

## HEPATOBLASTOMA, A WAR WON OR...CASE PRESENTATION

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## ABSTRACT

### INTRODUCTION

Hepatoblastoma is one of the ten most frequent tumors in children, who constitute 0.5-2% of all pediatric tumors. It occurs mostly in the first two years of life with an incidence 0.9 / 1x10<sup>6</sup> children. The average age at diagnosis is 1 year old, most are diagnosed under the age of 2 years old. It's often associated with genetic Syndromes: as Familial adenomatous Polyposis (FAP), Beckwith-Wiedemann Syndrom or hemi-hypertrophy or very low birth weight/VLBW.

### CASE PRESENTATION

A 13 month's child (F.F), couple first child, preterm delivery 31-32 gestational weeks (his mother had bicorn uterus), Bw= 1500 gr, was presented at the Paediatric Emergency Department with vomiting, loss of appetite, abdominal distention. After performing an abdominal Ultrasound, which result with hyperechogen formation 5x7.2 cm size, localized in the left hepatic lobe. We asked a CT (with contrast), which confirm Ultrasound's data, and a height AFP (103.887 ng/dl). Biochemical examination liver function tests was normal, ALT/AST, bilirubine, while LDH=442 U/L. Complete Blood count: microcytic anemia. Liver biopsy found " Fetal type Hepatoblastoma". Were pre-treated with chemotherapy (CISP+ Doxo ), according to the classification SIOPEL PRETEX, and after three cycles performed for reduction of the amount, surgery attended a left lobectomy. Post-operative complicated with an abdominal fistula and after an inefficient drainage the child undergoes another laparotomy for a hepato-duodenal by-pass, according Kasay he is an healthy child.

### CONCLUSIONS

Increasing the attention and care of paediatrician of infants during periodic controls. Hepatoblastoma, although is a rare type of liver tumors, not always presented in its clinical signs and not always associated with a genetic or metabolic syndrome. If is evaluated in time, the quality of life and chances of survival increase substantially.

**Keywords:** Hepatoblastoma, non-specific symptoms, survivance.

## CASE RELEVANCE

- Very rare occurrence in medical practice, increasing attention of medical staff in recognizing and diagnosing liver tumor, which brings us more good results in further treatment, in particular for the Fetal type of Hepatoblastoma. Extensive involvement of multidisciplinary
  - *Pediatrician.*
  - *Radiation oncologist.*
  - *Pediatric nurse specialist.*
  - *Rehabilitation specialist.*
  - *Psychologist.*
  - *Social worker*
  - *Trasplant centre*

## PRESENTING HISTORY

**Present history:** The child was presented to the emergency room at the Nene Tereza Hospital in Tirana with vomiting and abdominal distention, loss of appetite and high fever. The child has these signs of 2-3 days. It also came the other night and took parenteral infusion.

**Past history:** The child was couple first child of preterm delivery 31-32 gestacional weeks ( his mother had bicorn uterus) due to natural birth with weight 1500 g. His Apgar score was 6- 8. He cry at birth and was intubated nasally and transfer at NICU. As a premature birth he had Respiratory Distress, he had about 3 weeks hospital recovery. He was breastfed until 5 months later with the formula. In the three months had a viral infection and was in hospital for 2 days. Complete blood count result limfocytosis, then the abdominal ultrasound was normal, transfontanelar ultrasound also was normal.

## PATIENT EXAMINATION

### Physical Examination

#### Vitals:

Temp: 37.8°C, Pulse: 120, Resp. Rate: 24 BP: 85/48 mmHg O2 sats: 94% on room air.

#### General:

Alert, fretful, Height/weight proportionate for a premature birth child. No acute distress.

#### HEENT:

Pupils equal, round, reactive to light and accommodation. Extra-ocular movements intact. Moist mucous membranes in oropharynx.

#### Neck:

Supple, without lymphadenopathy or thyromegaly. No carotid bruits.

#### Lymph:

No axillary, cervical, supraclavicular, pre-auricular, submental, or occipital lymphadenopathy,

#### Cardiovascular:

Regular rate and rhythm, with normal S1 and S2. No murmurs, rubs, or gallops. No JVD. 2+ pulses bilaterally – dorsalis pedis and radial.

#### Lungs:

Vesicular breathing throw the two lung fields . No wheezes. No accessory muscle use or cyanosis. No tenderness to palpation.

**Abdomen:**

Normoactive bowel sounds. Distended abdomen , a palpable mass with right sided extended to the left side and midline until the umbilicus .

**Skin:**

Warm, dry, well-perfused. No rashes or other lesions.

**Extremities:**

2+ pulses in upper and lower extremities. No lower extremity pain or edema; legs are symmetric in appearance.

**Rectal:**

Deferred.

**Neuro:**

Alert and oriented to person, place, and time. Able to communicate well.

Cranial nerves 2-12 grossly intact. 5/5 strength in all extremities bilaterally. Sensation intact in all extremities. Normal gait. 1+ DTR's in biceps, triceps, supinator, knee, ankle. No clonus.

**Admission labs:**

WBC: 14.000, Hgb: 7.8, Hct: 34.0,

Iron=24. AFP( alfa phetoprotein) =103.887 ng/dl. LDH =442 UI/L.

**Imaging & other studies:**

Abdominal Ultrasound

CT scanning with contrast

Liver Biopsy

Surgery consultation

**INTERVENTION**

Chemotherapy followed by reassessment of surgical resectability. If possible, this is followed by surgical resection of primary tumor and extrahepatic disease. Additional chemotherapy will follow if the primary tumor was completely resected. The standard regimen is four courses of cisplatin/vincristine/fluorouracil [2] or doxorubicin/cisplatin combination chemotherapy [1,3] followed by attempted complete tumor resection. If the tumor is completely removed, two postoperative courses of the same chemotherapy should be given.

In a study employing a well-tolerated regimen of doxorubicin/cisplatin chemotherapy, about 50% of patients with metastases at presentation survived 5 years from diagnosis. Half of these survivors had developed progressive disease that was successfully treated with surgery and other interventions.[1] In another study, platinum- and doxorubicin-based multidrug chemotherapy induced complete regression in approximately 50% of patients, with subsequent 3-year EFS of 56%. [4]

Several studies have tested different chemotherapy regimens. A randomized clinical trial compared cisplatin/vincristine/fluorouracil with cisplatin/doxorubicin. Although outcome was nominally higher for children receiving cisplatin/doxorubicin, this difference was not statistically significant, and the combination of cisplatin/vincristine/fluorouracil was less toxic than the regimen of cisplatin/doxorubicin.[2] The cisplatin/doxorubicin used in the international studies appears to be less toxic than that in the North American study.[1] Addition of carboplatin to intensify the cisplatin/doxorubicin may have reduced its efficacy.[5] A regimen of intensified

platinum therapy with alternating cisplatin and carboplatin was associated with a decrease in EFS.[6] A combination of ifosfamide, cisplatin, and doxorubicin has also been successfully used in the treatment of advanced-stage disease.[7]

If possible, stage IV patients with resected primary tumor should have remaining pulmonary metastases surgically removed.[3] A review of patients treated on a U.S. Intergroup trial suggested that resection may be done at the time of resection of the primary tumor.[8]

## RESULTS

- After we perform a CT scanning and took the AFP results we conclude that the abdominal mass was a liver tumor and according to the radiology also biochemical probably was Hepatoblastoma which for sure we must ask a liver biopsy. Biopsy answer was Fetal type of Hepatoblastoma.
- Were pre-treated with chemotherapy (CISP+ Doxo ), according to the classification SIOPEL PRETEX, and after three cycles performed for reduction of the amount, surgery attended a left lobectomy. Post-operative complicated with an abdominal fistula and after an inefficient drainage the child undergoes another laparotomy for a hepato-duodenal bypass, according Kasay.
- After surgery we post-treated with chemotherapy ( CISP+ Adria).
- The child undergoes the follow up for AFP ( normalized ), abdominal ultrasound, X-Ray.
- Right now he is an healthy child.

## CONCLUSIONS

- Increasing the attention and care of paediatrician of infants during periodic controls.
- Hepatoblastoma, although is a rare type of liver tumors, not always presented in its clinical signs and not always associated with a genetic or metabolic syndrome.
- If is evaluated in time, the quality of life and chances of survival increase substantially.

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