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Myositis Ossificans- For Hip Deformity Correction; Anesthetic Implications.

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Abstract : *Myositis ossificans also known as "fibrodysplasia ossificans progressiva" is a severe disabling connective tissue disorder uncommonly seen. Hetrotopic ossifications appear spontaneously or flare up on trauma. These patients present for deformity correction and other surgeries. We report a case of 27 year old female, known case of myositis ossificans posted for hip deformity correction under regional blockade.*
Keywords : *Myosistis Ossificans, Neuraxial Blockade.*

I. Introduction

Myositis ossificans is a rare inherited connective tissue disorder. It is characterized by congenital malformations of great toes, thumbs and vertebrae1.There is no gender, ethnic, racial predisposition2.It is also called as "The stone man syndrome" as there is progressive ossification of skeletal muscles. There are no reported cases performed under regional blockade.After taking written informed consent ,we report anesthesia management under regional blockade in case of myositis ossificans and discuss anesthesia implications due to challenges in positioning, airway, pulmonary function after taking patient's consent.

II. Case Discussion

A 27 year old female, 50 kg; presented with restricted lower limb movements and difficulty in walking since one year.The patient suffered from malarial encephalitis in 9th month of gestation, was tracheostomized and on ventilatory support. She underwent lower segment caesarean section. She was weaned off ventilator and the tracheal stoma was closed after two months. Later she developed progressive difficulty in walking. On examination, abduction movement at both hip joints were restricted with flexion at knee joints. Diagnosis of myositis ossificans was made based on history, x ray imaging and biopsy. On investigation, hemoglobin was 12 gm/dl, other laboratory investigations and ECG were normal. X–rays of chest, neck and lumbar spine revealed no abnormality. Pulmonary function test and indirect laryngoscopy were normal.

A neuraxial anesthesia was planned. Both limbs were planned to be operated in single sitting. The severely affected right limb surgery was initiated. Patient was wheeled inside theatre and regular monitors were attached; NIBP, cardioscope, pulse oximetre and temperature probe.

Two wide bore intravenous cannulas were positioned taking care to avoid tissue trauma. Positioning for neuroaxial blockade was difficult and technically challenging. Patient was taken on the edge of table with assistants holding her and lower limbs supported on stool. Epidural catheter was inserted at L1-L2 interspace followed by spinal blockade at L3-L4. 3cc of 0.5% hyperbaric bupivacaine and 50ug of fentanyl were used for spinal blockade. Sensory dermatome level upto T10 was achieved. The patient was positioned supine for surgery and the pressure points were padded.

Anesthesia was maintained with 0.5% bupivacaine epidural infusion at 5ml/hr, started an hour after spinal blockade. Sensory level was maintained at T10.Inj hydrocortisone 100mg was administered intravenously. Intraoperatively vitals were maintained as MAP 60-70 mm Hg, HR 60-70 /min, SpO298%.

Total blood loss was two litres. Hemodynamics were maintained with crystalloids, colloid and two packed blood cells. Urine output was maintained at 1ml/kg/hr. The duration of surgery was five hours.

Due to blood loss and prolonged duration, other limb surgery was postponed. Post operative analgesia was given in form of epidural top ups of 90ug buprenorphine 12 hourly for three days. Patient was comfortable with VAS score of three. The other limb was operated after a period of 2 weeks under regional anesthesia. The patient was discharged home after a week. The patient was followed up for a period of 6 months without any evidence of progression of disease.

III. Discussion

Myositis ossificans progressiva" also called as "fibrodysplasia ossificans progressiva" is an autosomally inherited connective tissue disorder with less than 400 cases reported worldwide. There is heterotopic ossification of skeletal muscles induced spontaneously or by trauma3. The muscles of body spared are the tongue, larynx, eye, diaphragm, sphincters and perineum 4.

Cardiovascular and pulmonary function status should be assessed. Changes seen in ECG are RBBB, T wave inversion and ST segment changes5. Ossification of TM joints leads to reduced mouth opening with difficult intubation like situation6. Cervical spine fusion with scoliosis is observed in certain patients.

Due to ankylosis of the costovertebral joints and ossification of the chest wall, the patients eventually develop restrictive respiratory failure. Atelectasis and ineffective cough often lead to recurrent pulmonary infections7.Our patient had past history of tracheostomy with ventilator dependency for a month, so to avoid further airway manipulation and pulmonary complications, neuraxial anesthesia was planned. As there was no involvement of spine, the only difficulty to overcome was positioning of patient for regional blockade. Efficient positioning lead to successful neuraxial blockade in first attempt. By planning neuraxial blockade, we avoided difficulties of general anesthesia in a previously tracheostomized and ventilator dependent patient.

Careful positioning and padding is needed to prevent soft tissue injury, which was adequately taken care in our case. If a flare-up develops in the postoperative period, ice application for 24 h and steroids are recommended. Intramuscular injections are to be avoided as they can lead to ossification at site. Any dysfunctional indwelling intravenous catheters should be removed as they may also lead to heterotopic ossification6.

IV. Conclusion

Myositis ossificans though rare disorder pose great challenges to anesthetist. As an anesthetist we should be aware of its various forms and progression and should be able to deal with all difficult situations. All efforts should be made to avoid situations that lead to new localizations of heterotopic ossification, as that may further impair quality of life. Neuraxial anesthesia does not always lead to heterotopic ossification around the needle area and can be administered weighing the risk benefit ratio.The disease affects multiple joints and vertebral column, hence positioning the patient can be difficult. Appropriate positioning thus increases the rate of successful neuraxial blockade.

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