Abstract:
We report a 2-years-old boy presented with a liver mass, which on biopsy was proven to be hepatoblastoma. Chest x-ray and CT scan showed a mass lesion in right paravertebral region which on biopsy was proven to be neuroblastoma. Child was treated successfully by giving chemotherapy and surgically resected these two masses.

Keyword: Hepatoblastoma, Neuroblastoma

1. Case Report:
A 2 years old boy who presented with a liver mass (Figure 1) of 2 months duration. Initial evaluation showed elevated alpha fetoprotein (AFP) >300 IU/ml and trucut biopsy of the mass confirmed epithelial type of hepatoblastoma (HB). Initial chest X-ray and CT thorax revealed a right posterior mediastinal mass (Figure 2) and since the mass was large (8x5.2x3.4 cm, extending from D_3 to D_10), it was suspected to be separate malignancy rather than a metastasis from HB. CT guided needle biopsy of the mediastinal mass showed differentiating type neuroblastoma. Bone scan was negative for skeletal metastasis. His PRETEXT stage (Pretreatment extent of disease) was II V0P0E0C0M0 (V: Ingrowth venacava, all three hepatic veins; P: Ingrowth portal vein, portal bifurcation; E: Extrahepatic; C: Caudate; M: Metastasis). First he received 4 courses of PLADO chemotherapy (Cisplatin and Doxorubicin) for the treatment of HB. Subsequently he received 3 cycles of rapid COJEC protocol (Vincristine, Carboplatin, Etoposide, Cyclophosphamide) to treat the neuroblastoma. His POSTTEXT stage (Posttreatment extent of disease) was II V0P0E0C0M0. HB was located in segment V, VI of the liver. His AFP before surgery was 12400 IU/ml. He subsequently underwent nonanatomic resection of segment V and VI. Following this he received another 2 cycles of rapid COJEC. Then he underwent right thoracotomy and excision of neuroblastoma. Postoperatively he received one more cycle of rapid COJEC. At the end of his treatment, he was clinically well.
He came back for follow up after 9 months since completion of therapy with no evidence of residual or recurrent tumor. His AFP value was 1.13 IU/ml.

2. Discussion:
HB accounts around 80% of malignant liver tumor among pediatric age group. It is a rare pediatric neoplasm. Its incidence is about 1.5 per million. It comprises only about 1% of all pediatric malignancies. Different genetic syndromes associate with both hepatoblastoma and neuroblastoma. The association between Beckwith-Wiedemann syndrome (BWS) and neuroblastoma is quite strong. The genetic anomaly in HB is mapped to up 15.15 (P57 ki P2, WNT). They have increased risk of tumor development. The risk is greatest in first decade of life. The most frequently observed tumors in BWS are wilms’ tumor and hepatoblastoma, which comprise 43%, 12% of reported cancers, respectively. Neuroblastoma has also been observed in infants with BWS. The nuclear phosphoprotein gene TP53 has been recognized as an important tumor suppressor gene, perhaps the most commonly altered gene in all human cancers. Inactivating mutations of the TP53 gene also cause the TP53 protein to loose its ability to regulate the cell cycle. Heritable cancer associated changes in TP53 tumor suppressor gene occur in families with Li-Fraumeni syndrome (LFS). It is an autosomal dominant hereditary disorder. Malignancies seen in this syndrome include breast cancer, soft tissue sarcoma (Rhabdomyosarcoma), osteosarcoma, brain tumor-especially glioblastoma, leukemia, lymphoma and adrenocortical carcinoma. J.R.Gutweiler, D.C.Yu, H.B.Kim, et al. reported a 2 year old boy with stage IV neuroblastoma which was treated rigorously with chemotherapy, radiation and bone marrow transplant. On follow-up he developed a liver mass. Initial imaging and trucut biopsy of the liver mass were consistent with focal nodular hyperplasia (FNH). After left lateral segmentectomy, histopathological examination revealed the specimen mostly consistent with small cell undifferentiated hepatoblastoma as well as 3 foci of FNH in surrounding parenchyma. Here HB developed as second malignancy. But our case is unique because there was simultaneous presentation of two different solid tumors.

3. Conclusion: There is no report in the literature of simultaneous presentation of two different solid tumors.
hepatoblastoma and neuroblastoma. Synchronous malignancy or different solid tumors in a child can be diagnosed when there is a strong suspicion and investigation should be done accordingly.

References


