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Beware of lethal wernicke's encephalopathy after cytoreductive surgery with hipec for peritoneal pseudomyxoma: Case report of morbidity and mortality review

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Background: Pseudomyxoma peritonei (PMP) arising from the appendix is a rare entity. Complete cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC) is the only established curative treatment, and is reputedly linked to high morbidity and mortality.

We report, to our knowledge, the first case of delayed lethal Wernicke encephalopathy (WE) complicating CRS with HIPEC for an appendicular PMP. WE, caused by a thiamine deficiency, is characterized by ataxia, nystagmus and changes in consciousness.

Methods: A patient underwent complete CRS with HIPEC for a low grade mucinous appendicular tumor at the stage of PMP with a peritoneal index of 31, and was readmitted at POD 36 for persistent vomiting and vague neurological symptoms of mental confusion. The classic triad of WE appeared tardily. Although thiamine substitution was promptly applied, the patient died at POD53.

Conclusion: WE is an uncommon and severe neurological disorder with a mortality rate up to 20 % and only 16 % of treated patients can fully recover. This diagnosis should always be anticipated in patients undergoing major surgery such as CRS-HIPEC. Efficient treatment should be quickly introduced in order to avoid a lethal outcome

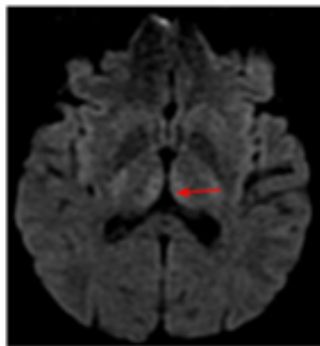


Figure 1: Cerebral MRI images showing hyperintense signal of the periaqueductal gray matter.

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