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Case Report

Diffuse villous hyperplasia of choroid plexus

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Summary

Diffuse villous hyperplasia of choroid plexus (DVHCP) is a rare condition which is characterized by the presence of diffuse enlargement of the entire choroid plexus throughout the length of the choroidal fissure and overproduction of CSF. The diagnosis of diffuse villous hyperplasia of choroid plexus can be established by the MR demonstration of diffusely large, contrast enhanced choroid plexus in the cases of overproduction hydrocephalus. Although some authors recommend choroid plexus excision or coagulation, ventriculo-atrial shunt insertion is a simple and effective treatment modality in cases of diffuse villous hyperplasia of the choroid plexus.

In this report we present a case of diffuse villous hyperplasia of the choroid plexus and a short review of the literature . To our knowledge, in the CT and MRI era only 5 cases of DVHCP cases have been reported.

Keywords: Choroid plexus; hydrocephalus; ventriculo-atrial shunt; villous hyperplasia.

Introduction

Diffuse villous hyperplasia of choroid plexus (DVHCP) is a rare condition which is characterized by the presence of diffuse enlargement of the entire choroid plexus throughout the length of the choroidal fissure and overproduction of CSF [1, 4, 5, 10, 12].

This clinical entity is distinct from bilateral choroid plexus papillomas and the patients usually present with clinical findings of hydrocephalus. Classic V-P shunt treatment is insufficient because the CSF production is higher than the resorption capacity of the peritoneum.

In this report we present a case of diffuse villous hyperplasia of the choroid plexus diagnosed by MRI and treated with V-A shunt insertion. To our knowledge, in the CT and MRI era only 5 cases of DVHCP cases have been reported.

Literature review

DVHCP was first described by Demselben as early as 1884, since then, limited cases have been reported [1, 4, 5, 10, 12]. In 1924 Davis described a case of diffuse enlargement of the choroid plexus in both lateral ventricles with normal histopathological examination.

It is well known that choroid plexus tumors can cause hydrocephalus related to the overproduction of CSF by the tumor itself [6, 9]. This is the same mechanism responsible for the hydrocephalus in the cases of DVHCP. The excessive CSF production documented in some cases was found to be 2–5 times of normal daily CSF production and the term villous hypertrophy of the choroid plexus was used. Some authors described similar cases. In the CT and MR era only 5 well documented cases of DVHCP have been reported (Table 1).

Analysis

The rarity of DVHCP is due to two factors; First, diagnosis of DVHCP could be established only at postmortem or by pathological examination before the CT and MR era. Second, some of the cases were probably reported as bilateral choroid plexus papillomas [2, 6, 9]. However, on pathological examination choroid plexus structure must be found normal, although they are diffusely enlarged macroscopically. The term diffuse hypertrophy of the choroid plexus described by Davis [4], is not appropriate because the size of the cells are not increased. Therefore, the term hyperplasia is more appropriate to describe the situation. Also, some cases of

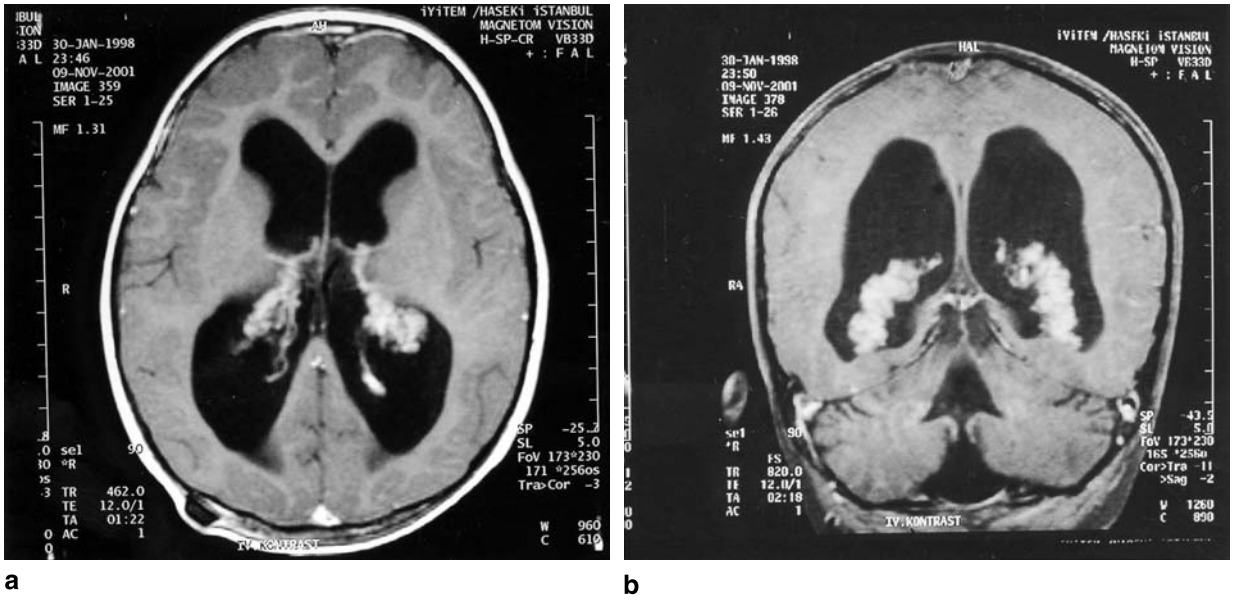
Table 1. *Previously reported cases of diffuse villous hyperplasia of choroid plexus (DVHCP)*

Author	Age/sex	Initial treatment	Treatment modality
Welch, 1983 [19]	10 d F	V-P Shunt	choroid plexus resection
Hirano, 1994 [12]	7 y F	V-P Shunt	choroid plexus resection
Britz, 1996 [4]	3 m M	V-P Shunt	V-A shunt monopolar coagulation
Philips, 1998 [11]	14 m F	V-P Shunt	endoscopic coagulation
Fujimoto, 2004 [8]	16 m M	V-P Shunt	left choroid plexus excision
Iplikcioglu, 2005 (present case)	5 y F	V-P Shunt	V-A shunt

overproduction hydrocephaly were thought to be associated with DVHCP [9, 10, 12].

Illustrative case

A five year-old girl was admitted to the hospital with complaints of headache, nausea and vomiting. Her medical history was unremarkable, although she was noted to have an enlarged head since birth. Her mental and physical development was described as normal. Physical examination was normal but the head circumference was 54 cm. On neurological examination, she had an ataxic gait and bilateral papilledema. An urgent CT scan showed communicating hydrocephalus.



a b

c

Fig. 1. (a) Axial contrast enhanced MRI shows hydrocephalus and diffuse enlargement of entire choroid plexus throughout the length of choroidal fissure. (b) Coronal contrast enhanced MRI shows diffusely enlarged choroid plexus with ventricular enlargement. (c) MR angiography shows dilated ant. and post. Choroidal arteries (arrows)

A ventriculoperitoneal shunt was inserted. Examination of CSF obtained at the operation revealed a protein level of 32 mgr/DL and a glucose level of 54 mgr/DL. Postoperative course was uneventful. A contrast MR examination revealed; enlarged, highly contrast enhanced choroid plexus (Fig. 1a, b). For the differential diagnosis of choroid plexus arteriovenous malformation MR angiography was performed where dilated anterior and posterior choroidal arteries were detected with no appearance of AVM (Fig. 1c). On the 7th postoperative day the patient was discharged.

Two weeks after the discharge, the patient was readmitted to the hospital because of abdominal swelling. On physical examination ascites was detected, although laboratory examinations, including blood counts, electrolytes and liver function tests were normal. Abdominal MR examination confirmed free fluid in the peritoneum (Fig. 2). Abdominal paracentesis was performed. Protein levels in the abdominal fluid and in the CSF sample obtained from shunt reservoir were 96 mgr/DL and 44 mgr/DL respectively. Gram stain and growth of culture from the abdominal fluid and shunt reservoir were negative.

The patient was reoperated and the abdominal catheter was withdrawn and ventriculoatrial (V-A) shunt was inserted. Postoperative course was uneventful and at the last follow up examination 3 years after the second operation, the patient was normal neurologically.

Fig. 2. Abdominal MRI displays massive free fluid in the peritoneum

Discussion

Today, MR demonstration of diffusely large, contrast enhanced choroid plexus without discrete mass, suggesting papilloma in the presence of overproduction hydrocephaly is sufficient for the diagnosis of DVHCP [1].

Bilateral choroid plexus papillomas are usually asymmetrical and appear as discrete masses [7]. Recently D'Ambrosio AL used proliferative index MIB to distinguish the bilateral choroid plexus papillomas from DVHCP [3].

DVHCP usually occurs in neonates or infants and is thought to be congenital. In the case of Welch *et al.*; hydrocephalus and large choroid plexuses were diagnosed in utero [12].

Patients with DVHCP, usually present with symptoms of hydrocephalus and ventriculoperitoneal (V-P) shunt treatment is performed as an initial surgical procedure. These patients usually develop ascites due to overproduction of CSF. In some of these cases daily CSF production was measured and found to be 4 or 5 times that of the normal amount of daily production. Welch measured the CSF production rate by a radionuclide assessment method. Hirano and Britz also measured the daily rate of CSF production by way of external drainage. This procedure can cause fluid and electrolyte imbalance due to large amount of fluid loss and carries a risk of infection. Therefore we performed a V-Atrial shunting procedure without an external drainage period. Today (Then) detailed neuroradiological examination including contrast enhanced MRI usually clinches the diagnosis of DVHCP.

Some authors prefer coagulation or surgical excision of the choroid plexus for the treatment of diffuse villous hyperplasia of the choroid plexus [8, 10–12]. This treatment has a surgical morbidity including hemianopsia, nystagmus and mortality [12]. Hirano *et al.* used a contact yttrium- aluminum garnet laser system before resection of the choroid plexus to decrease the bulk and fascilitate the operation [10]. In Fujimotos case after the resection of the left lateral ventricle choroid plexus, the patient's ascites resolved [8]. However V-A shunt is a well tolerated and simple treatment modality. In our case and in the case reported by Britz V-A shunt was used [1]. Surgical excision of choroid plexus should be performed in the cases of V-A shunt failure.

Conclusion

In our case MR angiography showed well developed choroidal arteries. Although the patients are usually

neonates or infants, embolization of choroid plexus via well developed choroidal arteries could be a treatment option in the future.

References

1. Britz GW, Kim DK, Loeser JD (1996) Hydrocephalus secondary to diffuse villous hyperplasia of the choroid plexus. Case report. J Neurosurg 85: 689–691
2. Coates TL, Hinshaw DB, Peckman N, *et al* (1989) Pediatric choroid plexus neoplasms: MR, CT and pathologic correlation. Radiology 173: 81–88
3. D'Ambrosio AL, O'Toole JE, Connolly ES Jr, Feldstein NA (2003) Villous hypertrophy versus choroid plexus papilloma: a case report demonstrating a diagnostic role for the proliferation index. Pediatr Neurosurg 39(2): 91–96
4. Davis LE (1924) A physio-pathological study of the choroid plexus with the report of a case of villous hypertrophy. J Med Res 44: 521–534
5. Demselben (1884) Hyperplasie der plexus choroider laterales bei hydrocephalus internus congenitus. Jahrb Kinderh 21: 419–420
6. Ellenbogen RG, Winston KR, Kupsky WJ (1989) Tumors of the choroid plexus in children. Neurosurgery 25: 327–335
7. Erman T, Göcer AI, Erdogan S, Tuna M, Ildan F, Zorludemir S (2003) Choroid plexus papilloma of bilateral lateral ventricle. Acta Neurochir (Wien) 145: 139–143
8. Fujimoto Y, Matsushita H, Plese JP, Marino R Jr (2004) Hydrocephalus due to villous hyperplasia of the choroid plexus. Case report and review of the literature. Pediatr Neurosurg 40(1): 32–36
9. Gudeman SK, Sullivan HG, Rosner MJ (1979) Surgical removal of bilateral ventricles with resolution of hydrocephalus. J Neurosurg 50: 677–681
10. Hirano H, Hirahara K, Asakura T (1994) Hydrocephalus due to villous hypertrophy of the choroid plexus in the lateral ventricles. Case report. J Neurosurg 80: 321–323
11. Philips MF, Shanno G, Duhaime AC (1998) Treatment of villous hypertrophy of the choroid plexus by endoscopic contact coagulation. Pediatr Neurosurg 28(5): 252–256
12. Welch K, Strand R, Bresnan M (1983) Congenital hydrocephalus due to villous hypertrophy of the telencephalic choroid plexuses. Case report. J Neurosurg 59: 172–175

Comment

The authors present the case history of a rare neuropediatric situation with hydrocephalus caused by diffuse villous hypertrophy of the choroids plexus without evidence of tumour in a 5 year-old child. Interestingly and, apparently, typical for this pathology, the CSF hyperproduction was not sufficiently treated by ventriculo-peritoneal shunting so that the patient had to be reoperated for replacement by a ventriculo-atrial shunt. In addition to the case report, the authors present a good review of the literature on this CSF pathology.

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