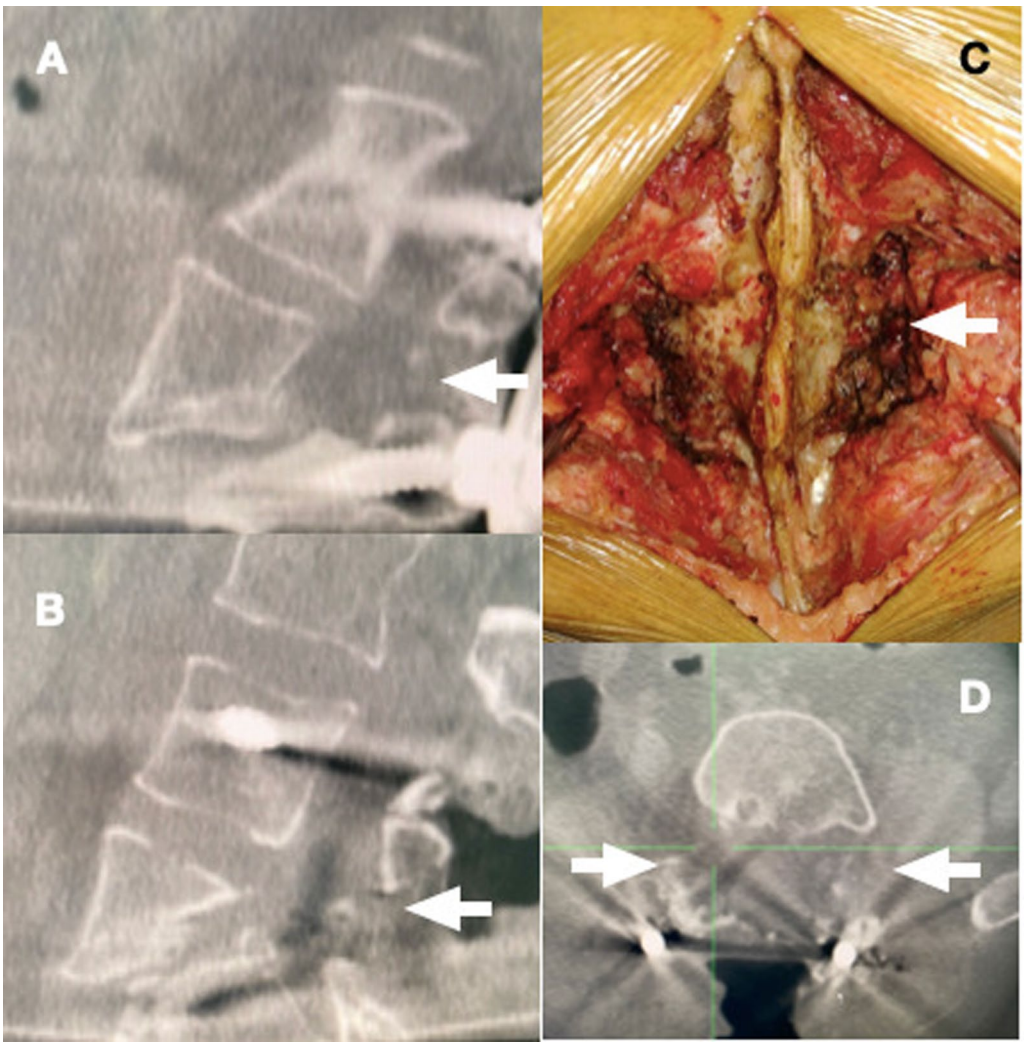

Fig. 2 **a**, **b**, **d** Intraoperative sagittal and axial CT scan of case 1 depicting L5-S1 anterolisthesis, absent peicles hypoplastic transverse process and posterior elements of the L5 vertebra. **c** Intraoperative clinical photograph showing similar findings of hypoplastic posterior elements and transverse process with absence of pedicles

investigations of ultrasound abdomen showed absent uterus, absent ovaries and vaginal atresia.

As the patient had severe mechanical back pain with chronic history of back pain, surgery was planned after detailed counselling and consent was taken. Surgery for non union of L2 fracture as well as L5-S1 anterolisthesis was planned. Intra-operatively the following findings were noted—Posterior elements of L4 lamina, facet joint of L4-5 and transverse process were hypoplastic (Fig. 2c). Pedicle screws were placed in L1-3 for stabilisation of fracture L2 and L4, S1 under the guidance of O-arm and navigation. L5-S1 interbody fusion was performed with autologous bone graft from L5 lamina. Gentle and thorough preparation of end-plates of performed as L5 vertebral body was unstable. Post-op imaging showed proper placement of screws and bone graft for fusion. Patient was mobilised on the 2nd post-op day and discharged on 4th post-op day. She had good relief of her mechanical back pain during follow-up visits.

Case 2

A 26-year-old male patient presented with lower back pain of 6 months duration and left-sided acute S1 radiculopathy for 15 days. ADL was restricted, with significant disability on walking for 5–10 min. Physical examination revealed positive Lasegue sign on left side with S1 hypoaesthesia without any motor deficit. Plain radiographs were noted to have facet joint abnormality on left L5-S1

joint. CT scan further revealed absent L5 inferior articular process and hypoplastic S1 superior articular process along with absent S1 pedicle on left side (Fig. 3a). MRI was suggestive of a left-sided L5/S1 contained disc herniation with abnormal hypoplastic facets and a large neural foramen (Fig. 3b). In view of severe intractable pain, L5-S1 transforaminal lumbar interbody fusion surgery was planned. Intra operatively, conjoint nerve root at left L5 level (type 2b- Neidre and MacNab) with absent S1 pedicle on left was noted on careful exploration (Fig. 4a). O-arm and CT navigation guided pedicle screws were inserted. Left S1 screw was placed using a lower entry point and divergent direction as compared to right (Fig. 4b). Interbody cage was inserted from right in contrast to left as planned. Postoperatively patient had good resolution of his symptoms.

Case 3

A 25 years old woman presented with low back pain since last 2 years and left lower limb radicular pain since 3 months. All modalities of conservative treatment failed to give her relief. On examination, lumbar spasm was noted and SLR on left side was 30^0 with normal power. Imaging revealed hypoplastic L5 pedicle, contained disc herniation on left side and L4/5, L5/S1 facetal arthropathy on right side (Fig. 5a–c). Limited microsurgical inter laminar decompression was planned. Through a 3 cm incision, left side exposure, fenestration decompression was done. Post-operative period was uneventful. At 30 months follow-up, she had no complaints and resumed her previous activities. At final follow-up, flexion/extension radiographs show no instability.

Discussion

Complete agenesis of the lumbar pedicle appears to be a rare entity [10]. According to Wortzman and Steinhardt, the terms 'hypoplastic' and 'agenesis' have been used synonymously with resultant confusion [10]. Dysgenesis or agenesis of the spinal pedicle is thought to be the result of a large retrosomatic cleft during embryological development [11]. This seems to occur in either membranous or cartilaginous stages of development [9, 11]. Developmental defects of the vertebral pedicle are reported to be persistent neurocentral chondrosis, retrosomatic defect (cleft pedicle) or retroisthmic defect. Agenesis of pedicle has been reported more commonly in the cervical spine and very rarely in lumbo-sacral spine. Pedicle agenesis can manifest unilaterally (case 2,3) or bilaterally (case 1). Congenital nature of the missing or absent or deformed pedicle is suggested by hypoplasia of the other elements of ipsilateral neural arch, as well as hypertrophy of various elements of contralateral neural arch. In our case series, one of them had facetal arthropathy on the contralateral side at both the adjacent levels and case 1 had

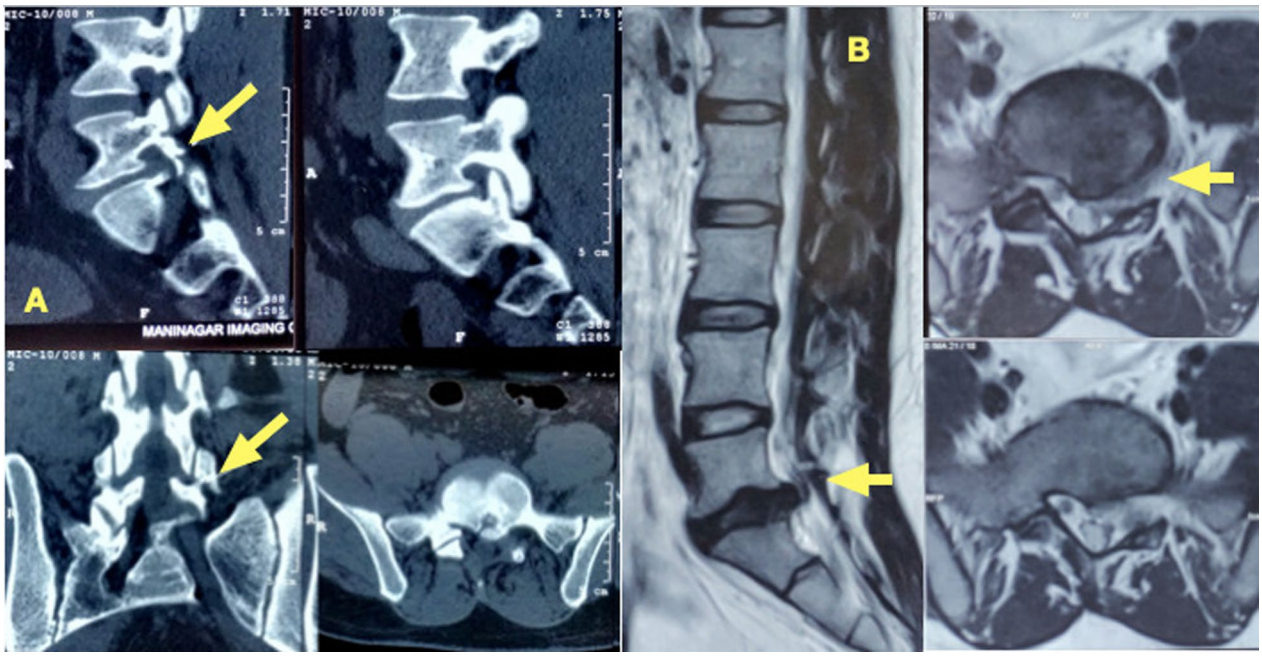

Fig. 3 **a** Sagittal, coronal and axial CT scan of case 2 depicting absent L5 inferior articular process and hypoplastic S1 superior articular process and absent S1 pedicle on left side. **b** Sagittal and axial MRI of the same patient showing L5-S1 paracentral contained disc herniation on the left side along with large neural foramen

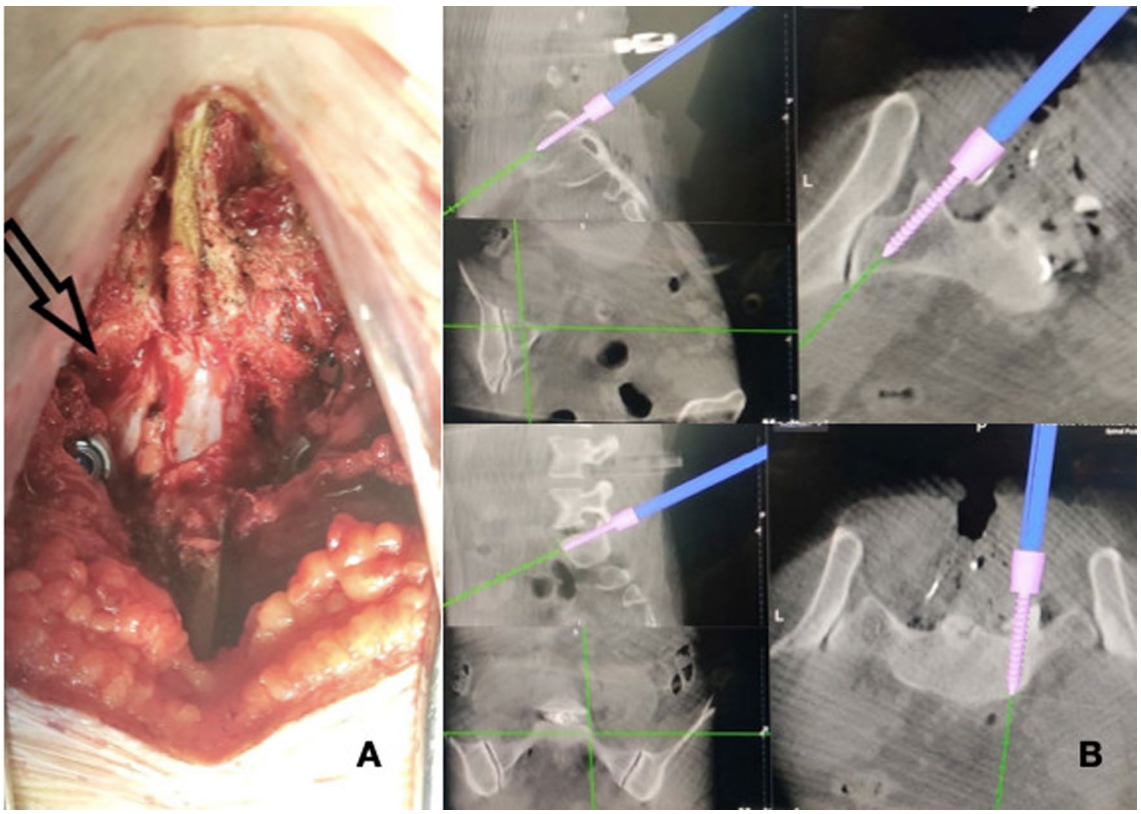

Fig. 4 **a** During intra-operative exploration in case 2 a conjoint nerve root at left L5 level (type 2b- Neidre and MacNab) with absent S1 pedicle on left was found. **b** Intra-operative O-arm and CT navigation picture showing placement of screws. The Left S1 screw was placed using a lower entry point and divergent direction towards the sacral ala

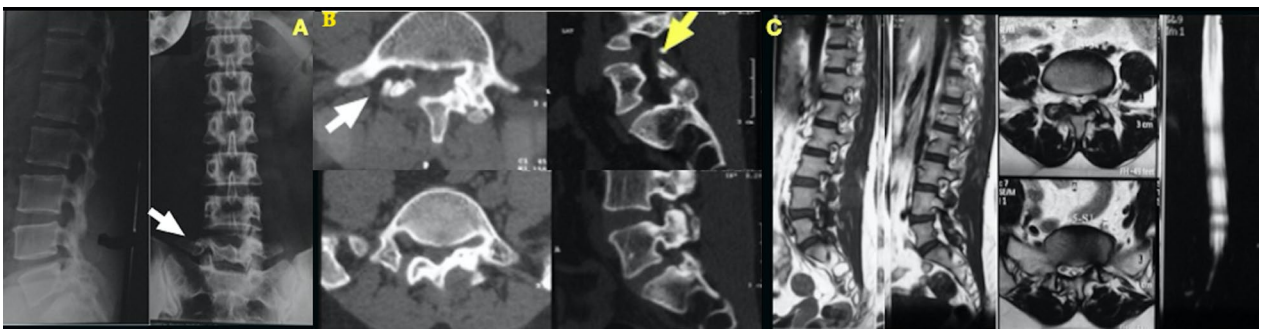

Fig. 5 **a** Plain radiographs of case 3 depicting hypoplasia of facet joints on right side and facet arthropathy on the left side. **b** Corresponding CT scan images showing corroborative findings and hypoplastic L5 pedicle on right side. **c** T2 Sagittal and axial MRI images demonstrating contained disc herniation on left side at L4/5 and L5/S1 facetal arthropathy on right side and hypoplastic L5 pedicle

absent posterior elements. Kaito et al. reported a similar case that was missing the right L5 pedicle, which led to severe degenerative changes in the contralateral facet joint [7]. Bilateral absence of pedicle usually results in spondylolisthesis (case 1) [12]. Low back pain is the frequently reported symptom, most of the times managed conservatively. Only few cases are reported in the literature with neurologic impairment, symptoms being present on the same side or contralateral side [7–9, 11]. Payer et al. reported a case of S1 pedicle aplasia in association with disc herniation that had ipsilateral leg pain [8]. Kaito et al. reported a congenital absence of the right

L5 pedicle and contralateral L5 radicular pain from an overloaded hypertrophic facet joint [7]. The exact pathomechanism of this radiculopathy is not yet explained but may be related to presence of disc prolapse, associated root anomalies or facetal hypertrophy. Most of the cases are asymptomatic or have minimal back pain and require only conservative management, on the other hand neurological involvement (radiculopathy) requires surgery mostly fusion surgery. In our case series, fusion was performed in two patients and limited decompression was done in case 3.

Plain radiographs often show pedicular anomalies more seen on oblique views. Listhesis, instability, tilting of vertebrae, displacement of spinous process need to be looked for. CT is the investigation of choice that depicts exact anatomical features of lamina, articular process, transverse process and neural foramen. Conditions such as infection / lytic spondylolisthesis / metastasis needs to be differentiated, margins of the defect gives the clue. MRI reveals disc herniation, facetal hypertrophy, anomalous roots and neural compression. The incidence of lumbosacral nerve root anomalies is 1.9–4% [13]. The conjoined nerve roots are occasionally associated with herniated disc, however, cases associated with spine anomalies are uncommon [14]. Pedicle screw insertion for fusion in such conditions poses a challenge for the operating surgeon. Case 2 had sacral pedicle agenesis with facet anomaly in association with lumbosacral conjoint nerve root and disc herniation. In the absence of the S1 pedicle and presence of conjoint nerve root at L5 exit foramen it becomes difficult in insertion of S1 pedicle screw as superior articular process of S1 is only attached to the posterior arch of S1. Use of advanced newer technology in such difficult cases helps a spine surgeon in identifying the bony landmarks in more accurate way. It also helps the spine surgeon to get good pedicle screw placement. Neurological injury can be avoided. Congenital pedicular agenesis has been associated with renal hypoplasia, absent kidney and imperforate anus in paediatric age group in a report by Yousefzadeh et al., however no relation has been seen in adults [15]. We had a case of malpositioned kidney in right iliac fossa, absent uterus, absent ovaries and vaginal atresia, all in one patient. Other case had root anomaly type 2b-Neidre and MacNab at the level of absent pedicle. These cases are reported for the first time.

Conclusion

Congenital agenesis of lumbo-sacral pedicle is very rare. Anomalous facet and conjoint nerve root need to be looked for intra-operatively. Pedicle agenesis increases the difficulty in pedicle screw fixation and fusion. Use of O-arm and CT navigation is of great help in getting safe and accurate anchor points for screw placement avoiding neurological injury.

Abbreviations

CT: Computed tomography; MRI: Magnetic resonance imaging.

Acknowledgements

None.

Authors' contributions

BRD—conception. DD—design of the work and drafting of manuscript. AK—Acquisition of data. SM—Drafting of manuscript. RR—Data analysis. AR—Editing. UM—Proof reading. All authors have read and approved the manuscript.

Funding

None.

Availability of data and materials

None.

Declarations

Ethics approval and consent to participate

Obtained.

Consent for publication

Obtained.

Competing interests

None.

Received: 14 May 2021 Accepted: 26 February 2022
Published online: 11 April 2022

References

1. Asakawa H, Yanaka K, Narushima K, Meguro K, Nose T. Anomaly of the axis causing cervical myelopathy. Case report J Neurosurg. 1999;91(1 Suppl):121–3.
2. Koyama T, Tanaka K, Handa J. A rare anomaly of the axis: report of a case with shaded three dimensional computed tomographic display. Surg Neurol. 1986;25(5):491–4.
3. Trivedi P, Vyas KH, Behari S. Congenital absence of the posterior elements of C2 vertebra report: a case. Neurol India. 2003;51(2):250–1.
4. Moon SJ, Lee JK, Seo BR, Kim SH. Traumatic subluxation associated with absent cervical pedicle: case report and review of the literature. Spine. 2008;33(18):663–6.
5. Wiener MD, Martinez S, Forsberg DA. Congenital absence of a cervical spine pedicle: clinical and radiologic findings. AJR. 1990;155:1037–41.
6. Sheehan J, Kaptain G, Sheehan J, Jane J Sr. Congenital absence of a cervical pedicle: report of two cases and review of the literature. Neurosurgery. 2000;47(6):1439–42.
7. Kaito T, Kato Y, Sakaura H, Yamamoto K, Hosono N. Congenital absence of a lumbar pedicle presenting with contralateral lumbar radiculopathy. J Spinal Disord Tech. 2005;18:203–5.
8. Michael P, Enrico T. L5/S1 disc herniation and S1 pedicle aplasia: case report and review of the literature. Acta Neurochir 2014;156.
9. Polly DWJ, Mason DE. Congenital absence of a lumbar pedicle presenting as back pain in children. J Pediatr Orthop. 1991;11:214–9.
10. Wortzman G, Steinhardt M. Congenitally absent lumbar pedicle: a reappraisal. Radiology. 1984;152:713–8.
11. Sener RN. Sacral pedicle agenesis. Comput Med Imaging Graph. 1997;21:361–3.
12. Patel AJ, Vadivelu S, Desai SK, Jea A. Congenital hypoplasia or aplasia of the lumbosacral pedicle as an unusual cause of spondylolisthesis in the pediatric age group. J Neurosurg Pediatr. 2013;11:717–21.

13. Postacchini F, Urso S, Ferro L. Lumbosacral nerve-root anomalies. J Bone Joint Surg Am. 1982;64:721–9.
14. Okuwaki T, Kunogi I, Hasue M. Conjoined nerve roots associated with lumbosacral spine anomalies. A case report. Spine 1991;16:1347–49.
15. Yousefzadeh DK, El-Khoury GY, Lupetin AR. Congenital aplastic-hypoplastic lumbar pedicle in infants and young children. Skeletal Radiol. 1982;7:259–65.

Publisher's Note