Clinical Images and Case Reports

Received 25 Dec 2023 | Revised 26 Dec 2023 | Accepted 28 Mar 2024 | Published Online 29 Mar 2024

Published By: Vision Publisher

ISSN 2942-0776

CICR 02 (3), 15-19

Choroid Plexus Papilloma

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Abstract

Choroid plexus papillomas (CPP) are benign tumor originating from the epithelial cells of the choroid plexuses. Choroid plexus tumors are categorized into three groups based on some histological findings. We present a case of prenatally diagnosed at 34 weeks of gestation hyperechoic formation in the area of the choroid plexus in a girl born by Caesarean section with an Apgar score of 6/7 at term. After birth, she has delayed cardio-pulmonary adaptation. No abnormalities were observed on neurological examination. Ultrasound examination showed a hyperechoic formation with an inhomogeneous structure occupying 3/4 of the right lateral ventricle and notmal cerebrospinal fluid drainage. During the neonatal period, an increase in the size of the formation was registered. Operative treatment was performed. Postoperatively, right-sided ventricular dilatation with a formed porencephalic cyst was observed at the operative access site. Subsequently, a decrease in the size of the ventricle and growth of the choroid plexus was recorded in the sixth month. By the age of two, the child showed normal neuropsychological development

Keywords: choroid plexus papilloma, newborn, ultrasoun

Supplementary information The online version of this article (https://doi.org/xx.xxx/xxx.xx) contains supplementary material,which is available to authorized users.

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Background

Choroid plexus papilloma (CPP) are benign tumor originating from the epithelial cells of the choroid plexuses, most common in children, often accompanied by hydrocephalus[1]. Choroid plexus tumors are categorized into three groups based on some histological findings: Grade I or CPP, which is a slow-growing and benign tumor; Grade II or atypical CPP (aCPP), which is a mid-grade tumor with a higher probability of recurrence; and Grade III tumors of choroid plexus carcinoma (CPC) as the malignant neoplasm of choroid plexus [2]. The prognosis after CPP is widely variable and depends on the histological findings. Approximately 10% of all brain tumors in infants and 5% of perinatal brain tumors are of choroid plexus etiology[3]. Tumor of the choroid plexus is a rare type of intracranial tumor in children, but accounts for 42% of cerebral tumors in neonates [4]. The lateral ventricles are the main site of tumor origin followed by the third and fourth ventricles, respectively [5].

Case report

We present a case of prenatally diagnosed at 34 weeks of gestation hyperechoic formation in the area of the choroid plexus (Figure 1).

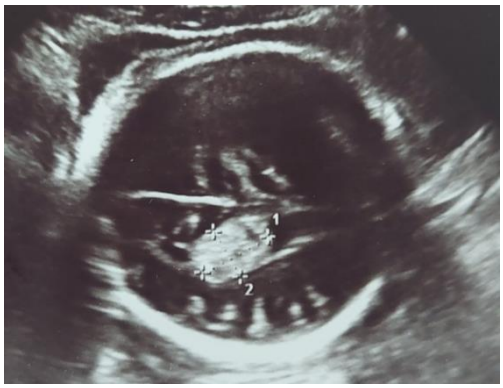

Figure 1. Ultrasound at 34 weeks

The mother was a 23-year-old primigravida. At the age of 15, she underwent treatment for acute leukemia, after which she was in remission. In the first trimester, the mother had Covid-19. The birth was by operation, due to the prenatally established formation and initial fetal distress of the fetus.

The newborn was a girl with a gestational age of 38 weeks, weight - 3410 g and height - 50 cm, Apgar score 6/7. After birth, she has delayed cardio-pulmonary adaptation and hypoglycemia. No abnormalities were observed on neurological examination. Moderate neonatal jaundice was reported. Ultrasound examination showed a hyperechoic formation with an inhomogeneous structure occupying 3/4 of the right lateral ventricle with dimensions of 34mm/24mm, does not compress adjacent structures. Normal CSF drainage. External CSF spaces not dilated. Non-dilated third and fourth ventricles and left lateral ventricular (Figure 2).

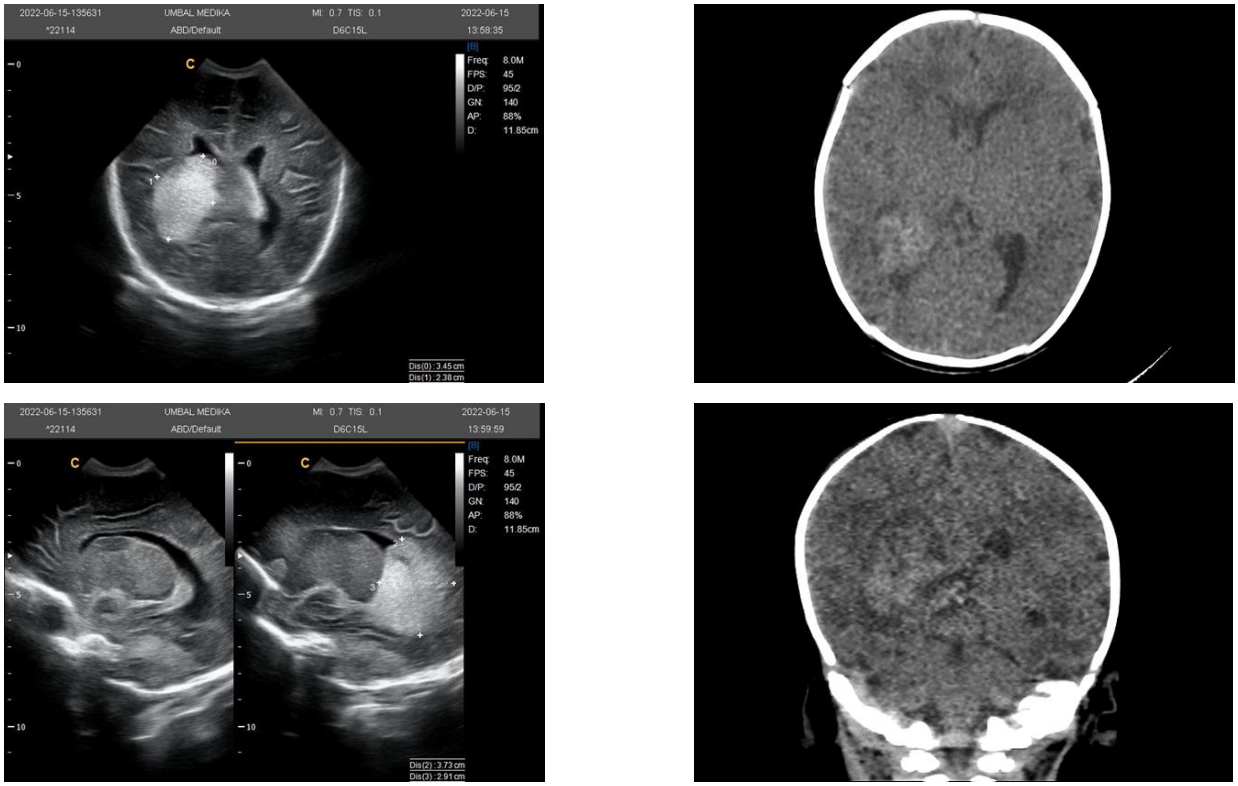

Figure 2. Ultrasound at Day 1 and CAT at Day2

During the neonatal period, an increase in the size of the formation was registered (Figure 3). This necessitated surgical treatment with removal of the structure. The histological variant was an aCPP.

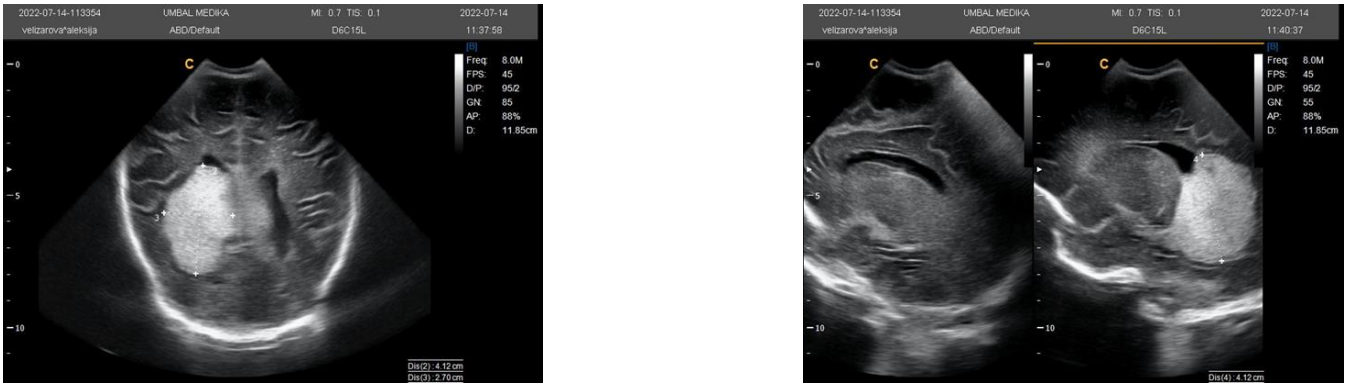

Figure 3. Ultrasound on 28th day

Postoperatively, right-sided ventricular dilatation with a formed porencephalic cyst was observed at the operative access site (Figure 4).

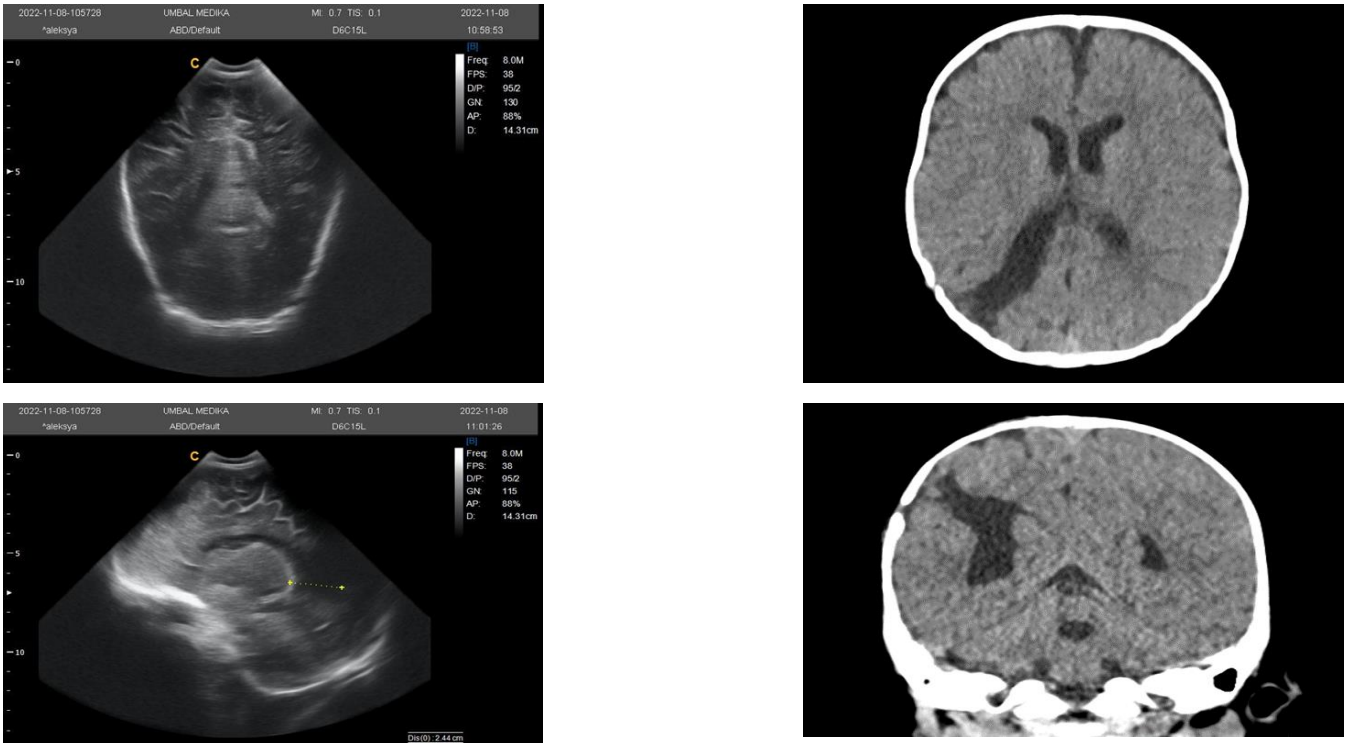

Figure 4. Postoperative ultrasound end CAT

Subsequently, a decrease in the size of the ventricle and growth of the choroid plexus was recorded (Figure 5). The child developed symptomatic epilepsy, which was controlled with monotherapy for the first year. Neurological development followed after discontinuation of anticonvulsant therapy during the second year showed no abnormalities. No relapse was recorded during this period.

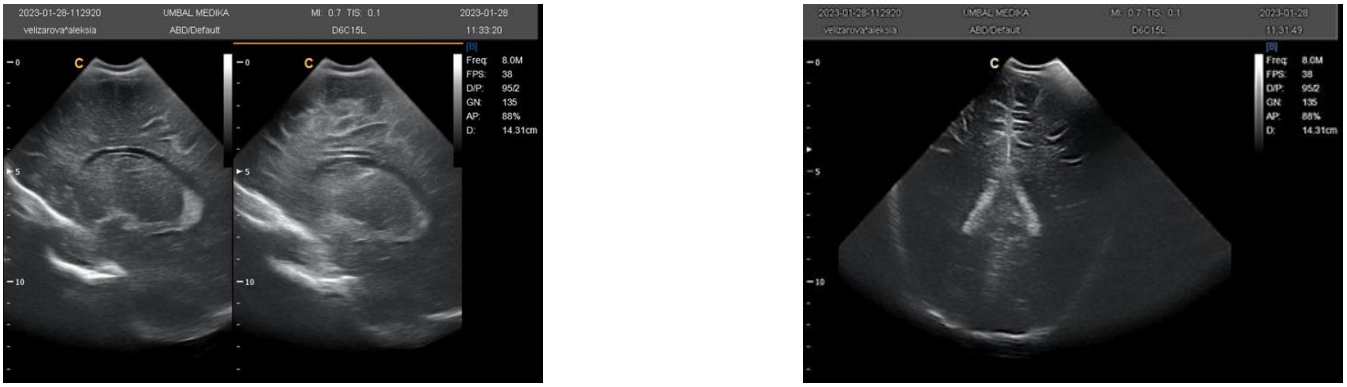

Figure 5. Ultrasound at 6th months

Discussion

A few case reports of congenital choroid plexus tumors diagnosed in utero exist in the literature [3]. Antenatal choroid plexus tumors are described most often during the third trimester, presenting as a large homogeneous mass, as in our case. They can also be revealed by the presence of hydrocephalus or ventriculomegaly because of overproduction of cerebrospinal fluid or obstruction of the foramen of Monro by the mass [4]. We found no difficulty in CSF drainage, probably due to the earlier surgical intervention in our patient. In pediatric patients, CPPs are found in the lateral ventricle in 71% of all cases, rarely are found in the third and fourth ventricles [6]. About half of them are located in the atrium of lateral ventricle [7]. The localization of the tumor in our patient was similar. In the pediatric population, there is significant (12%) perioperative mortality, mainly from catastrophic blood loss. Total removal is possible for 96% of papillomas and for 61% of carcinomas [4]. Hydrocephalus in patients with CPP located in the lateral or third ventricle was significantly higher than in those with tumors located in the fourth ventricle, while the incidence of acute hydrocephalus was much higher in the fourth ventriclehe[8]. Prognosis is widely variable between CPP and CPC, with a survival rate at 5 years of approximately 65-100% for papillomas and 40% for carcinomas [9, 11]. Continued follow-up of our patient is needed to account for his development at 5 years of age with regard to histological outcome (aCPP). Improvements in surgical and intensive care techniques have vastly improved the prognosis of patients. At presentation, children may have features of prolonged raised intracranial pressure such as papilledema, optic atrophy, and visual loss, which may not recover after surgery. Some may develop cognitive defects, bleeding, and seizures that may persist postoperatively. We also observed several epileptic seizures postoperatively, for which the child was on therapy during the first year. Management of CPT requires an interprofessional team that includes the neurosurgeon, various specialists (anesthetist, pediatrician, ophthalmologist, neurologist), and neuroscience specialty-trained nurses. Low-grade choroid plexus tumors treated with total resection have excellent long-term survival and function. Recurrences are uncommon (8.7%), appear within the first few years after primary surgery [10].

Conclusion

We present a case of prenatally diagnosed choroid plexus papilloma in a girl with early surgical treatment and a favorable evolution during the follow-up period without late neurological abnormalities.

Acknowledgments:

I thank G.Arasheva, M.Radulova for participation in prenatal and postnatal imaging.

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