

Severe visceral injuries following consecutive births : A possible link to underlying peripheral neuromuscular disorder “Charcot Marie Tooth Disease”?

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Objective

Charcot-Marie- Tooth (CMT) disease is a group of hereditary sensory and motor neuropathy resulting from demyelination, axonal dysfunction or both. We present an interesting case of CMT, where patient developed ureteric injury and Ogilvie syndrome following successive childbirths. We suspect whether these complications are potentiated by neuromuscular pathology secondary to CMT.

Case report

Our patient, with background of CMT, had an uneventful Kiwi omnicup delivery due to maternal exhaustion in her first pregnancy.

On the third day following delivery, she developed constant abdominal pain and CT urogram confirmed right ureteric rupture. She did not have any renal issues during antenatal period and intrapartum bladder care was appropriate. She underwent retrograde pyelogram which showed tortuous right ureter with mid-ureteric injury and stenting performed. The stent was removed two months later with no further complications.

Five years later, in her second pregnancy, she required full dilatation caesarean section following failed Forceps for suspected fetal compromise. The Caesarean section was complicated with bilateral broad ligament tears and Postpartum haemorrhage of 1.4 Lt. Postnatally, she developed Ogilvie syndrome with significant colonic dilatation and required loop colostomy on day 4 postnatal. She recovered well and reversal of colostomy performed after 2 months.

Discussion

CMT is one of the most common hereditary neuromuscular disorders affecting the peripheral nervous system. Usually autosomal dominant inheritance pattern but can also be autosomal recessive or X-linked. Most of the clinical symptoms are those related to walking, gait and balance secondary to motor and sensory loss causing muscle wasting and weakness. Among the many types of CMT, CMT2 results from abnormalities in the axon of the peripheral nerve cell, whereas CMT1 is due to the involvement of myelin sheath. Our patient did not have significant motor or sensory neuropathy but developed pelvic girdle pain in second pregnancy. Although uncommon, Ogilvie syndrome may happen following Caesarean section, but ureteric rupture following childbirth is one of the rare complications. Therefore, our strong suspicion that the underlying neuromuscular weakness could be linked to the recurrent complex visceral injuries.

Conclusion

Exacerbation of CMT disease can occur in pregnancy and they may have more operative deliveries, malpresentations and postpartum bleeding than the general obstetric population. Multidisciplinary team management is advised where appropriate involving obstetricians, neurologists, geneticists, anesthetists and midwives. Mode of delivery following previous outcome could have been more carefully considered in this instance.

References :

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