

Histologic evaluation revealed polytypic lymphoplasmacytic infiltration with focal spindle-shaped cells which were found to be EBER positive. EBV-associated IPT was diagnosed. The patient had no post-operative complaints, and one month after surgery, the platelet count was $386,000 \times 10^9/\text{ml}$ with no recurrence of thrombocytopenia. Serum EBV-DNA results remained negative before and after diagnosis. **Discussion:** The IPTs of the spleen can develop either via proliferation of myofibroblasts or FDC that may be infected by EBV. They may be discovered by investigation of another disorder similar to our case as ITP, leukemoid reaction or hypercalcemia. Total resection of the tumor results in general improvement.

<https://doi.org/10.1016/j.htct.2024.11.054>

PP 27

CHOROID PLEXUS CARCINOMA AND CHOROID PLEXUS PAPILLOMA; RARE CASES

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Case Report: Choroid plexus carcinoma (CPC) is a rare and aggressive intracranial neoplasm, constituting 1–4% of all brain tumors and approximately 40% of choroid plexus tumors. Classified as a WHO Grade III malignancy, CPC is characterized by a poor prognosis, with reported 5-year survival rates around 40%. In contrast, choroid plexus papilloma (CPP), classified as a WHO Grade I tumor, is a benign and slow-growing lesion originating from the epithelial cells of the choroid plexus. This report presents four cases of choroid plexus tumors: two diagnosed as choroid plexus carcinoma (WHO Grade III) and two as choroid plexus papillomas (WHO Grade I). The CPP cases were managed with observation and followed up without active treatment. Among the CPC cases, a 3-year-old patient received initial radiotherapy followed by chemotherapy based on the CPT-SIOP-2000 protocol. A 7-month-old patient with CPC was treated with chemotherapy (CPT-SIOP-2000 protocol), while radiotherapy was deferred due to her age of less than 3 years. Multidisciplinary treatment strategies for CPC include maximal surgical resection followed by chemotherapy and radiotherapy. The CPT-SIOP-2000 study has demonstrated that the Carboplatin/Etoposide/Vincristine (CarbEV) chemotherapy protocol is effective in treating CPC.

<https://doi.org/10.1016/j.htct.2024.11.055>

PP 28

CLINICAL AND GENETIC FEATURES IN CONGENITAL GLYCOSATION DEFECTS PRESENTING WITH HEREDITARY HEMOLYTIC ANEMIA AND PROLONGED JAUNDICE

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Objective: Congenital glycosylation disorders (CGD) are a large group of genetic diseases that occur due to a decrease or increase in the glycosylation of glycoconjugates. Congenital glycosylation disorders; They can be grouped under 4 groups: protein N-glycosylation, protein O-glycosylation, combined N- and O-glycosylation and lipid glycosylation disorders. Congenital glycosylation disorders are divided into 2 main groups: Type I and II (CGB-1 and GB-2). In this article, we would like to present a cases of CGB with an atypical presentation, presenting clinical findings with hemolytic anemia and prolonged jaundice, and diagnosed by clinical exon panel genetic study, since it is very rare in the literature. **Case Report:** Our first patient, H1, was a 6-month-old male infant who received erythrocyte transfusion at an external center at the age of 14 days due to jaundice and anemia during the neonatal period (when HB: 5 g/dl), and then applied to the pediatric hematology clinic of our hospital with the same complaints at the age of 43 days. As a result of molecular tests, he was diagnosed with CGD type 2. Our other patient, H2, is a 10-year-old male, our third patient, H3, is a 13-year-old male, and our last patient, H4, is a 17-year-old male; These 3 patients were siblings. All three of them were hospitalized at an external center with jaundice and anemia during the neonatal period, but after diagnostic genetic tests, H4 was diagnosed after 3 years of age, but the other siblings were diagnosed after 6 months of age due to the oldest sibling's history. C.657c>A homozygous mutation was detected in the GSS gene in these siblings. **Methodology:** The diagnostic difficulties and treatment options of 4 patients (H1, H2, H3, H4), who received inpatient treatment with anemia and jaundice in the pediatric hematology clinic between 2022 and 2024 and were ultimately diagnosed with CGD, were obtained from the hospital information processing system and presented because they are very rare in the literature. **Results:** Our first patient, H1, was a 6-month-old male infant who received erythrocyte transfusion at an external center at the age of 14 days due to jaundice and anemia during the neonatal period (when HB: 5 g/dl), and then applied to the pediatric hematology clinic of our hospital with the same complaints at the age of 43 days. As a result of molecular tests, he was diagnosed with CGD type 2. Our other patient, H2, is a 10-year-old male, our third patient, H3, is a 13-year-old male, and our last patient, H4, is a 17-year-old male; These 3 patients were siblings. All three of them were hospitalized at an external center with jaundice and anemia during the neonatal period, but after diagnostic genetic tests, H4 was diagnosed after 3 years of age, but the other siblings were diagnosed after 6 months of age due to the oldest sibling's history. C.657c>A homozygous mutation was detected in the GSS gene in these siblings. **Conclusion:** Although prolonged jaundice and anemia are quite common, we wanted to emphasize with this very unique study that metabolic diseases may be among the differential diagnoses that are very rare in the literature. CGD has been diagnosed in only 40 cases in the last 30 years; Diagnostic evaluation with genetic consultation is very important for diagnosis. Literature data on rare diseases will be strengthened with new studies.

<https://doi.org/10.1016/j.htct.2024.11.056>