



Case Report

Giant Hypertrophy of the Fifth Cervical Spinous Process with associated Hemivertebra presenting as Posterior Neck Mass

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Abstract

Background: Congenital anomalies of the spine are relatively common and can affect both upper and lower limbs. However, hypertrophy of the spinous process is an extremely rare anomaly of the cervical spine. The majority of these vertebral abnormalities are often incidental findings in radiography with few causing pain, and aesthetic problems to the patients.

Patient main concern and important clinical findings:

Thus, we present a 12-year-old female who presented to the paediatric outpatient clinic on account of swelling at the lower back of her neck of 4 years duration, and recurrent neck pain of one month duration. The patient noticed the mass 4- years before presentation, as a painless, slow-growing lump behind her neck which appears more prominent whenever she flexes her neck. The USS revealed a linear hyperechogenicity within the hypochoic soft tissue (muscle) of the posterior neck and a plain radiograph of the neck was subsequently obtained and showed abnormal elongation with downward curvature of the spinous process of the sixth cervical vertebra. The patient's main concern was the sustained increase in the size of the mass with associated pain and unknown cause. A diagnosis of giant hypertrophy of the fifth cervical vertebra spinous process with hemivertebra at C6 was made. She was managed conservatively with satisfactory outcomes.

Conclusion: This case report illustrates a possible differential diagnosis of congenital anomalies of the hypertrophy of the spinous process in the evaluation of posterior neck mass in Nigeria

Keywords: Spine, cervical spine, computed tomography, hypertrophy of the spinous process

Introduction

Congenital anomalies of the vertebral column are relatively common in medical practice. Cervical spinal abnormalities are seen in both upper and lower cervical spines and can occur in association with other anomalies.^{1,2} However, hypertrophy of the spinous

process is an extremely rare anomaly of the cervical spine with only a few cases reported.^{1,2} Various anomalies of the cervical spines that are commonly encountered include persisting apophyses of the transverse processes, persisting epiphyses, vertebral platyspondylia, vertebral hypoplasia, and dysplasia of the arch of the vertebra.² Most of these vertebral abnormalities are often incidental

findings in radiography with few causing pain, and aesthetic problems to the patients.^{1,2} There are documented associations of cervical vertebra anomalies with Klippel-feil syndrome, Morquio syndrome, dystrophic dwarfism, spondyloepiphyseal dysplasia, and osteogenesis imperfecta.³ These syndromic effects may partly explain the reason for various configurations of anomalies documented in the cervical spine.

This case report is a documentation of giant hypertrophy of the fifth cervical (C5) vertebra spinous process with hemivertebra in a 12-year-old girl who was presented with a history of posterior neck swelling of 4-year duration. It is an extremely rare finding with only a few cases documented in English literature.

Thus, we present a rare case of giant hypertrophy of the spinous process and literature review as noted in the index patient.

Case Presentation

Miss N.M. is a 12-year-old female who presented to the paediatric outpatient clinic on account of swelling at the lower back of her neck for 4 years duration, and recurrent neck pain of one-month duration. The patient noticed the mass 4- years prior to presentation, as a painless, slow-growing lump behind her neck which appears more prominent whenever she flexes her neck. There was no history of trauma, surgery, or previous lesions involving her neck. There was associated pain-which started one month before the presentation which has been recurrent, and worse on flexion. There was no similar mass or other masses anywhere in her body, and no history of surgical excision of any mass lesion in the past. There was no loss of movement, no past history of neck stiffness, and no loss of sensation. There was no weight loss, jaundice, and no significant change in her usual activities. The patient is a student in junior secondary school and maintains above-average grades in school. She presented to the hospital because of the sustained increase in the size of the swelling, and the neck pain that started recently.

Physical examination revealed a young girl in no obvious distress, not pale, afebrile, acyanosed, anicteric, not dehydrated, and no pedal oedema. The temperature was 36.1°C, the Pulse rate of 89 b/min, and BP was 110/60 mmHg. Neurological examination was unremarkable.

Musculoskeletal revealed an immobile mass measuring 4.1x2.4cm as shown in Figure 1. The mass was not tender, no differential warmth, was firm-hard in consistency, and was not attached to the overlying skin. It was non-reducible, appeared more prominent with flexion of the neck, was non-pulsatile, and had no loss of sensation.

There was no associated restriction (stiffness) in the movement of the neck.

Investigations and Treatment

The routine tests: urinalysis, full blood count, and retroviral screening were not remarkable. Ultrasonography (USS) of soft tissue of the posterior neck was requested to evaluate the mass. The USS revealed a linear hyperechogenicity within the hypoechoic soft tissue (muscle) of the posterior neck as shown in Figure 2. A complementary plain radiograph of the neck was subsequently obtained and showed abnormal elongation with downward curvature of the spinous process of the fifth cervical vertebra with distinct continuous cortices and medullae through the entire length; there was also subtle abnormal curvature of the cervical spine towards the right side noted in the anteroposterior radiograph with associated hemivertebra at the left sixth cervical vertebra (C6), and osteophytic bridging of the transverse processes of C6 and C7 on the left as demonstrated in figure 2, 3 and 4 below. There was no evidence of pseudo-articulation. Echocardiography and correlative abdominal ultrasonography show normal findings.

A diagnosis of giant hypertrophy of the fifth cervical vertebra spinous process with hemivertebra at C6 was made based on imaging findings. Computed tomography and magnetic resonance imaging scans of the cervical spine were requested for further evaluation- to rule out spinal cord and nerve root involvement. However, these investigations were not procured due to lack of funds. The patient and her parents opted for a non-operative treatment – due to financial constraints and the fact that the patients' activities were not significantly affected. She is currently on conservative management for relief of pains and on monthly follow-up with satisfactory outcomes.

Discussion

Congenital vertebral anomalies of the cervical spines may cause clinical symptoms and signs which include reduction in the range of movement of the cervical vertebra, pain, neurologic deficit, and aesthetic problems for the patients.¹

The pathogenesis of spinous process hypertrophy is not known. It is believed to be due to developmental abnormalities and faulty fusion process at the 3rd – 6th embryonic weeks which marks the period for the intrauterine development of the vertebral column.^{1,2,3} The spinous processes lack an intrinsic ossification center, and are formed by the fusion of endochondral osseous extension from both vertebral arches around the first year

of life. The tips of the spinous processes develop from secondary ossification centres formed after the fusion.^{2,3,4} Abnormal extension of chondrification and ossification of the vertebral arches beyond the normal development processes are believed to be responsible for the occurrence of the hypertrophic spinous process.^{5, 6} Cervical vertebra anomalies are associated with syndromes may include: Klippel-Feil syndrome, Morquio syndrome, diastrophic dwarfism, spondyloepiphyseal dysplasia, and osteogenesis imperfecta.³ In the present case, echocardiography, and abdominal ultrasonography were used to rule out associated abnormalities of the heart and kidneys- which can be found in these syndromes. In addition, other imaging and clinical findings in this index patient were not suggestive of syndromic associations.

The diagnosis of the hypertrophic spinous process is based on imaging with plain radiography playing major roles. Computed tomography and magnetic resonance imaging may sometimes help to rule out other spinal complications.^{1,5,6} This is true of the index case where a suggestion of bony echogenicity was made on ultrasonography and confirmed by plain radiographic findings in the cervical spine of abnormally elongated C5 spinous process with hemivertebra at C6, and osseous bridging of the left transverse processes of C6 and C7. These findings could account for the patient's clinical presentation of neck pain as documented by Farooqi et al in which the patient also had hemivertebra in addition to abnormal elongation of the spinous process.¹ However, in the index case, the hemivertebra was located at the C6 level- which is a level below the abnormal spinous process, unlike the patient in Farooqi et al who had it at the C4 level- a level above the abnormal spinous process.

Hemivertebra occur commonly in the thoracic and lumbar vertebra and are associated with congenital scoliosis. Cervical hemivertebrae occur rarely in literature are detected early in children, and are treated with surgical excision and fusion of the involved vertebral level.^{1,4} Hemivertebrae have been associated with neurological signs from severe angulations of the spine, stenosis of the spinal canal, or spinal instability.⁴ Moghaddam and colleagues documented a case of intradural lipoma associated with hemivertebra.⁷ Some of these associations

could not be ruled out in the index case due to non-procurement of computed tomography, and magnetic resonance images of the cervical spine. However, the patient showed no clinical or neurologic symptoms and signs suggestive of significant spinal canal or neural involvement. This was equally concordant with the findings in other reported cases.^{1,5,6}

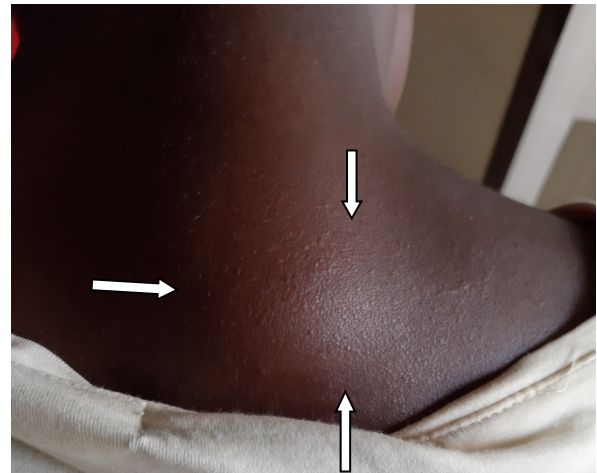


Figure 1: Gross picture of the posterior neck showing posterior neck swelling (white arrows)

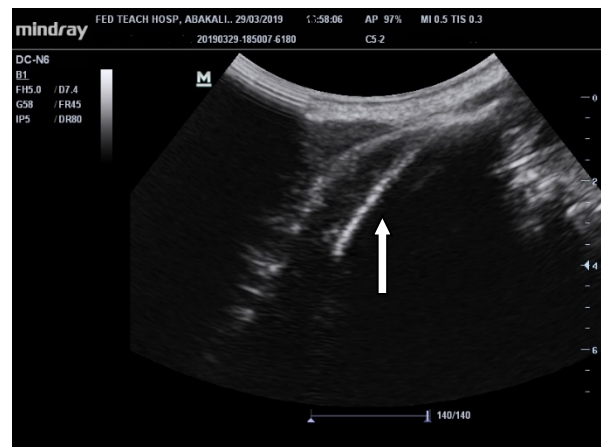


Figure 2: Longitudinal view ultrasonographic image of the posterior neck showing a linear hyperechoic structure suggestive of bone within the soft tissue of the neck. (white arrow)

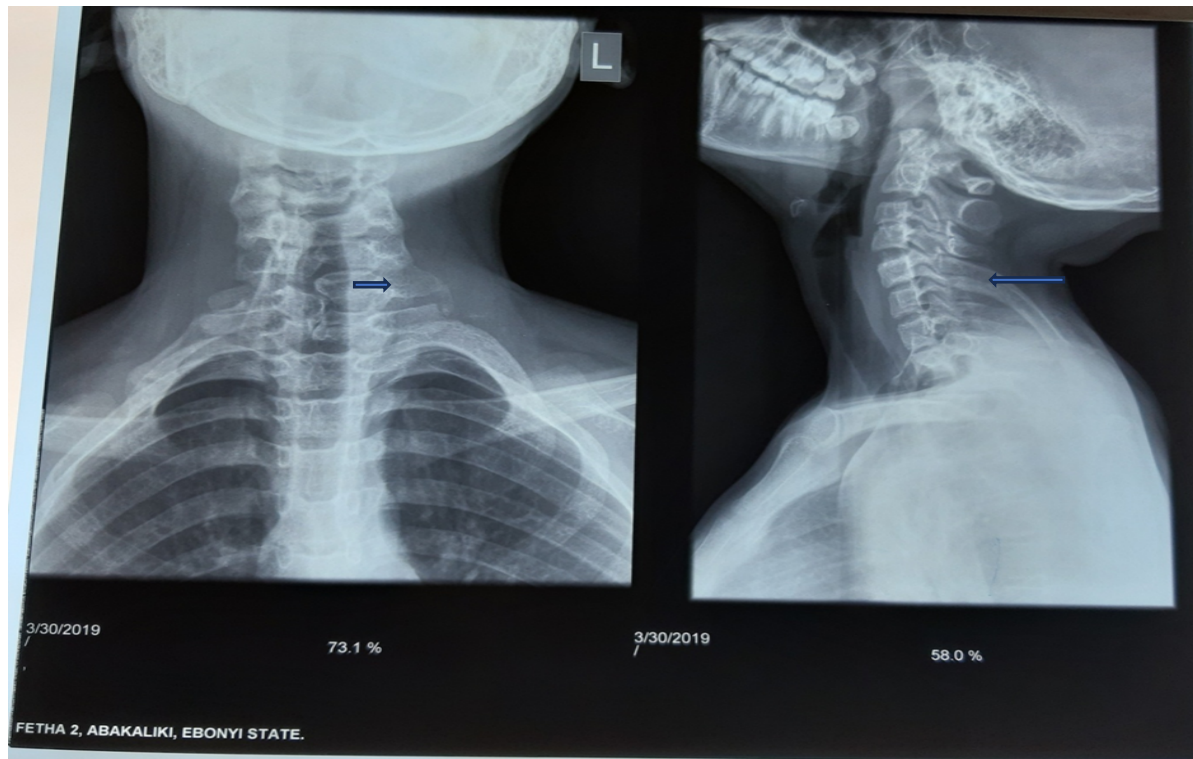


Figure 3: plain radiographs of the cervical spine (AP & Lateral) showing a giant spinous process of C5 curving downwards with well-defined medulla and cortical outlines (long horizontal white arrow). The AP radiograph shows subtle abnormal curvature towards the right with hemivertebra at C6 level (black arrowhead) and osteophytic bridging of C6/7 transverse processes (short white arrow).

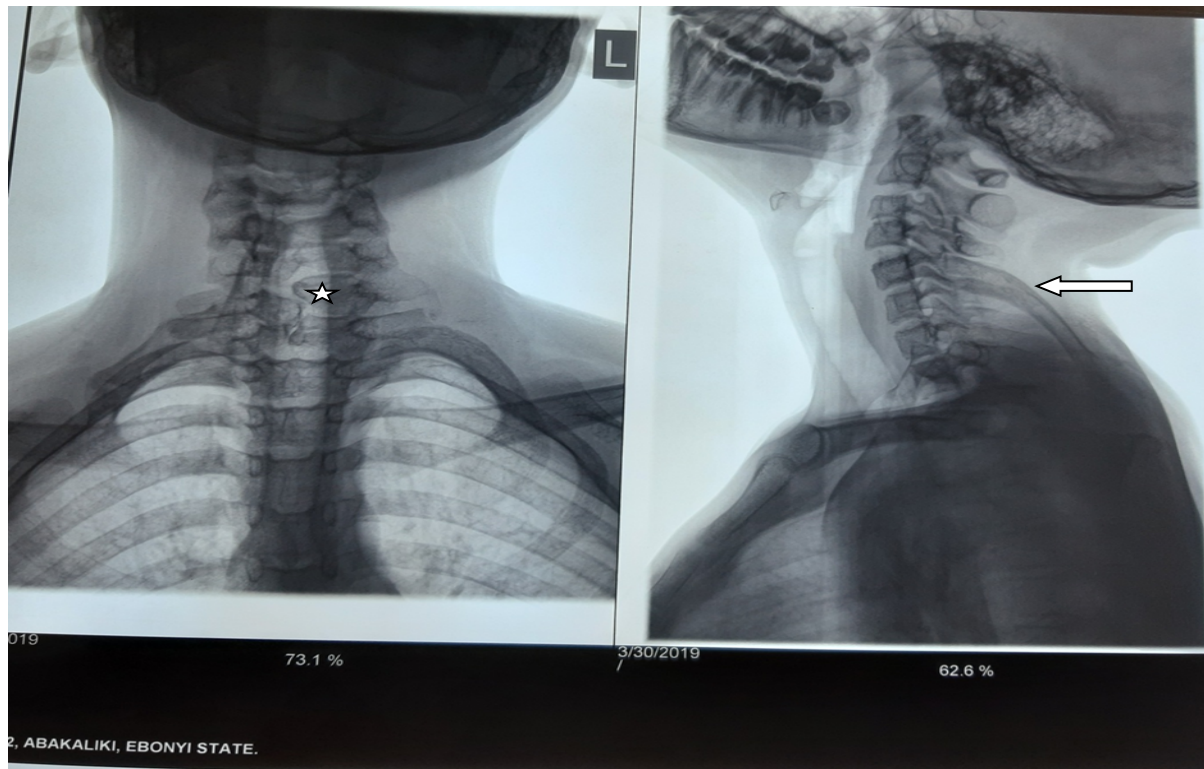


Figure 4: Positive image of Figure 3A: Rendering the bony outlines in a positive background demonstrating the elongated spinous process (white arrow), and hemivertebra (white star).

Furthermore, Esposito et al documented hyperplasia of the spinous process of the C6 vertebra with schisis at the same level. Hypertrophy of the spinous process is the common denominator between the index case and those of Esposito et al.⁶ The associations are however different. The index case is associated with the hemivertebra whereas in Esposito et al there is vertebral schisis. In addition, Mesut et al published a case of giant hypertrophy of the C5 spinous process with associated occipitalization of the atlas and congenital atlanto-axial fusion which was consistent with Klippel-Feil syndrome.⁸ The index case did not show vertebral fusion, no renal or cardiac abnormalities, and no clinical evidence of mental anomalies that maybe associated with the syndrome documented in the case of Mesut et al. Similarly, Reddy et al reported a case of giant C5 spinous process in a 7-year-old male child which was associated with partially blocked vertebrae at C2, C3, and C4 levels, unlike the index case.⁹ Report by Reddy et al mirrored the same spinal level with the index case. However, the present case is associated with hemivertebra at C6; unlike the partial fusion of C2, C3, and C4 vertebrae documented by Reddy et al.⁹

The radiographic differential diagnoses of giant spinous hypertrophy include post-traumatic changes like non-union of the spinous process, ossifying haematoma, and pelvic digit/rib.^{6,10} These are usually associated with pseudo-articulation and a history of trauma.⁶ Pelvic digit is a rare abnormal finger-like osseous outgrowth near a normal spinal skeleton which has been described in the pelvic, thoracic, and very rarely in the cervical region.^{10,11,12} The present case has no prior history of trauma to the spine, and no radiographic evidence of non-union or pseudo-articulation noted in the hypertrophied spinous process.

The management options for the hypertrophied spinous process are conservative management, and surgical excision for significantly symptomatic ones associated with spinal complications, and those with aesthetic problems. Conservative management protocols are reserved for those whose symptoms are mild, and not associated with neurologic deficit, severe pain, or aesthetic problems.^{1,5,6,7,8,9} The index patient did not show any clinical sign of neurologic deficit on examination, no aesthetic worries, with mild to moderate pain. She was therefore placed on conservative management with monthly follow-up plans and satisfactory outcomes.

Strengths and Limitations

This case report on congenital anomalies of the spinous process which presented as posterior neck mass could

mimic other soft tissue pathology in our local environments. Although, it's very rare, without thorough evaluation with relevant radiological investigation, the diagnosis of giant hypertrophy of the fifth cervical vertebra spinous process with hemivertebra might have been missed. More importantly, this case report itemizes successful conservative management which shall serve as database for future study in Nigeria. The limitation of this study is that it's impossible to establish a cause effect relationship and management options available cannot be generalized.

Implications of the findings

The diagnosis of giant hypertrophy of the fifth cervical vertebra spinous process with hemivertebra should be considered as a possible differential diagnosis of posterior neck swelling.

Conclusion

Hypertrophy of the spinous process is a rare cause of posterior midline neck mass and may be complicated by other associations or syndromes. It is usually an incidental finding. Imaging is the mainstay for diagnosis and should be utilized in evaluating its various associated anomalies and syndromes.

Declarations

Ethical consideration: The case report was conducted in compliance with the guidelines of the Helsinki declaration on biomedical research in human subjects and informed consent of the patient for publication was obtained.

Authors' contribution: OSM conceived and designed the study, contributed to data collection and manuscript writing. RMW contributed to data collection, data analysis tools, analysis of data, and manuscript writing, critical editing for intellectual approval OJE contributed to data analysis. LTO-O contributed to manuscript writing. All authors approved the final copy of the manuscript.

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