Resident Research Night

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The Anesthetic Management of a Pregnant Patient with Multiple Pterygium Syndrome (Escobar Type)

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Multiple Pterygium Syndrome

• Rare heterogeneous disorder that can be either autosomal recessive or autosomal dominant.

• Has multiple subtypes depending on the combination of abnormalities.

• In general, it is characterized by:
  • **pterygia** (excessive webbing)
  • **arthrogryposis** (congenital contractures)
  • **ankyloglossia** (adhesions of the tongue to the palate)
  • **syngnathia** (congenital bands of tissue between the maxilla and mandible)
  • **cleft palate, micrognathia, and scoliosis** (1).

• Variably expressed features = short stature, craniofacial dysmorphism, arachnodactyly, and congenital respiratory distress.
Multiple Pterygium Syndrome
Escobar Type

• The Escobar subtype
  • webbing of the neck that increases with age
  • lumbar lordosis
  • webbing of the knees and elbows =before adolescence (2).

• Less than 70 cases of this syndrome reported in the literature.

• 1\textsuperscript{st} case report of a parturient with Escobar syndrome requiring general anesthesia for an elective cesarean section.
Multiple Pterygium Syndrome
Escobar Type

- Extreme Form
Our Patient
Multiple Pterygium Syndrome
Escobar Type

My Case

21 year old Caucasian female G1P0, 40 kg, and 144 cm tall.

35 weeks of gestation with contractions and subsequently premature rupture of membranes.

PMed/Surg Hx = significant for Escobar syndrome diagnosed in early childhood. - Cleft palate, - ASD (repaired as a young child)

- Multiple corrective orthopedic surgeries for her congenital scoliosis, with posterior spine fusion from L2 to the sacrum, including a sacral block fixation.

- Several bilateral knee and hip surgeries as well as lower extremity contracture releases.
Multiple Pterygium Syndrome
Escobar Type

Past Anesthetic History:
Of the available records = past GA was provided via an LMA #3 (uneventful)

Meds = PNV and PRN Tylenol.

Physical Exam = pterygium colli with ~ 30 degrees of leftward rotation and slight flexion of the neck.

Significant limitation of the cervical extension, micrognathia, and a repaired cleft palate.

Pharyngeal exam = Mallampati III with maxillary prognathism. The cricothyroid membrane was easily palpable and the thyromental distance was 3.0 cm.
Multiple Pterygium Syndrome
Escobar Type

Low nasal bridge and small nares measuring 0.4 cm diameter.

Chest = pectus carinatus with thoracic kyphoscoliosis. BS were clear bilaterally. Cardiac exam unremarkable. EKG = NSR. Recent ECHO = mild MVP, no residual intracardiac shunt (post ASD repair), and a left ventricular ejection fraction of 51%.

LABS = WNL for pregnancy.

Anesthetic Plan =
OBs opted for C/S due to the maternal pelvic abnormalities.

We considered general anesthesia as the optimal anesthetic. Neuraxial was not feasible due to her lumbar hardware.

The increased likelihood of a difficult intubation along with pregnancy and its associated risk of aspiration = Awake FOB
Multiple Pterygium Syndrome
Escobar Type

Aspiration prophy = Bicitra 30 ml PO, famotidine 20 mg IV, and metoclopramide 10 mg IV.
Topical = 5 mLs of nebulized 4% lidocaine.

OR =
Standard Monitors.

Peds Oto resident in room.

Abdomen prepped and draped.

IV sedation with midazolam (total = 4 mg IV) and ketamine ~2 mg/kg (total = 100 mg IV).

An oral fiberoptic bronchoscope revealed significant edema of the arytenoids and anterior displacement of the larynx.
Multiple Pterygium Syndrome
Escobar Type

Difficulty = degree of anterior flexion of the scope impossible to achieve without exaggerated “sniffing position”.

5.0 ETT placed (confirmed) and incision made after induction of general anesthesia with propofol 1.5 mg/kg.

Maintenance = 0.5 MAC desflurane in an oxygen and air mixture with a propofol infusion at 100 mcg/kg/min.

The heart rate (100-120 beats/min) and blood pressure (110-145/50-80 mmHg) remained stable with an SpO2 of 99% throughout the procedure.

Sedation to skin incision time interval = 4 minutes.
Sedation to uterine incision = 6 minutes.
A vigorous and apparently healthy male infant was delivered weighing 2035 grams.

Apgar scores = 8 and 9
Multiple Pterygium Syndrome
Escobar Type

Fentanyl 3 micrograms/kg given after the baby was delivered.

No muscle relaxants were used.

Prior to emergence from general anesthesia, we performed a direct laryngoscopy with a 2.0 Miller blade = Grade 2 visualization of the larynx.

The patient was extubated in the operating room and recovered uneventfully in the post anesthesia care unit.
Multiple Pterygium Syndrome
Escobar Type

• First case involving the anesthetic management of a pregnant patient with multiple pterygium syndrome (Escobar type) who underwent general anesthesia for an elective cesarean section delivery.

• Previous reports involving GA were adequately NPO and younger than 40 months of age. Our patient = full stomach and 21 y/o.

• Escobar syndrome in the pregnant patient can pose several anesthetic challenges: difficult airway anatomy

• unpredictable reaction to depolarizing and non-depolarizing muscle relaxants

• possible association with malignant hyperthermia

• possible undiagnosed congenital cardiac anomalies

• difficult neuraxial blockade due to either anatomical deformities or prior spine surgeries requiring hardware implantation.
Multiple Pterygium Syndrome
Escobar Type

- **Airway**
  Obviously needs to be secured with ETT.
  History of LMAs = no direct visualization of the larynx since her palate and ASA repair. Syndrome can include webbing in and around the larynx.
  Even with good view . . .

Most Concerning = With increasing size and age . . . Overall decreased cervical mobility.

How did the asleep DL change our thoughts?
The validity = low predictive value = “stenting effect”
Multiple Pterygium Syndrome
Escobar Type

What did we have available?
Assortment of laryngoscope blades
✓ LMAs
✓ a bougie
✓ an LMA-Fastrach™
✓ a jet ventilator
✓ pediatric cricothyrotomy kit.

5.0 ETT

Small nares = oral approach only.

Alternate approach = Fastrach™ LMA
Multiple Pterygium Syndrome
Escobar Type

Couple of reasons muscle relaxants not used.

1\textsuperscript{st} = relaxation not needed for the surgical procedure.

2\textsuperscript{nd} = None used in past available anesthetics and the pharmacodynamics are potentially altered (5).

The safe use of vecuronium, however, has been reported in this population (1).

3\textsuperscript{rd} = previous isolated report of malignant hyperthermia associated with Escobar syndrome (4) = questionable and no specific recs in lit
References


