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CASE REPORT

Unilateral hyperplasia of the left posterior arch and associated vertebral schisis at C6 level

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Abstract We report on a 5-year-old girl with unilateral hyperplasia of the left posterior arch of C6 associated with spina bifida occulta at the same level. Anteroposterior and lateral radiographs of the cervical spine showed hypertrophy of the left lamina as well as overgrowth and elongation of the left spinous process of the sixth cervical vertebra. Computed tomography (CT) examination better depicted this congenital variant and clearly showed the associated schisis of the posterior arch at the same level. Magnetic resonance (MR) imaging examination ruled out other spinal anomalies. The neck pain, the young age of the patient and the local aesthetic abnormality contributed to the surgical indication. To the best of our knowledge, this is the first case in the English literature of unilateral hyperplasia of a posterior cervical arch. Only one previous study has reported a similar congenital anomaly, but it was limited to the left side of the spinous process.

Keywords Spine · Spine, congenital abnormalities · Spine, CT · Vertebral schisis

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Introduction

Congenital spinal anomalies are relatively common. Several of these spinal abnormalities occur uniquely in the lower cervical region [1, 2]. Examples of relatively common disorders of the lower cervical spine include persisting apophyses of the transverse processes, persisting epiphyses, vertebral platyspondylia, vertebral hypoplasia, and dysplasia of the vertebral arch. Congenital abnormalities of the transverse and spinous processes are often incidental radiological findings rarely associated with clinical manifestations namely back pain, or constitute aesthetic problems [1, 2].

In contrast to the aforementioned disorders, unilateral hyperplasia of a cervical spinous process is an extremely rare congenital anomaly having been illustrated only once in the English literature [3]. Chitkara et al. [4] described, but did not clearly illustrate two "rib-like swellings" at the back of the neck. Furthermore, unilateral hyperplasia of a posterior cervical arch has not previously been described in the English literature, which is why the observation of a 5-year-old girl harboring a giant spinous process associated with hypertrophy of the left lamina and spina bifida occulta at C6 level prompted us to the present report.

Case report

A 5-year-old girl presented to the neurosurgery clinic with a posterior midline cervical mass and increasing neck pain, which her parents stated as having been present for at least 2 years. There was neither history of trauma nor family history of relevance. One year before the admission, the pain had increased in intensity. At the time of presentation, her pain was evoked by the pressure on the spinous process of the sixth cervical vertebra and not related to physical

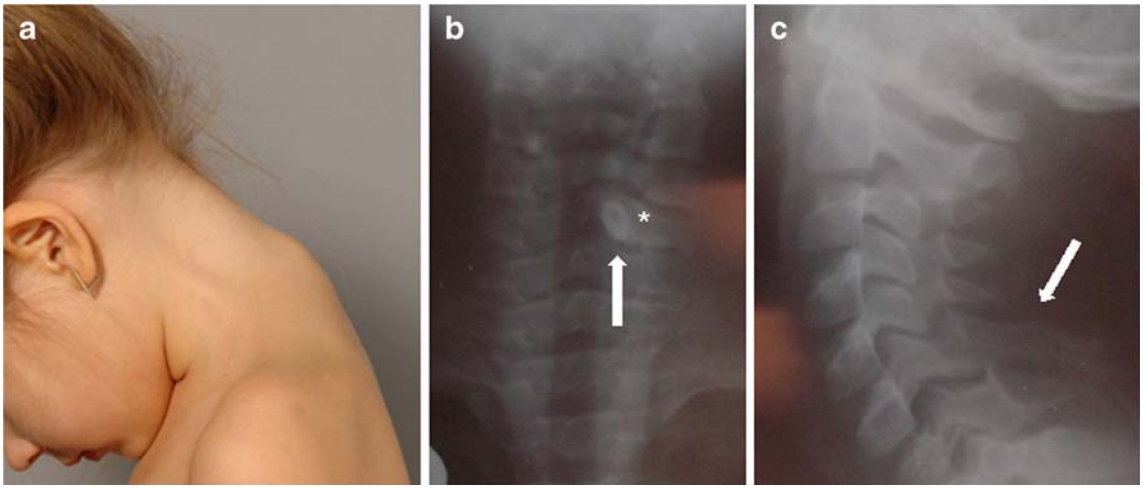

Fig. 1 **a** Photograph showing the posterior midline lower cervical protuberance at presentation. **b** AP radiograph of the cervical spine shows the off midline position and sclerosis of the bifid C6 giant spinous process (*arrow*), and hypertrophy of the left lamina at the same level (*asterisk*). **c** Lateral radiograph of the cervical spine clearly shows overgrowth and elongation of the spinous process of the sixth cervical vertebra (*arrow*)

activity. The neurological examination at the admission was unremarkable. The full range of cervical movements was preserved. Physical examination pointed out the presence of a posterior cervical midline hard mass, more evident after neck flexion (Fig. 1a). Laboratory findings were normal.

Radiographs of the cervical spine revealed hypertrophy of the left lamina as well as an abnormal elongated and thickened spinous process of the sixth cervical vertebra (Fig. 1b,c). Computed tomography (CT) examination with reformatted images on the sagittal plane and three-dimensional reconstructions better defined the presence of a C6 giant spinous process that was directed posteriorly and downwards, and associated with hypertrophy of the left lamina, and schisis of the posterior arch at the same level (Figs. 2 and 3). There was no evidence of pseudo-articulations and the giant spinous process had well-defined cortices and medullae on CT scan (Figs. 2 and 3). Subsequent magnetic resonance (MR) imaging examination of the cervical spine revealed a well-circumscribed cortical bone within the dorsal neck muscles and did not demonstrate morphological or signal abnormalities of the spinal cord. Both the congenital variant and the surrounding soft tissues showed normal signal intensities. No other pathological findings were detected in the cervical spine.

Surgical excision of the C6 spinous process was planned and carried out through a posterior approach without any intra-operative or post-operative complications (Fig. 4). Histological examination of the gross specimen revealed normal bone tissue. At the 3-month postoperative follow-up, the patient was pain-free and aesthetically satisfied with the procedure.

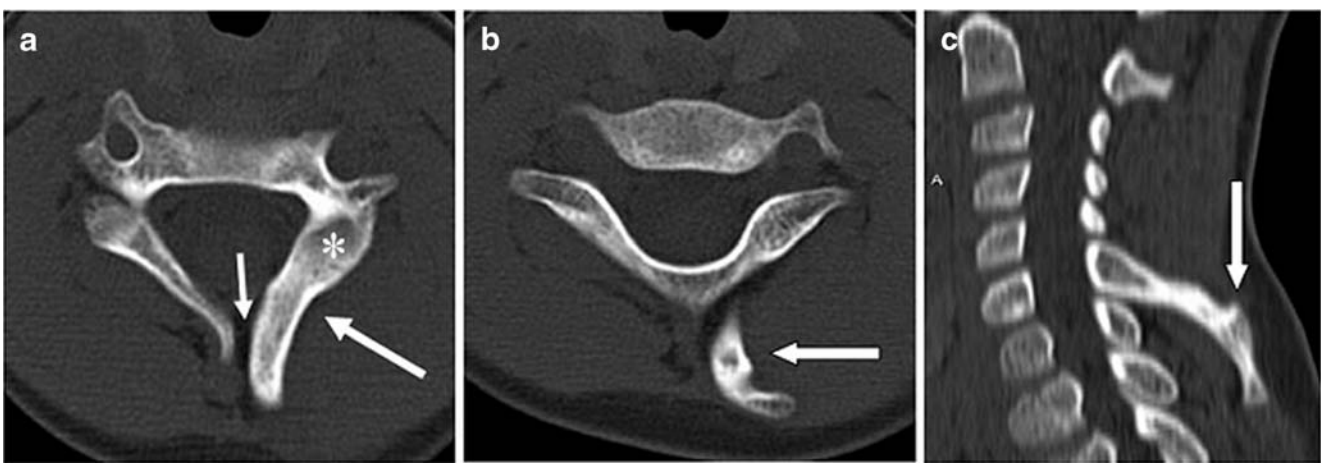

Fig. 2 **a**–**b** Axial CT scans passing through C6 vertebra show the absence of pseudoarticulations at the base of the spinous process and within it (*arrow* in **a** and **b**). Schisis of the vertebral posterior arch (*small arrow* in **a**), and hypertrophy of the left lamina (asterisk in **a**) are also evident. **c** Sagittal MPR image of the cervical spine shows hyperplasia of the left C6 posterior arch with the giant spinous process directed posteriorly and downwards (*arrow*)

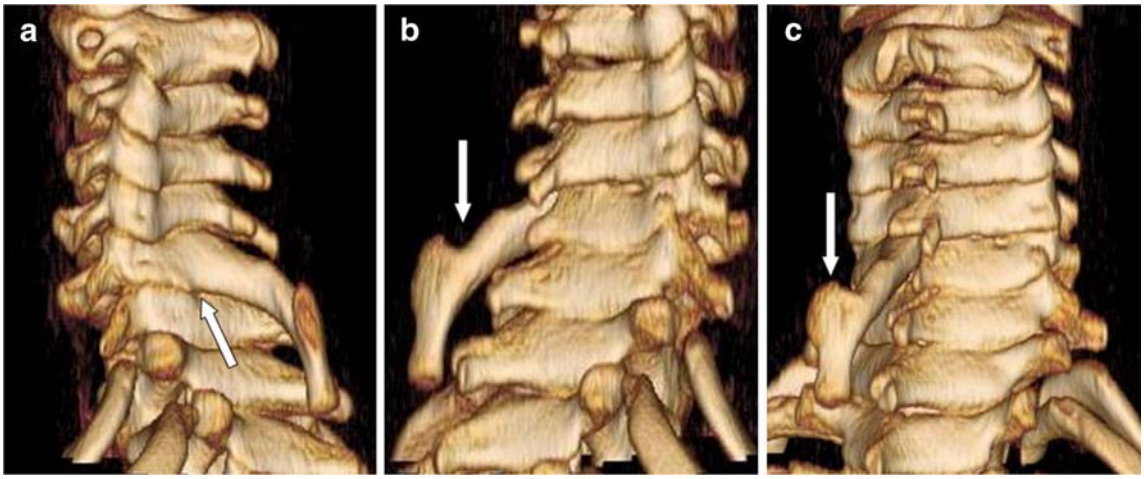

Fig. 3 **a**–**c** Oblique sagittal three-dimensional volume-rendered CT images from left (**a**), right (**b**), and back (**c**) respectively, clearly confirm the absence of pseudoarticulations at the base (*arrow* in **a**) and within (*arrow* in **b** and **c**) the left C6 giant spinous process

Discussion

Congenital osseous abnormalities of the cervical spine can occasionally cause clinical symptoms such as limitations of the full range of cervical movement, cervical pain, neurological deficits and aesthetic anomalies [2]. Precise diagnosis of a congenital spinal anomaly is primarily based on imaging studies. Although radiographic examination can depict spinal abnormalities in most cases, radiographic examination may be unsatisfactory and differentiation between congenital osseous anomalies and post-traumatic changes may be difficult. Therefore, if uncertainty as to the diagnosis exists, CT examination is indicated. However, the combination of CT and MR imaging examinations is the best way of evaluating spinal anomalies because patients with congenital spinal deformities have a high incidence of intraspinal pathological conditions (30–35%) even in the absence of clinical findings [5, 6]. In our patient, CT scan clearly demonstrated a marked unilateral hyperplasia of the left posterior arch of the sixth cervical vertebra with no evidence of pseudo-articulation, and associated with spina bifida occulta at the same level (Figs. 2, 3).

Typical cervical spinous processes are short, down-turned, and bifid at their apices to afford a greater surface for muscular attachment. Their length increases progressively from the fourth to the seventh vertebrae. The spinous process of C7 vertebra differs from the others; it is thick, nearly horizontal, and not bifurcated. Furthermore, the spinous process does not have its own ossification center (except for its tip); it is formed by fusion of the endochondral growing osseous extensions from both vertebral arches during the first year of life. The tip of the spinous process develops from a secondary ossification center at puberty.

Unilateral hyperplasia of cervical spinous process is a rare radiological finding; only one case, described by Heyer et al. [3], has been clearly illustrated in the English literature so far. The authors reported on a 24-year-old woman with a distinct unilateral left-sided hyperplasia of the spinous process of the seventh cervical vertebra, who

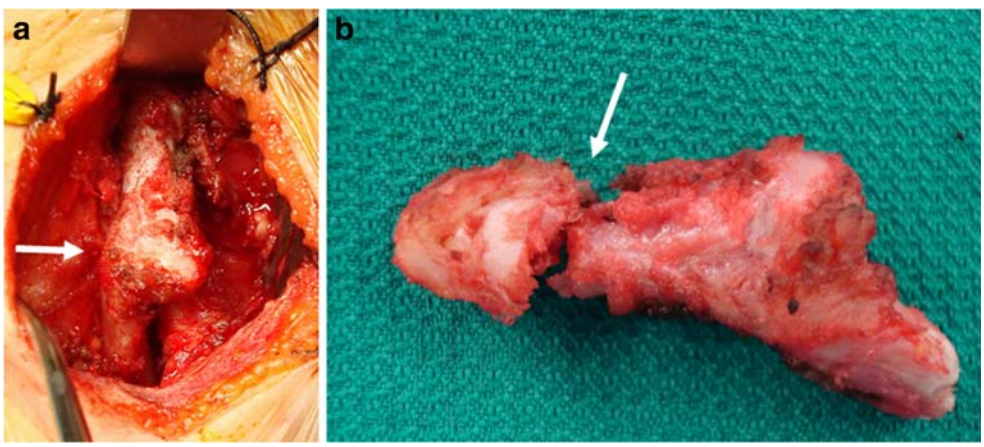

Fig. 4 **a** Intraoperative digital photograph showing C6 giant spinous process (*arrow*; top of image is proximal). **b** Digital photograph of the gross specimen that has been fractured during surgery for easier removal (*arrow*)

suffered from chronic dorsal neck pain, in the absence of pathological findings at the physical examination and with a normal neurological status. The role of cross-sectional imaging was emphasized. No information about the treatment was provided. In contrast to the previous case, our pediatric patient showed an evident aesthetic abnormality, hypertrophy of the left lamina, and a posterior arch schisis at the radiological examination, besides the dorsal neck pain. Furthermore, the neck pain, the local aesthetic abnormality, and, above all, the young age of the patient contributed to the surgical indication. In a young patient, this spinal anomaly may cause secondary pseudoarthrosis which might prevent normal vertebral alignment during growth of the body.

The exact etiologic factors involved in the pathogenesis of unilateral hyperplasia of a posterior cervical arch are unknown; however, abnormal extension of chondrification and ossification of one vertebral arch with normal development of the other arch can explain the present anomaly. The radiological differential diagnosis of giant spinous process primarily includes post-traumatic changes, and pelvic digit/rib. CT scans easily rule out post-traumatic spinal changes such as non-union spinous process fracture or ossifying hematoma; furthermore, in our case, there was no history of trauma. Pelvic digit/rib is an unusual developmental anomaly in which bony formations arise in soft tissue around the normal skeletal bone. Radiologically, it resembles a rib- or a phalanx-like bone with a clear cortex and medulla, and often with a characteristic pseudoarticulation at the base, or one or more pseudoarticulations within it [7–13]. The mechanism of formation of the pelvic digit/rib is not yet established. The most likely theory is that the anomaly arises in the mesenchymal stage of bone growth within the first 6 weeks of the embryonic stage. If the cartilaginous costal primordium of the first coccygeal vertebra does not fuse with the spine, the cartilaginous center may develop independently, forming a rudimentary "rib" [12]. Segmentation of these cartilaginous centers might cause the digit-like appearance [8].

Several digit/rib-like structures in the pelvic region have been reported [7–13]. Ishikawa et al. [14] identified a digit-like structure in the thoracic region. Atalar et al. [15] first described two bony formations resembling a coccygeal segment or a phalanx, posterior to the spinous process of the C6 vertebra. They had well-defined cortices and medullae, but a characteristic pseudo-articulation between each other and with the spinous process was also demonstrated.

In our patient, unilateral left-sided hyperplasia of the C6 spinous process, with no evidence of pseudo articulation, was associated with schisis of the posterior vertebral arch at the same level. Vertebral schisis is the simplest neural tube defect caused by the failure of the fetus's spine to close properly during the first month of pregnancy. Schisis of the spinous processes in the lower cervical spine is a common finding often associated with other osseous anomalies such as congenital absence of a cervical spine pedicle [16] and cervical spondylolysis [17]. Furthermore, schisis of the lower cervical spinous processes represents a very consistent finding in mild diastrophic dysplasia [18, 19] and recessive multiple epiphyseal dysplasia [20]. Mutations in the diastrophic dysplasia sulfate transporter gene result in a family of recessively inherited chondrodysplasias including, in order of decreasing severity, achondrogenesis 1B, atelosteogenesis 2, diastrophic dysplasia, and recessive multiple epiphyseal dysplasia. Diastrophic dysplasia is a distinct autosomal recessive disorder of cartilage and bone development which is characterized by short stature, progressive scoliosis, joint limitation, clubfoot deformity, brachydactyly, symphalangism, "hitchhiker's thumb", malformed pinnae with calcification of the cartilage, cauliflower deformity and cleft palate in some cases. A distinct group of patients who have similar but less severe involvement have been referred to as having "diastrophic variants". However, there is wide variability in the phenotypic expression of diastrophic dysplasia and those individuals identified as having "diastrophic variants" should be referred to as having mild diastrophic dysplasia [18, 19]. Recessive multiple epiphyseal dysplasia is characterized by joint pain, malformations of hands and feet, multilayered patella and scoliosis [20, 21]. In contrast to other multiple epiphyseal dysplasia types [22, 23], pre-pubertal children with recessive multiple epiphyseal dysplasia usually do not show short stature [20].

In conclusion, we describe the first case of marked unilateral hyperplasia of a left posterior cervical arch associated with spina bifida occulta at the same level. This finding may be of clinical interest to radiologists, neurologists, orthopaedic surgeons, anthropologists and forensic personnel. We suggest that in dealing with abnormalities involving the lower cervical spine, the possibility of anomalies such as the presence of a giant spinous process or unilateral hyperplasia of a posterior arch should be considered and the treatment planned accordingly.

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