

## Peri Operative Management of a Case of Paget's disease with Hemorrhagic Shock: About a Case

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### Abstract

### Case Report

Paget's disease of bone (PD) is a rare disease with late onset whose current prevalence reaches, after the age of 55, PD is characterized by bone fragility, hypertrophy and deformities of the affected bones, caused by foci of increased bone remodeling that contain overactive and large osteoclasts. The consequences are: thickening, decrease in its flexibility, hypervascularization, pathological fractures. Often asymptomatic, Paget's disease never affects the whole of the skeleton but each bone can be affected, it affects in order of frequency the pelvis, the sacrum, spine, femur and skull. We report here the peri operative management of a case of Paget's disease who presented with hemorrhagic shock during intermediate hip prosthesis for femoral neck fracture.

**Keywords:** Paget's disease and anesthesia, hemorrhagic shock, fracture.

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## INTRODUCTION

Paget's disease of bone (PD) is a late-onset condition whose current prevalence reaches, after the age of 55, up to 3% of the population in Europe, North America, Australia and New Zealand [1]. This disease, more common in men, sees its prevalence increase with age to exceed 6% after 80 years. It therefore remains, by its frequency, the second metabolic bone disease after osteoporosis. PD is characterized by bone fragility, enlargement and deformities of the affected bones, caused by foci of increased bone turnover that contain overactive and large osteoclasts. A few years ago, the observation of a secular decline in the prevalence and severity of PD raised the question of the possible role of environmental factors in this evolution [1].

We report here the perioperative management of a case of Paget's disease who presented with hemorrhagic shock during an intermediate hip prosthesis. Institutional ethics board clearance and patient consent were obtained prior to reporting.

## CASE REPORT

A 63-year-old woman, 148 cm, 86 kg, type 2 diabetic on insulin since, ASA 2, This present for support of a fracture following a fall from its height.

She was diagnosed with a fracture of the upper end of the femur (Fig 1).



Figure 1: Chest radiology for pre-anaesthetic assessment

The general examination finds a conscious patient, normocolored connectives and a blood pressure with 120/60mmHg. auscultation: without anomalies. Chest X-ray (Figure 1) and ECG: unremarkable.

X-ray of the lumbosacral spine: aspect in favor of Paget's disease with deformation of the lumbar spine and diastasis fracture of the left femoral neck with ipsilateral diaphyseal ascent, site of two fracture lines (Figure 2).



**Figure 2: X-ray of the hip showing a diastasis fracture of the femoral neck**

His hemoglobin was 12.2 g/dl, serum urea - 25mg/dl and creatinine – 6 mg/dl. The coagulation profile, blood glucose and electrolytes were normal.

Mouth opening was adequate but neck extension was limited with class 3 mallampati. so we decided to go for general anesthesia . The risks were explained to him and informed written consent was obtained.

Pre-induction monitoring included electrocardiogram (ECG) leads II and V, invasive blood pressure, heart rate (HR), and peripheral oxygen saturation (SpO<sub>2</sub>). Venous access was established at the help of a large caliber VVP and glycemic monitoring every hour.

On the table, she received intravenous propofol (150mg) and fentanyl (200 µg). After checking for adequate ventilation inj. Rocuronium (0.6 mg/kg) was intubated using a video laryngoscope. The tube was fixed after checking bilateral air entry and the right internal jugular vein was cannulated. Anesthesia was maintained with Air:O<sub>2</sub>: 50:50 , sevoflurane 1–2% blood was about 1200 ml.

The duration of the surgery was four hours. The patient was extubated on the table after stabilization and weaning from norepinephrine and transferred to intensive care for monitoring and additional management. She was discharged from the hospital on the 7th postoperative day.

### 3. DISCUSSION

Paget's disease is a bone remodeling and remodeling disorder of unknown etiology but likely caused by infection with a common and widespread virus superimposed on genetic variation. Excessive breakdown and formation of bone tissue leads to weak bones leading to pain, bone deformities, fractures and arthritis in the joints near the affected bones [2].

It is marked by anarchic bone remodeling associating active bone resorption (osteoclastic activity) followed by excessive bone formation (osteoblastic activity). The consequences are: thickening, decrease in its flexibility, hypervascularization, pathological fractures [3, 4].

#### Description

Often asymptomatic, Paget's disease never affects the whole of the skeleton but each bone can be affected, it affects in order of frequency the pelvis, the sacrum, the spine, the femur and the skull. Pain is the main symptom.

- \* Spine: spinal cord compression, spinal root compression, kyphosis, atlo-occipital instability.
- \* Skull: enlarged with headache, hearing loss, optic nerve neuropathy, all cranial nerves may be compressed, cerebellar compression in the posterior fossa, hydrocephalus, hypertrophy of the maxillary bone.
- \* Deformities of the upper and lower limbs.
- Increased cardiac output linked to hypervascularization, with arterial hypertension, end-stage heart failure is exceptional.
- Calcemia is rarely high, if it is, this increase is discreet, serum alkaline phosphatase, witness of osteoblastic activity, and 24-hour hydroxyprolinuria, witness of osteoclastic resorption are high.
- Rare complication: sarcomatous degeneration.
- Treatment: antiosteosclastics: calcitonin, bisphosphonates (some of them inhibit mineralization and promote fractures). Mithramycin is prescribed in case of hypercalcemia and in refractory bone pain, it is responsible for nausea, vomiting, platelet function abnormalities, liver and kidney toxicity [5, 6].

#### Major Issues:

- Pathological fractures
- Neurological compressions
- Intubation difficulties
- Cervical instability
- Increased cardiac output

**Anesthetic Implications:**

- No contraindications
- Assess cardiac function: ECG, cardiac ultrasound,
- Respiratory assessment
- Assess the risks of neck instability, spinal damage, neurological compression
- Consider treatments and their consequences
- Intubation can be made difficult due to deformed jaws, caution during laryngoscopy due to
- cervical instability
- Evaluate serum calcium, phosphorus
- Pay attention to position on the table
- Beware of bleeding: hemorrhagic bone
- Landmarks for ALR can be made difficult by medulla.

**CONCLUSION**

In conclusion, our experience with this patient suggests that success in these patients depends on a thorough preoperative assessment, optimization of cardiac status, a perioperative plan including prompt diagnosis and management of complications as well as appropriate postoperative monitoring. However, intensive invasive monitoring can be individualized according to patients.

**Conflict of Interest:** We have no conflicts of interest to declare.

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