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Mis-diagnosed giant ovarian cyst and ventriculoperitoneal shunt malfunction

Kyste ovarien géant mal diagnostiqué et dysfonctionnement d'un shunt ventriculo-péritonéal

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Running title: Shunt malfunction related to ovarian pseudocyst

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Highlights: Abdominal pseudocysts are common complications of ventriculoperitoneal shunt, but rare differential diagnoses may be occasionally encountered. To our knowledge, this is the first description of shunt dysfunction related to cystic ovarian tumor. The interest is also to show the potential role of endoscopic ventriculocisternostomy in shunt dysfunction.

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Abstract

Abdominal pseudocysts are common complications of ventriculoperitoneal shunt (VPS). We report the case of a 37-year-old woman treated by VPS for congenital obstructive hydrocephalus, who presented shunt dysfunction related to a voluminous abdominal cyst initially diagnosed as cerebrospinal fluid pseudocyst. The cyst was drained and the VPS was removed after endoscopic third ventriculocisternostomy (ETV). A few months later, a large new abdominal cyst appeared and was operated on. Diagnosis was rectified as massive ovarian mucinous cystadenoma. In any intra-abdominal cyst, differential diagnoses need considering to avoid mis-diagnosis in shunted patients, especially if the cyst is very large. The etiology of the hydrocephalus should also be investigated in case of shunt dysfunction. Even in case of longstanding shunt, ETV can be an alternative to shunt revision surgery in obstructive hydrocephalus, enabling VPS withdrawal and treatment of the hydrocephalus.

Résumé :

Les pseudo-kystes abdominaux sont des complications fréquences des dérivations ventriculo-péritonéales (DVP). Nous présentons ici le cas d'une patiente de 37 ans traitée par une DVP pour une hydrocéphalie obstructive congénitale et présentant un tableau de dysfonctionnement de valve en relation avec un volumineux kyste abdominal initialement considéré comme un pseudo-kyste de LCS. Le kyste a été drainé et la DVP a été retirée après réalisation d'une ventriculo-cisternostomie endoscopique. Quelques mois plus tard, un nouveau kyste abdominal, volumineux, est réapparu dont la patiente a été opérée ; le diagnostic de cystadénome mucineux ovarien a été fait. Devant toute collection intra-abdominale, surtout massive, dans les suites d'une dérivation, une démarche diagnostique systématique doit être adoptée pour éviter une erreur d'orientation diagnostique. Il est également souhaitable, devant un tableau de dysfonctionnement de valve, de retrouver l'étiologie de l'hydrocéphalie. Même chez un patient valvé depuis très longtemps, la ventriculocisternostomie endoscopique peut être une alternative à la révision de valve en cas d'hydrocéphalie obstructive, permettant de sevrer le patient de la DVP et de traiter l'hydrocéphalie.

1. Background and importance

Ventriculoperitoneal shunt (VPS) is the commonest answer to long-term hydrocephalus. It is associated with a high rate of complications, estimated at 17-33% [1]. Dysfunction may be due to malfunction due to blockage, disconnection, migration or infection, in turn due to abdominal effusion. These have various causes, developing in any abdominal organ, and can be very large. In females, ovarian cysts are frequent, due to the high activity of ovarian tissue. Abdominal effusion can also develop in the peritoneum itself in case of adherence, due, for instance, to previous infection or surgical intrusion. In these cases, this abdominal effusion is called pseudocyst.

One such is cerebrospinal fluid (CSF) abdominal pseudocyst. CSF pseudocyst is sometimes symptomatic, mostly with abdominal symptoms in adults [2,3,4]. It can lead to shunt dysfunction and acute hydrocephalus.

2. Clinical presentation

A 37-year-old epileptic woman was implanted with a VPS at the age of 1 year for a cerebral acqueductal stenosis. The reference CT-scan showed small ventricles (Fig 1A). She had complained of abdominal heaviness for about 6 months when she was admitted for vomiting and impairment of consciousness, with Glasgow coma scale 8. Clinical examination revealed a dilated abdomen. Cerebral CT revealed large tri-ventricular hydrocephalus (Fig 1B). Abdominopelvic CT revealed a very voluminous intra-peritoneal effusion pushing back the viscera (Fig 1C) with a catheter that remained extra-cystic. At this point, the team suspected abdominal pseudocyst.

The patient was sedated and intubated for intracranial hypertension and respiratory difficulty due to increased abdominal pressure (Fig 1D and E). To resolve the hydrocephalus, the peritoneal catheter was externalized; the collected CSF was sterile. Paracentesis of the abdominal cyst was also performed, collecting 15 liters of clear sterile liquid. A week after onset of dysfunction, endoscopic third ventriculocisternostomy (ETV) was performed (Fig 1F). Postoperative imaging showed small ventricles and ETV permeability (Fig 1G and H). The externalized VPS was removed secondarily.

When she awoke, the patient presented only isolated cognitive slowdown. She was admitted to a rehabilitation center and radiological and clinical follow-up was instituted.

Eight months later, the patient was once again admitted for abdominal heaviness and pain, progressing for 1 month. Initial exam revealed dilated abdomen without symptoms of intracranial hypertension. Abdominal effusion was again seen on CT (Fig 1I), as voluminous as previously. Pelvic MRI suggested

ovarian origin of the cyst. Laparotomy was performed, revealing a 14-liter cyst developed from the right ovary. Appendectomy was performed, and the cyst wall was removed despite strong atypical adhesion to the peritoneum. Anatomopathological analysis diagnosed mucinous cystadenoma, with no risk of malignancy.

3. Discussion

Abdominal pseudocyst is a rare complication of VPS, at 1-4.5% of VPS complications [2,3], or slightly higher in the pediatric population. All factors modifying CSF fluidity (higher proteins content) or peritoneum absorption capacity (abdominal surgery, peritonitis or intra-abdominal infection, including CSF or peritoneal catheter infection, inflammation and immunoallergic phenomena, multiple shunt revision) may induce pseudocyst [2,3,5]. Most large unilocular cysts are sterile, and most small or multilocular cysts are infected.

The symptoms associated with pseudocysts are varied. Adults mainly present abdominal symptoms: abdominal heaviness, pain or increased abdominal perimeter [2,3,4]. Symptomatic shunt dysfunction, with neurological impact, is of late onset, seen only in 30% of cases [2]. In case of pseudocyst, CSF is analyzed from the pre-shunt reservoir, and abdominal surgery is scheduled. In case of meningitis, the whole VPS is removed and replaced by ETV. In case of isolated infection of the abdominal cyst, the peritoneal catheter is exteriorized and a new shunt can be implemented after 2-3 weeks' antibiotic therapy, in case of communicating hydrocephalus. Onset of CSF pseudocyst is of poor prognosis for the shunting in the peritoneal cavity, and ventriculoatrial shunt (VAS)should be considered.

In case of obstructive hydrocephalus, an ETV can be considered. Finally, in the absence of signs of pseudocyst infection, the peritoneal catheter can be repositioned at the contralateral side in the same surgical step.

The initially suspected diagnostic was wrong, given the recurrence of the cyst a few months after VPS removal, and diagnosis was rectified as massive ovarian mucinous cystadenoma. The initial diagnosis could have been corrected if we had paid attention to the extra-cystic situation of the peritoneal catheter, which did not end inside the cyst, as would usually be the case in pseudocyst [6, 7].

Ovarian cysts represent a large proportion of intra-abdominal cysts in the female population. Most are benign, especially before the menopause [8]. They can be functional or organic. Organic cysts may develop from the epithelial or the stromal part of the ovary [8]. Mucinous cystadenomas are less frequent than serous cystadenomas, and have specific characteristics: most are voluminous, with a mean diameter of 18 cm [9]; they are often pluri-locular, with thin walls. Ultrasonography is the first-line radiological

exploration in abdominal cyst [10]. Pelvic MRI is indicated if cyst diameter is >7 cm or if there are signs of malignancy on ultrasonography. Surgery should be performed if the cyst is voluminous, symptomatic or presents radiological signs of malignancy [10].

In the present case, the symptoms of shunt dysfunction completely masked the chronic ovarian pathology and left the final diagnosis overlooked. Genital cysts in pre-menopausal women are far more frequent than pseudocysts in patients with VPS. Here, the priority was to manage the life-threatening acute hydrocephalus. Nevertheless, after resolving the shunt dysfunction, the abdominal effusion could have been analyzed, including all the clinical and radiological elements already present and suggested by the guidelines [10].

Otherwise when VPS is dysfunctional because of a pseudocyst, the way to treat the hydrocephalus has to be discussed. Intra-abdominal reimplantation of the shunt is possible, depending on pseudocyst infectious status and size and the degree of peritoneal adherence. Another solution can be VAS, avoiding the peritoneum. The etiology of hydrocephalus is also important to consider in case of shunt dysfunction. When hydrocephalus is caused by an obstruction in the CSF flow path, as in the present case, ETV can be a precious alternative, allowing VPS removal. This option should be considered even in longstanding shunt or risk of impaired CSF absorption. The rate of chronic complications associated with ETV is lower than with VPS or VAS, due to the absence of foreign body [11].

4. Conclusion

Implanting a ventriculoperitoneal shunt in young patients with hydrocephalus amounts to replacing one subacute or chronic pathology by another. The rate of complications is significant, and some can be very severe and life-threatening.

In case of suspected abdominal pseudocyst in shunted patients, especially in women and if the cyst is very large, differential diagnoses need to be considered. Abdominal effusion should be systematically explored, as therapeutic options may be radically different and inappropriate management can have important consequences such as tumoral spread due to a puncture instead of en-bloc removal.

Finally, ETV should be favored in case of obstructive hydrocephalus, due to its lower risk of chronic complications. The absence of foreign body avoids most mechanical complications, abdominal complications are non-existent, and the risk of infection is lower than with ventriculoperitoneal shunt.

Figure 1: A: Reference CT-scan, far from the previous revision surgery, in asymptomatic patient. B: Emergency CT-scan showing a clear increase in the size of the ventricles. C: Abdominal CT-scan showing a very large cyst. The peritoneal catheter is pushed back to the side (yellow arrows) and seems to remain extra-cystic. D: Chest X-ray showing the mass effect of the cyst on the diaphragmatic cupolas. E: normalization of chest X-ray after cyst evacuation. F: MRI, axial slice, T2 sequence, confirming stenosis of the aqueduct with a ventriculo-cisternal pressure gradient, as shown by the floor of the 3rd ventricle thinned and lowered 3rd ventricle (red arrow). G: CT-scan after ETV, with ventricular size normalization. The shunt will be removed secondarily. H: Postoperative MRI after ETV, with ventricular size reduction and 3rd ventricle floor in anatomical situation (green arrow). I: Abdominal CT-scan showing recurrence of the bulky cyst that finally proves to be a massive ovarian mucinous cystadenoma.

Statement regarding patient consent: The patient authorized the publication of her clinical case (written agreement).

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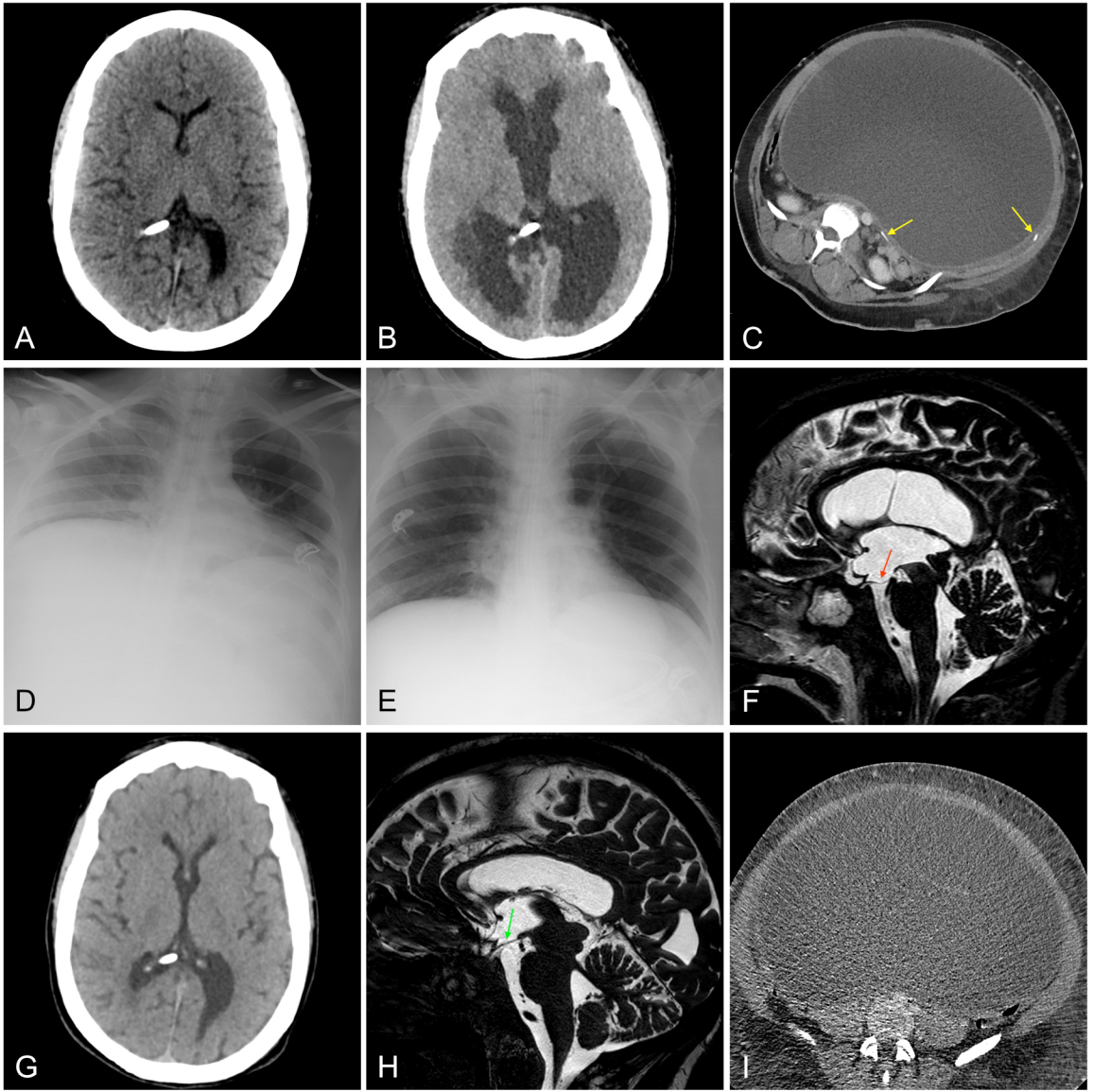
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