Unexpected outcome (positive or negative) including adverse drug reactions

A child with suprasellar mass and ascites

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Summary

A 7-year-old child having short stature presented with headache and vomiting of 6 months' duration. MRI of the brain showed a sellar and suprasellar mass suggestive of craniopharyngioma with hydrocephalus. He underwent a right ventriculo-peritoneal (V-P) shunting followed by a subtotal resection of the tumour. A year later, he presented with progressive ascites and umbilical hernia. Systemic examination was unremarkable except for massive ascites. Ultrasound abdomen confirmed free-fluid in the peritoneal cavity and the ascitic fluid was transudative. Ventriculocysternography revealed a functional and patent V-P shunt. A diagnosis of cerebrospinal fluid ascites (shunt ascites) was made and he underwent ventriculo-atrial shunting. After treatment the patient improved with the gradual disappearance of ascites.

BACKGROUND

Ventriculo-peritoneal (V-P) shunting is a relatively common procedure in neurosurgical practice. Though shunt occlusion and infection are common complications, development of cerebrospinal fluid (CSF) ascites (shunt ascites) is very rare and may develop at variable time intervals following the procedure. It needs strong index of suspicion and careful evaluation since the removal of the V-P shunt and the placement of a ventriculo-atrial (V-A) shunt is curative.

CASE PRESENTATION

A 7-year-old child presented to us due to failure to gain height for the past 3 years and progressive headache associated with vomiting of 6 months' duration. Imaging (CT scan and subsequently MRI brain) revealed large sellar and suprasellar mass with cystic component and calcification suggestive of craniopharyngioma with hydrocephalus (figure 1). The patient underwent immediate right V-P shunting. Six months later, right pterional craniotomy and subtotal excision of the tumour was done. Postoperative dynamic testing found him to be hypothyroid, hypocortisolic and growth hormone deficient. The child received replacement doses of levothyroxine and hydrocortisone. However, he could not afford to receive growth hormone.

One year later, the patient presented with progressive distension of the abdomen and respiratory difficulty. On examination, the child was afebrile and vital parameters were stable. He was short (height=96 cm, <3rd centile) and emaciated. Abdominal examination revealed a distended abdomen with demonstrable fluid thrill, umbilical hernia and bilateral hydrocele suggestive of massive ascites (figure 2). Cardiovascular, respiratory and neurological examination revealed no abnormality.

INVESTIGATIONS

His renal function and liver function revealed no abnormality. Viral markers for hepatitis B and C were negative. Ultrasound of the abdomen and pelvis revealed normal hepatic size and echotexture, normal portal vein diameter and significant free-fluid in the abdominal cavity without septa-

tion or any mass lesion. Echocardiography of the heart was normal. Ascitic fluid was transudative (protein content of 0.9 g/dl and high serum-ascitic fluid albumin gradient) with no signs of infection (total leucocyte count was 18 with 60% lymphocytes, negative culture and low adenosine deaminase level) and no malignant cells. Ventriculocysternography revealed functional and patent V-P shunt.

DIFFERENTIAL DIAGNOSIS

A diagnosis of shunt ascites following V-P shunt was made after excluding other aetiologies like chronic liver disease, constrictive pericarditis and nephrotic syndrome.

TREATMENT

With a diagnosis of CSF ascites, a V-A shunt was placed and the V-P shunt was removed.

OUTCOME AND FOLLOW-UP

Following the procedure, the ascites resolved gradually after 1 month.

DISCUSSION

The treatment for hydrocephalus is either by a V-P or a V-A shunt procedure. V-A shunting is used infrequently because of life-threatening complications like venous thrombosis, bacteraemia and sepsis; hence, the V-P shunt is preferred. Occlusion of the shunt tube and infection are frequently observed complications of V-P shunt.¹ Other rare complications include intracranial haematoma, intestinal perforation by the tube,² and tube migration to the thorax and scrotum.³

CSF ascites is a very rare complication of V-P shunt. To the best of our knowledge, only 28 cases of shunt ascites have been reported in the world literature⁴ and, out of these, 4 cases had craniopharyngioma. Several aetiological factors have been discussed in literature, but all are elusive. An imbalance between peritoneal absorptive capacity and amount of CSF production is implicated as a major cause of shunt ascites. Therefore, patients with excessive amounts

Figure 1 Non-contrast CT head showing suprasellar mass with calcification.

Figure 2 Child with massive ascites, umbilical hernia and hydrocele after ventriculo-peritoneal shunting. Note shunt tube is visible along the chest wall.

of CSF production like choroid plexus papilloma are at the risk of developing CSF ascites following V-P shunt.⁵ Similarly, patients with high CSF protein due to chronic infection (tuberculous meningitis)⁶ or brain tumours, especially optic glioma,⁷ may have difficulties in CSF absorption through the peritoneum due to high viscosity. Peritoneal inflammation due to repeated shunt revisions,⁸ or non-specific inflammatory response to shunt material,⁹ may also play a role in decreasing the absorptive ability of the peritoneum. Recently, vascular permeability factor secreted by both low and high grade gliomas are implicated in the formation of shunt ascites.¹⁰

However, in most reported cases, the aetiology was unknown and V-A shunt was effective. Out of the four cases of shunt ascites reported in patients with craniopharyngioma, only two had resolution of ascites following V-A shunting.⁴ Our patient had shunt ascites 1 year following V-P shunt. Non-specific peritoneal inflammation to shunt material or some putative inflammatory factor released from craniopharyngioma might be the cause for his ascites, which successfully resolved after V-A shunt.

Learning points

- ▶ Shunt ascites is a rare complication following V-P shunt procedure.
- ▶ The diagnosis requires a high index of suspicion.
- ▶ Removal of the V-P shunt and placement of a V-A shunt is curative.

Competing interests None.

Patient consent Obtained.

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