Subarachnoid block in a patient with Machado-Joseph disease

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Background and Aims

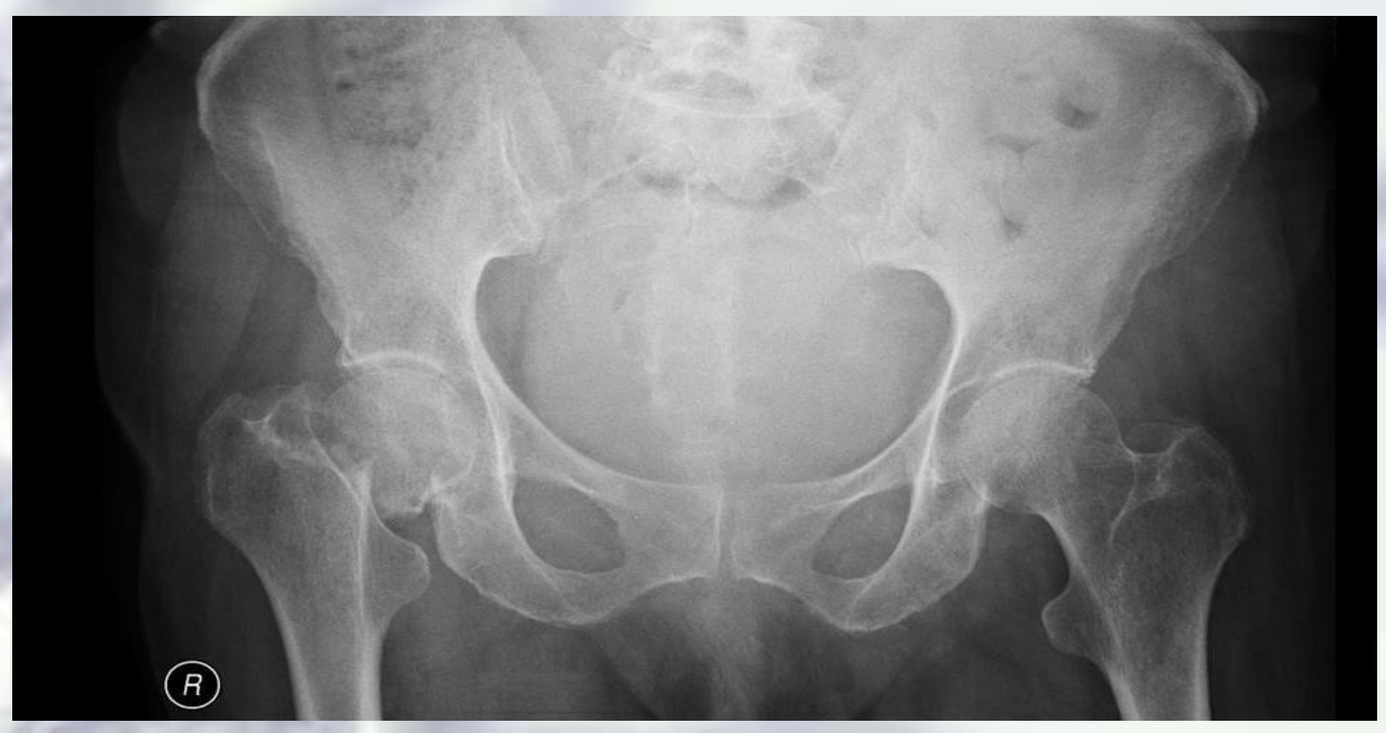
Machado-Joseph disease (MJD) or spinocerebellar ataxia 3 is a rare, autosomal, dominantly inherited neurodegenerative disorder characterized by varying degrees of ataxia, ophtalmoplegia, peripheral neuropathy, pyramidal dysfunction and movement disorder.

The use of regional techniques in patients with preexisting central nervous system disorders has been controversial, and the patients with MJD are no exception given the concern of worsening the neurological symptoms. In the other hand, an increased risk of pulmonary aspiration is reported in these patients during general anesthesia.

Results

A 77-year-old female patient with MJD is proposed for a right hemiarthroplasty due to fracture of the femur.

The MJD disease was diagnosed 3 years ago and is manifested mainly by imbalance, lower limb strength and muscle fasciculations. She had also previous medical history of high blood pressure and diabetes mellitus.



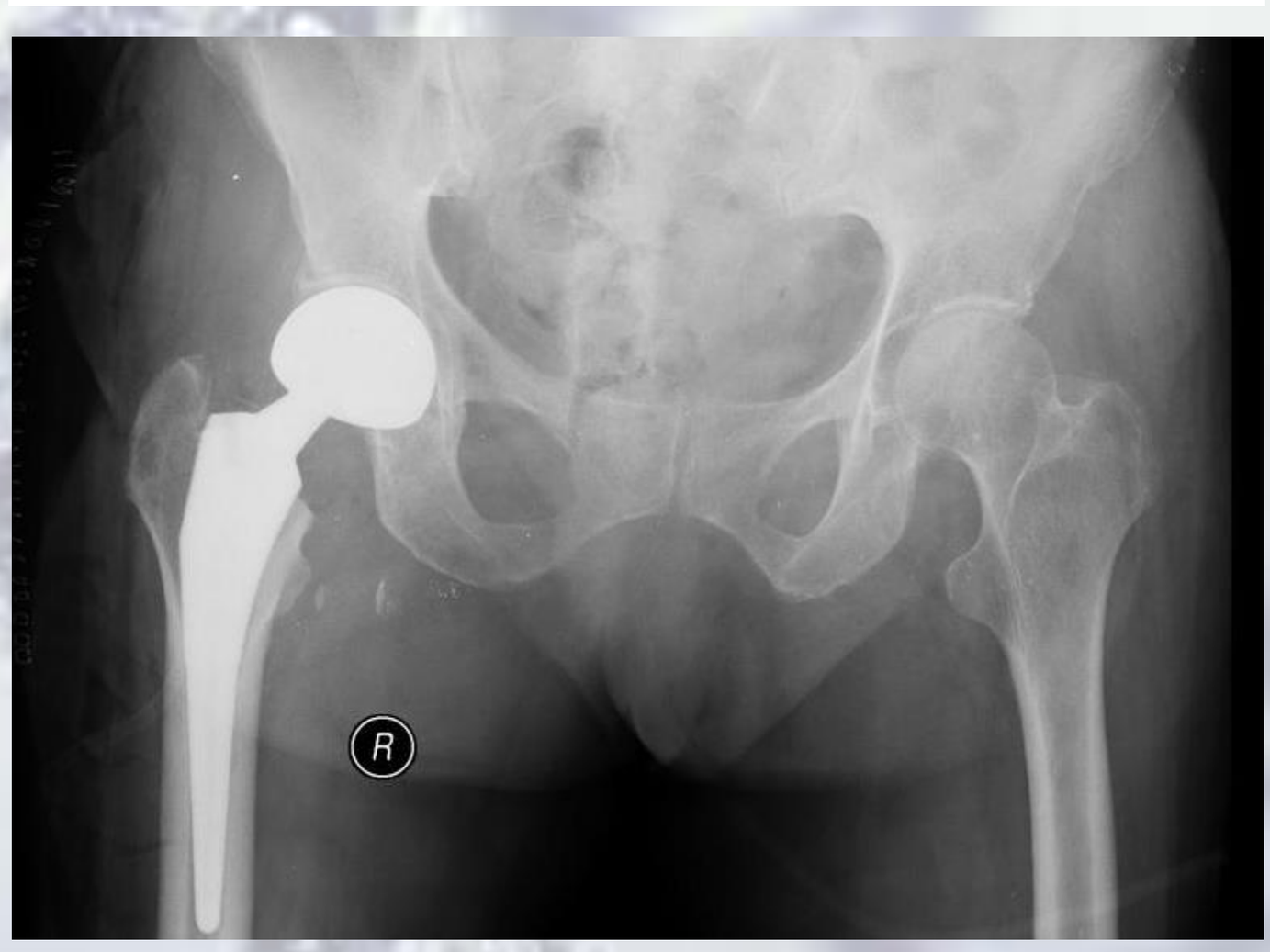
After informed consent, subarachnoid blockade was performed with 10mg of bupivacaine and 0.025mg of sulfentanil.

Sensory block reached a T10 level.
Surgery proceeded uneventfully.

There was full recovery of block by the third postoperative hour.

General anestesia was avoided regarding the increased risk of of aspiration reported in these patients, unpredictable response to muscle relaxants and the risk of ventilatory depression.

In the other hand, regional anestesia avoid airway and ventilation related problems and is associated with better postoperative analgesia.



Conclusions:

Considering the risks of general anesthesia in these patients, central neuroxial anesthesia should be an option for patients with MJD presenting for lower extremity operations.

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