Case Report

Anesthesia in a Dwarf Lady- A Case Report

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Abstract

Anesthesia for individuals with dwarfism requires careful consideration due to potential anatomical and physiological variations. These variations can lead to challenges with airway management, regional anesthesia, and other anesthetic procedures. Preoperative evaluation is crucial to identify potential risks and plan for appropriate anesthetic management.

In this report, we present the case of a 33-year old dwarf lady, Diagnosed case of Fibroid Uterus and treatment plan is total abdominal hysterectomy. Who was scheduled for Total Abdominal Hysterectomy. Preoperative evaluation revealed Curved spine (kyphoscoliosis) and fixation of the *lumbo sacral spine, restricted movement of atlanto occipital joint, with a short neck with possible atlantoaxial instability. Special attention should be given to positioning during anesthesia to avoid undue strain on the spine and prevent atlantoaxial instability. Special attention and precautions must be taken before induction for general anesthesia with a view to rescue from difficult intubation or failed intubation. Should have adequate idea about difficult intubation drill. For example, keeping fiberoptic laryngoscope, LMA, Stellate etc.*

***Key words**: Dwarfism, Anesthesia.*

Introduction

Dwarfism, often caused by skeletal dysplasia, involves skeletal deformities and disproportionate short stature. This means the limbs and trunk don't grow at the same rate, leading to short limbs, a large head, and other characteristic features. Achondroplasia, the most common type, results from a gene mutation affecting bone growth, causing short limbs and a prominent forehead.

Dwarfism occurs when an individual person or animal is short in stature (less than 147 cm or 4 feet 10 inches) resulting from a medical condition resulting in abnormal (slow or delayed) growth[1].

In the United States, 2.5 percent of the population is short. The Utah Study, a population-based survey of growth in children, concluded that endocrine disorders constitute only a small number of cases with short stature. People of both sexes are affected, but males are more likely to be brought to medical attention because of the prevalence of social expectations[2].

Human height is related to a combination of genetics, hormones, nutrition, environment, and other factors. It is a complex process involving multiple genes and multiple factors. Genetic factors are the main factors affecting individual height differences, and the heritability of human height accounts for approximately 80%[3]. Dwarfism may also result from inadequate nutrition during crucial phases of growth and development. Malnutrition is a notable etiology of retarded growth in kids, especially in developing countries. Many efforts have been in process by the government and non-profit organizations to curb this[4]. A leading nutritional cause of dwarfism is vitamin-D-resistant rickets during childhood; intelligence is not affected by this condition. Here we reported a rare case of Total Abdominal Hysterectomy (TAH) in the dwarf lady with successful outcome.

Case Report

Mrs. Komola, a dwarf lady of 33 years, Muslim, housewife of lower class family was admitted in Gynae and obstetrics department of Kumudini Women's Medical College Hospital with diagnosed case of fibroid uterus. According to her statement, she has only one brother but died before 24 years back. She told her mother also dwarf lady and all of her family members are in average height and body built. She had regular menstrual cycle with average flow and duration but had delayed menarche at 17 years. She is married for 12 years, para-0+1(ab), 109 cm tall komola having developed desmenorrhoea for last 3 years .On G/E-Pulse (78b/min), BP(120/80 mmhg), Anaemia-mild, oedema –absent. Routine investigations including ECG & ECHO were found normal .On; P/S/E: Cervix –healthy, B/M/E-Uterus-Impacted on pelvis, 14-16 weeks size. An ultrasound measurement shows of fibroid uterus about (5.9 x 4.7) cm. She had no history suggestive of bronchial asthma, heart disease, chronic renal disease, malabsorption syndrome, diabetes mellitus or thyroid disorder. To find out the possible causes of dwarfism, thyroid function test and growth hormone assay were done and the results were within normal range. On clinical examination, she was a proportionate dwarf (body parts being proportional). Musculoskeletal system revealed low muscle bulk and bony mass, but muscle power was normal. Cardio-respiratory and other systems were normal.

On X ray shows Lumbo sacral spine: Curved spine (scoliosis or kyphosis): The spine may develop abnormal curves-kyphoscoliosis and fixation of the lumbo sacral spine, it is difficult to perform spinal anesthesia, restricted movement of atlanto occipital joint, so it is difficult to intubation. My plan of anesthesia was-spinal, if failure then general anesthesia. I failed to perform spinal anesthesia, then I gave general anesthesia, though it was difficult to intubate patient. But eventually I intubated. Initiating anesthesia by 0.5L Isoflurane, 4 Litre 0_2 and 1 Litre N_20. At the end of the operation I reversed the patient, complete reversal. But in the post- operative ward it was difficult to maintain 0_2 saturation. Without oxygen saturation was going down below 90% .I continued nasal 0_2 spraying first postoperative night, and next day approximately after 30-35 hours of operation, patients 0_2 saturation become normal.


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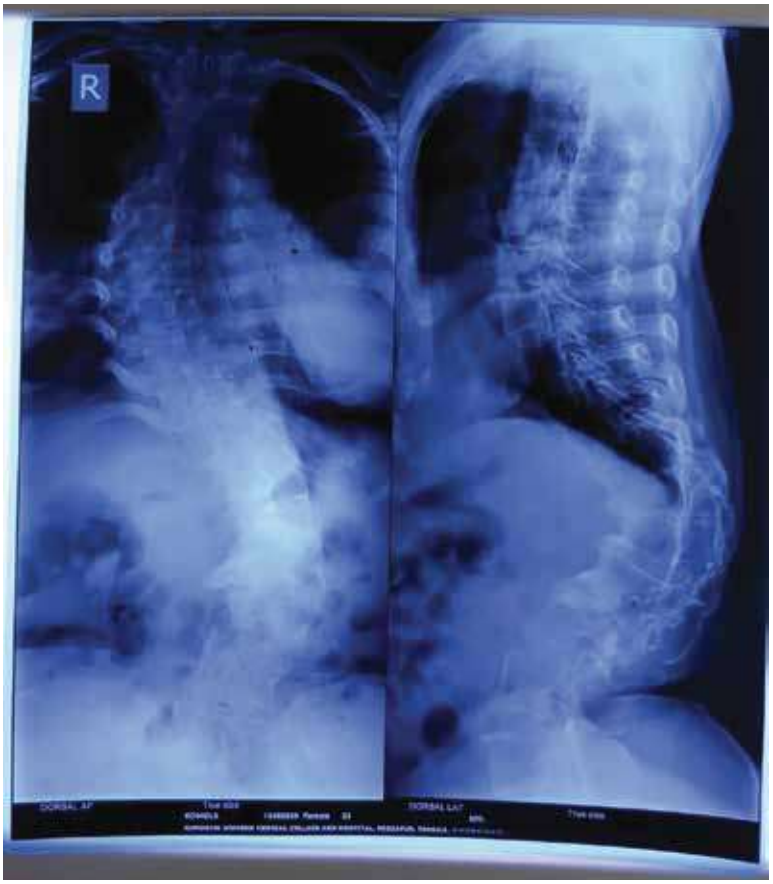
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Discussion

Anesthetic challenges in patients with achondroplasia include potentially difficult airways, often complicated by sleep apnea due to obesity, altered respiratory mechanics, and difficult neuraxial access with the unpredictable spread of local anesthetics. Restrictive lung disease and pulmonary hypertension can develop from chronic hypoxemia or hypercarbia secondary to thoracic scoliosis, airway obstruction, or sleep apnea. Preoperative arterial blood gas may be warranted in these cases.

Neuraxial access can be difficult due to kyphoscoliosis and narrow epidural space. The spine in patients with achondroplasia has several unique anatomical features, such as hypertrophy of superior and inferior articular facets, short and thickened pedicles of the vertebral body from premature fusion with scalloping on the posterior surface, prominent bulging of intervertebral discs, spinal canal tapering caudally from decreases in the interpedicular distance in the lumbar region (as opposed to widening caudally in normal anatomy), and thin dura[5]. These mechanical features can lead to spinal stenosis and nerve root compromise. In patients who had previously undergone lumbar decompression surgeries, scar tissue and altered anatomy can further complicate neuraxial techniques. Because of these expected anatomical challenges, we utilized ultrasound to facilitate access to the intrathecal space. Determining the dose and volume of local anesthetic is further complicated by accentuated lumbar lordosis, spinal stenosis, engorged epidural veins, and narrow epidural and intrathecal spaces, which can result in the unpredictable spread, iatrogenic dural puncture, difficult catheter placement, air embolism, or a high spinal/epidural block[5, 6].

In our case, however, the patient presented to the operating room for a relatively brief operative procedure, and spinal injection was chosen but if failed do perform general anesthesia. Given the risk for brainstem compression, preoperative screening for central obstructive apnea and cervicomedullary compression is critical for the prevention of sudden death from central respiratory failure[7].

In a prospective series of achondroplastic infants, radiographic evidence of craniocervical stenosis was present in 58% of patients and cervicomedullary compression in 35% [8]. Similarly, a 7.5% risk of sudden death in the first year of life was observed in a retrospective study of achondroplastic infants[9]. Presenting signs of cervicomedullary compression include upper or lower extremity paresis, apnea, cyanosis, hyperreflexia, hypertonia, abnormal plantar response, or delay in milestones versus other achondroplastic infants. Awake fiberoptic intubation may be the safest method of securing the airway in these patients if general anesthesia is needed but is not feasible due to a lack of patient cooperation, anxiety, and anatomical challenges secondary to smaller airways. As a result, in-line stabilization with video laryngoscopy may be a good alternative[10].

Dwarfism can be caused by about 200 distinct medical conditions; such that the symptoms and characteristics of individuals with dwarfism vary greatly[11]. Disproportionate dwarfism is characterized by one or more body parts being relatively large or small in comparison to those of an average-sized adult.

Achondroplasia is the commonest cause of short-limbed dwarfism and characterized by proportionately shorter limbs than the trunk, increased spinal curvature, and distortion of skull growth with characteristic facial features. It is a rare genetic disorder inherited as autosomal dominant trait. In cases of proportionate dwarfism, extreme shortness with proportional body parts usually has a hormonal cause, such as growth-hormone deficiency (also known as pituitary dwarfism), defect in receptor for growth hormone and Hypothyroidism. Growth hormone deficiency has no single definite cause. It can be caused by mutations of specific genes, damage to the pituitary gland, Turner's syndrome, poor nutrition[3], or even stress (leading to psychogenic dwarfism). Serious chronic illnesses may also produce dwarfism as a side effect. In cases of proportionate dwarfism the sexual development is often delayed or impaired into adulthood. Hypotonia, or low muscle tone, is common in dwarfs, but intelligence and lifespan are usually normal. It is rare for achondroplasia sufferers to conceive and when they do the pregnancy does not last long because of the uterus size [12, 13].

Conclusion

Dwarfism is one of the most common diseases in the endocrine system. This case represents the successful use of general anesthesia in an achondroplastic patient without neurologic sequelae. Managing the general anesthesia with successful intubation and reduced the

risk of airway obstruction in the postoperative period. Understanding the anatomical features associated with achondroplasia is critical for avoiding potential problems with airway management.

References

1. Dwarfism. FromWikipedia.Retrieved2009-02-22.

2. Grimberg A, Feemster KA, Pati S, Ramos M, Grundmeier R, Cucchiara AJ, et al. Medically underserved girls receive less evaluation for short stature. Pediatrics. 2011 Apr;127(4):696-702. [PMC free article] [PubMed]

3. Ornitz DM, Legeai-Mallet L. Achondroplasia: Development, pathogenesis, and therapy. Dev Dyn. 2017;246:291–309. doi: 10.1002/dvdy.24479. [DOI] [PMC free article] [PubMed] [GoogleScholar

4. Pérez-Ríos M, Santiago-Pérez MI, Malvar A, Hervada X. [Prevalence of malnutrition in Spanish schoolchildren]. An Pediatr (Engl Ed). 2019 Apr;90(4):259-260. [PubMed]

5. Carson BS, Groves M, Yassari R. Schmidek and Sweet's Operative Neurosurgical Techniques. Vol. 1. Amsterdam, Netherlands: Elsevier; 2012. Neurologic problems of the spine in achondroplasia; pp. 2091–2099. [Google Scholar]

6. Spinal anaesthesia for emergency caesarean section in an achondroplastic dwarf. Ravenscroft A, Govender T, Rout C. Anaesthesia. 1998;53:1236–1237. doi: 10.1046/j.1365-2044.1998.0716q.x. [DOI] [PubMed] [Google Scholar]

7. Foramen magnum stenosis and spinal cord compression in achondroplasia. Ok MS, Nagaraj UD, Mahmoud M. Anesthesiology. 2019;131:396. doi: 10.1097/ALN.0000000000002757. [DOI] [PubMed] [Google Scholar]

8. Cervicomedullary compression in young patients with achondroplasia: value of comprehensive neurologic and respiratory evaluation. Reid CS, Pyeritz RE, Kopits SE, et al. J Pediatr. 1987;110:522–530. doi: 10.1016/s0022-3476(87)80542-5. [DOI] [PubMed] [Google Scholar]

9. Mortality in achondroplasia. Hecht JT, Francomano CA, Horton WA, Annegers JF. https://pubmed.ncbi.nlm.nih.gov/3631079/ Am J Hum Genet. 1987;41:454–464. [PMC free article] [PubMed] [Google Scholar]

10. Spiegel JE, Hellman M: achondroplasia - implications and management strategies in anesthesia. [Jun;2021];Spiegel JE, Hellman M. https://www.anesthesiologynews.com/Review-Articles/Article/10-15/Achondroplasia-Implications-and-Management-Strategies-in-Anesthesia/33892/ses=ogst Anesthesia experts. 2015.

11. Kozma, Chahira. Causes of Dwarfism. American Journal of Medical Genetics 2005; 140A (4): 303–311.

12. Dwarfism: Symptoms. MayoClinic.com. MayoFoundation for Medical Education and Research. Retrieved 2009-01-29.

13. Ghumman S, Goel N, Rajaram S, Singh K.C,Kansal B, Dewan P. Pregnancy in an achondroplstic dwarf. Journal of the Indian Medical Association 2005; 103(10):536-538.