Case report

A left-sided cerebrospinal fluid hydrothorax and a right ventricular-peritoneal shunt: a unique clinical case study

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SUMMARY

We describe the case of a 84-year-old woman with a right ventricular-peritoneal shunt and a left-sided pleural effusion, the analysis of which was positive for cerebrospinal fluid. We consider the potential causative mechanisms. Our patient was managed conservatively due to her frailty, the effusion being asymptomatic and her preference not to pursue further invasive diagnostic testing. This case report is unique due to the contralateral nature of the effusion to the shunt, which has not been described before in the literature.

BACKGROUND

Cerebrospinal fluid (CSF) in the pleural space, known as CSF hydrothorax, has been described in numerous case reports and is more frequent in the paediatric population. This is probably because hydrocephalus is more common in children and treated with ventricular shunts. These shunts can be atrial, pleural or peritoneal.

The most common reason for the development of CSF hydrothoraces is shunt migration into the thoracic cavity. Very large pleural effusions can also be formed due to fistulas between the pleural cavity and the subarachnoid space. However, the aetiology of those is usually through trauma, cardiothoracic and spinal surgeries.

Identification of beta 2 transferrin in any fluid is a highly specific and sensitive test, consistent with the presence of CSF.

In the existing literature, CSF hydrothoraces have always been described on the same side as the ventricular-peritoneal (VP) shunts, usually due to ipsilateral shunt migration.

CASE PRESENTATION

An 84-year-old female patient was referred to the frailty assessment unit by her general practitioner.

In 2009, she had a right-sided acoustic neuroma resected and a right VP shunt inserted for secondary obstructive hydrocephalus. In March 2018, she tripped, fell and sustained a right subdural haematoma that required surgical evacuation with a burr hole. She had a prolonged hospital stay on the neurosurgical unit and on discharge was referred by her general practitioner to the frailty assessment service. Further systemic enquiry revealed no weight loss, chest pain or breathlessness.

Her medical history included treated hypertension and short-term memory impairment.

Her medication included indapamide 2.5mg once a day, amlodipine 10mg once a day, atorvastatin 80mg once a day, doxazosin 4mg once a day and paracetamol 1g four times a day, as required.

On assessment, she had a low body mass index (BMI 19.5) and normal observations, including normal oxygen saturations on air.

Cardiovascular examination was normal. There was dullness to percussion in the left hemi-thorax and reduced air entry. Neurological examination was normal. Generalised muscle wasting was noted with no fasciculation. Further examination revealed no lymphadenopathy, no breast masses or skin changes.

Her blood tests including haemoglobin levels and kidney function were normal.

INVESTIGATIONS

A chest X-ray was performed and showed a left pleural effusion. There were no previous X-rays available for comparison.

A CT scan of her head, neck, chest and abdomen was done. [Figure 1](#) shows a left-sided effusion with a smooth pleural margin with no thickening, nodularity or tumour. The VP shunt was sited appropriately from her cerebral space into her abdomen where there was no fluid collection. The shunt was not broken and had not migrated ([figures 2–5](#)).

The patient was thus referred to the specialist pleural clinic. A thoracic ultrasound showed a large left-sided hyperechoic effusion. Under asepsis and local anaesthetic, a pleural tap was done and 50 mL of straw-coloured fluid was removed. Analysis revealed a lymphocytic exudate with a protein count of 49 g/L and fluid lactate dehydrogenase (LDH) at 172 units/L. pH was normal at 7.4. Glucose levels were not sent. The plasma LDH levels were 250 (<270) units/L, her total serum protein level was 75 g/L (range 60–80 g/L) and albumin level 37 g/L (range 35–50 g/L). Cytological and microbiological analyses were negative. Acid fast bacilli were not seen or grown. A full autoimmune panel screen was negative. A biopsy of the pleural margin was considered. Our centre runs a well-established pleural service offering medical thoracoscopy and image-guided closed pleural biopsy.

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Figure 1 Chest X-ray showing right-sided ventricular-peritoneal shunt and left pleural effusion.

The patient refused further interventions citing progressive frailty and being asymptomatic from an incidental finding.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for an exudative lymphocyte-rich effusion is wide: occult malignancy needs to be considered as well as infections such as tuberculosis and autoimmune conditions such as rheumatoid arthritis. However, as discussed below, they were all felt to be unlikely.

CSF hydrothorax was also thought to be a remote possibility and the fluid was sent for analysis for beta 2 transferrin. This was positive.

Figure 2 CT scan showing well-sited right ventricular-peritoneal shunt with no migration and left-sided pleural effusion with no nodularity on the pleural surface.

Figure 3 Coronal image showing ventricular-peritoneal shunt in right side of neck.

OUTCOME AND FOLLOW-UP

The patient was asymptomatic from her effusion and hence no immediate intervention was required. She has now been under follow-up for over a year and there has been no increase in the size of her pleural effusion and there have not been any symptoms. After discussion with the neurosurgical team and more imaging showing no defects in the shunt (CT head, X-ray abdomen and cervical spine), no further action was planned.

She continues to be actively monitored.

DISCUSSION

Application of Light's criteria help in the differentiation between an exudate and transudate.¹ Our patient fits the criteria for an exudate with the protein ratio of 0.65 and an LDH ratio of 0.69.

The presence of lymphocytes in the fluid suggests a long-standing effusion and can be associated with infections such as tuberculosis and malignancy.¹

Consideration was given to whether this could be tuberculosis but the patient had no weight loss, cough or haemoptysis. She had also resided in a rural region of the North East of England with an extremely low incidence of tuberculosis. She had not travelled in the last 10 years and no tuberculosis contact. Hence, the possibility of this being a tuberculous effusion was dismissed. An assay for adenosine deaminase was not sent as it was not thought to be indicated given the low likelihood of tuberculosis. Acid fast bacilli were not seen or grown.

Further consideration was given to whether this represented a malignancy. A pleural effusion is a common presentation of metastatic malignancy.

CT scanning has a relatively low sensitivity for the detection of pleural tumours, but there was no thickening, nodularity or enhancement of the pleural margins which are strong predictors of a malignant effusion.¹ Pleural fluid cytology was negative. The sensitivity of that particular test is very variable (10%–80% sensitivity) and as such, the next preferred investigative step would be a pleural biopsy under direct vision at medical thoracoscopy. This was declined by the patient as explained in the foregoing. A

Figure 4 Further coronal image showing ventricular-peritoneal shunt going subcutaneously and into the abdominal cavity.

closed pleural biopsy under ultrasound guidance was considered. The previous thoracic ultrasound had shown a smooth pleural margin with no thickening or discrete masses to target and as such the potential sensitivity of such a biopsy was deemed low. Furthermore, the patient refused this. Repeat thoracentesis was also considered for repeat cytological analysis but again, further procedures were declined by the patient.

Other causes of pleural effusions were considered such as her medications but none of her prescribed medications are associated with pleural effusions.

Detection of beta 2 transferrin confirmed that the effusion contains CSF.

Figure 5 Further coronal image showing ventricular-peritoneal shunt going subcutaneously outside the thorax.

Transferrin is found in most body fluids. The CSF-specific variant migrates into the beta 2 fraction when separated by electrophoresis and is hence referred to as beta 2 transferrin. Beta 2 transferrin is only detectable in CSF, perilymph and aqueous humour. It is formed by the loss of sialic acid due to the presence of neuraminidase in the central nervous system. It is utilised extensively to diagnose CSF rhinorrhoeas and has a sensitivity of 94%–100%, and specificity of 98%–100% to detect CSF.^{2,3}

A transferrin band was clearly seen in the beta 2 region when electrophoresis was performed on the pleural fluid. False positive results are rare but are possible in patients with chronic liver disease or inborn errors of glycoprotein metabolism.^{2,3} In the absence of any of other cause for the effusion, although with limited diagnostics performed, we concluded that CSF migrated into the pleural space and caused a CSF hydrothorax.

A review of the sparse literature reveals that the pleural effusion due to a CSF hydrothorax tends to be transudative, clear and colourless with a paucity of cells on cytology.^{4–11} However, pleural fluid eosinophilia and lymphocytosis have been noted and remain unexplained. Lymphocytosis in a pleural effusion implies chronicity—our patient had not had a chest radiograph for over 10 years and there is no way of determining when her pleural effusion developed.

Over 60% of cases of CSF hydrothoraces are due to distal catheter tip migration into the thorax.⁴ Once the shunt has migrated, the resorptive capacity of the pleura is overwhelmed and effusions develop. The normal pleural space is capable of reabsorbing 3 mL/kg of fluid each day and normal choroid plexus produces approximately 600 mL/kg of CSF. Chronic damage to the pleural surface reduces the resorptive capacity.⁴

There are numerous case reports in the literature of patients with VP or atrial shunts developing CSF hydrothoraces.^{4–11} A selection of them are described. Sekiguchi *et al.*⁴ describe a 61-year-old woman with many revisions of a right VP shunt for previous aqueductal stenosis who presented with a right-sided pleural effusion and ascites. The effusion was a neutrophilic exudate and positive for CSF. Bacon *et al.*⁵ describe a 35-year-old woman who presented with a large left-sided pleural effusion but do not elaborate whether it was exudative or not. She had a left VP shunt inserted aged 8 for spina bifida and hydrocephalus and imaging revealed intrapleural tracking of the shunt. Shipley *et al.*⁶ describe a case of a large right-sided transudative pleural effusion due to displacement of a ventricular-atrial shunt into the mediastinum. All of these patients had various neurosurgical procedures such as shunt removal, shunt revisions, third ventriculostomies as well as installation of anti-siphon valves to prevent accumulation of the effusion.

There was no evidence of shunt migration or fistula formation in this case. A shuntogram¹² was not performed as the patient was asymptomatic, frail and did not want any further procedures. As such, we hypothesise on how the effusion could have developed: CSF from the ventricles is diverted into the peritoneal space and subsequently migrated into the pleural space through diaphragmatic defects.

The diaphragm has various foramina to allow passage of structures such as the right and left greater and lesser right splanchnic nerves, the hemiazygous vein, the sympathetic trunks, the subcostal nerve and vessels, the left phrenic nerve and the vena cava.

Migration of peritoneal fluid, even in the absence of detectable ascites, through those foramina into the pleural space, is well described in patients with decompensated liver disease, chylothorax, yellow nail syndrome, urinothorax and pancreaticopleural fistulas. Factors implicated are the generation of

Patient's perspective

Relative's perspective: Since the head injury requiring surgical intervention, my mother has been very frail and the initial diagnosis was a surprising one. However, my mother does not seem to be affected by the effusion and as such, she is quite happy for the situation just to be monitored. Both my parents are very relieved that she does not need to go into hospital for further investigations as she does not fare well as an inpatient, does not eat and becomes very anxious.

I am worried that if my mother has a chest infection or similar ailment, that the fluid would have potential to aggravate the situation.

Learning points

- Identification of beta 2 transferrin in a fluid is a highly specific test for the presence of cerebrospinal fluid (CSF).
- CSF hydrothoraces are commonly due to ventricular-peritoneal or ventricular-atrial shunt migration.
- Peritoneal fluid can migrate into the pleural spaces through diaphragmatic defects and in patient with decompensated liver disease, pleural effusions can develop in the absence of peritoneal fluid. CSF hydrothorax should be considered as an aetiology for an unexplained pleural effusion in a patient with a ventricular shunt.

negative intrathoracic pressures, increased abdominal pressures, a piston-like effect of the liver and in cirrhotic patients, thinning of the diaphragm. Herniation of the peritoneum into the pleural space can also develop due to an increase in the gaps between the diaphragmatic muscle fibres with increased intra-abdominal pressure. These herniations are known as pleura-peritoneal blebs and can rupture to increase passage of fluid into the thoracic cavity. These are usually visible at thoracoscopy and can be repaired. Such effusions are right-sided in 85% of cases, but left ones occur in approximately 12%; the rest are bilateral.^[13–16]

Hence, in this case, we hypothesise that any CSF deposited in the peritoneum is migrating directly up into the pleural space.

As such, this is the first description of a CSF hydrothorax on the contralateral side to a VP shunt.

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