PERIOPERATIVE MANAGEMENT IN PATIENT WITH Mc CUNE-ALBRIGHT SYNDROME WITH PATHOLOGICAL PROXIMAL FEMUR FRACTURE.

[1*]**Dr.Akanksha Patil**, [1]Department of Pharmacy Practice, Krishna Institute of Pharmacy, Krishna Vishwa Vidyapeeth (Deemed to be University), Karad, Maharashtra -415539,
[1]**Jisha Annie Geevarghese** Department of Pharmacy Practice, Krishna Institute of Pharmacy, Krishna Vishwa Vidyapeeth (Deemed to be University), Karad, Maharashtra -415539
[1]**Dr.Gaurav Memane**,Sahyadri Super Speciality hospital Nashik Maharashtra

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Corresponding author
Dr.Akanksha Patil, Department of Pharmacy Practice, Krishna Institute of Pharmacy, Krishna Vishwa Vidyapeeth (Deemed to be University), Karad, Maharashtra -415539

KEYWORDS
McCune Albright syndrome, polyostotic/monostotic fibrous dysplasia (PFD), cafe-Aulait skin pigmentation, autonomous endocrine hyperfunction

ABSTRACT
The McCune- Albright syndrome is a rare disease diagnosed by the clinical triad of fibrous dysplasia (monostotic or polyostotic), cafe-au lait skin pigmentation, and endocrine hyperfunction. Those patients with bone issues mainly involve long bones and craniofacial anomalies. A 9-year-old male having pathological proximal Left femur fracture with hyperthyroidism, hyperthyroid induced hyperthermia with precocious puberty posted for internal fixation and open deduction for the same fracture. The procedure was done under neuraxial anesthesia without any complications.

INTRODUCTION

McCune Albright syndrome is an uncommon disease first reported in 1937 separately by Donovan McCune & Fuller Albright with an estimated prevalence between 1/1,00,000 & 1/10,00,000 [1]. MAS is a genetic and nonhereditary syndrome comprised of a minimum of two of the subsequent three features: (1) polyostotic/monostotic fibrous dysplasia, (2) cafe-au lait skin pigmentation, and (3) precocious puberty. Other endocrine syndromes could even be present, including acromegaly, hyperthyroidism, Cushing syndrome, and Mazabraud syndrome (myxomas) [2,3,4,5,6]. Fibrous dysplasia is observed in Femur (91%), tibia, pelvis, and cranio facial region[7,8].Non-endocrine abnormalities like cardiac arrhythmias, sudden cardiac death, and chronic liver disease also can be present [9]. In MAS, patients have pathological fractures or post-traumatic fractures secondary to trivial injury as the osseous complex is replaced by fibrous tissues [10]. MAS is the result of a sporadic, early postzygotic somatic mutation within the GNAS1 gene at locus 20q13.113.2, coding for the G protein subunit Gs alpha [11].

CASEREPORT

A 9-year-old, 30 kg, 130 cm male child was admitted with a history of slip and fall leading to pain, swelling over the left thigh, and inability to bear weight on the left lower limb.In birth history,he is the first child born out of non-agnate marriage with a normal younger sibling. The patient had fibrous dysplasia, diagnosed at the age of 4years when parents noticed abnormal gait and walking difficulty.Orthopedic consultation was done and started on oral calcium and vitamin supplements.

On preoperative assessment, it was noticed that child is having cafe-au-lait skin pigmentation (brown) on the left side of the chest wall antero laterally extending from T 2 to T6 level along with enlarged testicles and penis. The child was having tachycardia (heartratewas115/m),a VAS score of 7/10.Nocraniofacial abnormality was noted. The airway on assessment was normal. No obvious swelling in the neck was

present. Liver and renal function tests along with complete blood counts were normal. Thyroid function tests, ECG, serum electrolytes with calcium and magnesium levels, urine routine were advised.
A femoral nerve block was given with15cc of 0.2%Ropivacaine with 4mg Dexamethasone for analgesia.

On day 1 of admission, tachycardia continued despite a VAS score of 3/10. Serum electrolytes were normal. The urine routine was normal, and ECG showed sinus tachycardia. Thyroid function test showed a hyperthyroid picture with TSH - 0.007 µcg IU/ml, free T3 - 12.2 pmol/l, free T4 - 2.16ng/dl. The endocrinologist assessed the case and started tablet Propranolol 20mg TD Sandtablet Carbimazole10mg BD and he ordered for FSH, LH,and testosterone levels which came out to be increased testosterone 36.3-µg/ml, decreased LH- 0.10 mIU/ml, and decreased FSH- 0.56 mIU/ml levels. On day 3 of admission patient developed a fever of 102^0F with tachycardia of heart rate120/m. The pediatrician diagnosed it as thyrotoxicosis induced hyperthermia and advised sleeping heart rate monitoring on ECG and started him on antipyretic tablet Ibuprofen 200mgTDS. The endocrinologist reviewed the case and increased the dose of tablet Propranolol 40mg QID.

ECGshowedTwaveinversioninanteriorleadswithsinustachycardiaforwhichcardiologistopinionwas taken, 2D echo was done which showed ejection fraction 60%, mild pulmonary hypertension (PAH) of 33mmHg, normal systolic and diastolic function with doubtful ASD/PFO. Infective endocarditis prophylaxis with Inj. Cefuroxime 1.5 g and Inj. Amikacin 500mg 1 hr before surgery was advised.
For the next 2days, heartrate was monitored 2hourly for 24hrs which settled to 90-100/m,repeat thyroid function tests showed TSH-0.009µIU/ml,FreeT3-10.66pmol/l,freeT4-2.16ng/dl.The endocrinologist gave clearance for surgery with a high risk of thyroid storm.
On the day of surgery, informed written consent for curettage and open reduction internal fixation with a bone allograft of left proximal femur fracture to be performed under spinal and epidural anesthesia was taken with high-risk consent of intraoperative thyroid storm.
Infective endocarditis prophylaxis was given with Inj. Cefuroxime 1.5 gm and Inj. Amikacin 500 mg iv. Sedation was givenwithInj.Fentanyl60µcgiv.Inj.Dexamethasone4mg iv,Inj.Paracetamol500mgiv,
Inj. Ketorolac 15 mg iv was given. Under all sterile conditions in sitting position, spinal anesthesia was given after local anesthesia infiltration with 25G Quincke's needle with 2cc of 0.5% (H) Bupivacaine at L3-L4 level and the epidural catheter was placed with 19G Tuohy`s needle at L3- L4 level and fixed at mark 9cm.

Intraoperative surgery was uneventful, and vitals were stable with a heartrate between 90-100bpm. Blood loss was 150ml. Postoperatively epidural pump was started with Inj.Bupivacaine0.125%andInj.Fentanyl 1µg/cc at 3 ml/hr and analgesics Inj. Paracetamol 500mg IV TDS and Inj. Ketorolac 15 mg IV BD was continued for1 day followed by tablet Paracetamol 500mgTDSand tablet Aceclofenac50mg BD.VAS-score less than 3 was maintained. The epidural catheter was detached on day 2 and non-weight-bearing mobilization was started. The patient was discharged on day 5with the continuation of antithyroid drugs, bisphosphonates, and regular endocrinologist, cardiologist and orthopaedician follow up.

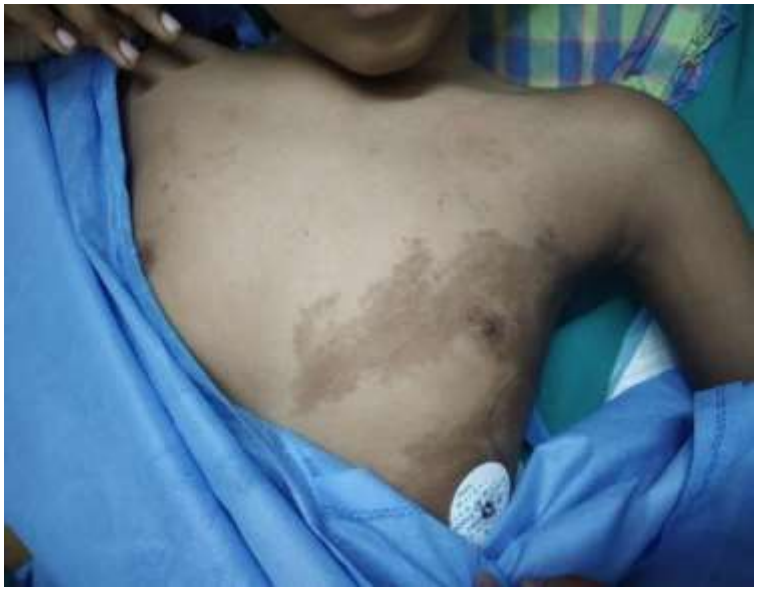

Fig.1

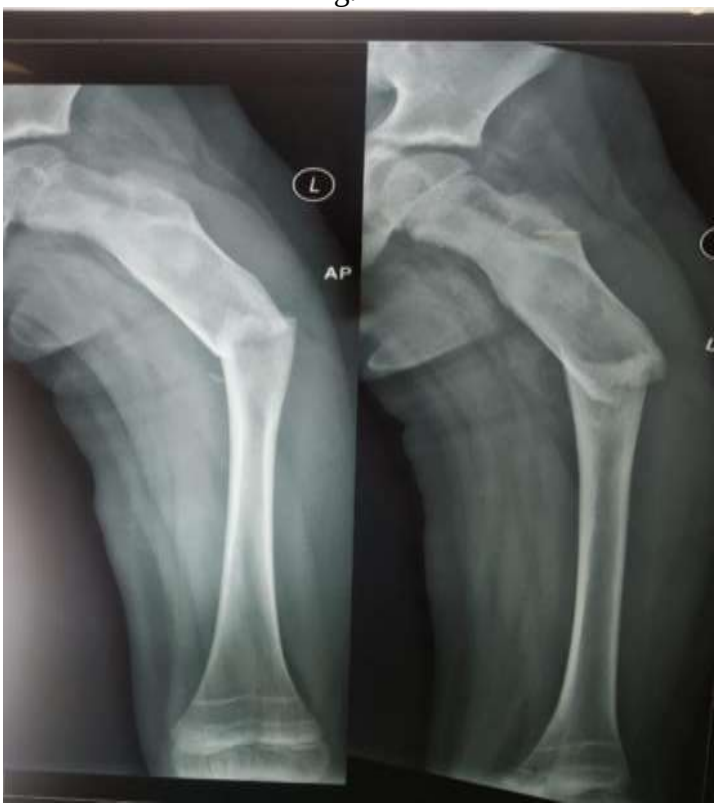

Fig.2

CASE DISCUSSION

Patients with thyrotoxicosis given out for surgery should preferably be made clinically and biochemically euthyroid before surgery, so on reduce the danger of perioperative thyroid storm. The danger of perioperative thyroid storm is often elevated following an acute event like surgery, trauma, pain, or infection[12].Patients with hyperthyroidism who aren't adequately clinically prepared for surgery are at serious risk [12]. Elective surgery and treatment should be postponed until the patient becomes euthyroid. Treatment typically lasts for a minimum of seven to ten days, according to the half-life of free T4 [12]. Preoperative Propranolol comprised the first-line treatment within the present case along with antithyroid medications [13]. MAS requires perioperative management by anesthesiologists, endocrinologists, orthopedics. The anesthesiologist should select the most appropriate method with the risks and benefits of both general and regional anesthesia with the preparation of hyperthyroid crisis. In patients with MAS polyostotic fibrous dysplasia, acromegaly, cervical spine instability, large neck swelling poses airway management difficulties and positioning difficulties. There is an altered stress response to general anesthesia.risk of cardiac arrhythmias,sympathetic response to tracheal manipulation,thyroid Storm offers a variety of challenges to anesthesiologists while giving general anesthesia.These challenges can be avoided to some extent by administering regional anesthesia whenever possible.

CONCLUSION

In the present case though, regional anesthesia was the preferred option as it minimizes stress response to surgery, provides better analgesia which will limit sympathetic and stress response to surgery. MAS is less often in boys than girls. In this case, the patient was having treatment for precocious puberty (symptoms) but the treatment for it that is testolactone, antiandrogen, and aromatase inhibitors like spironolactone or flutamide were not given. Another missing parameter was vitamin D report, phosphate level, and radiological examination which is required for the assessment of limb deformity for prophylactic surgery. Antithyroiddrugs, bisphosphonates, regular endocrinologist, and orthopedics follow-up are recommended to all patients postoperatively.

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