Hepatoblastoma : A rare presentation in elderly.

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Abstract:

Hepatoblastoma is one of most common types of liver cancer. However it is comparatively a rare variety of solid tumor in paediatric age. The disease usually affects children below the age of 3 years. Hepatoblastoma (HB) accounts for 79 % of all the liver tumors in children. Though it is usually seen in childhood, few cases of hepatoblastoma has also been reported in adults worldwide. Studies suggest the disease is more common in males as compared to females. The etiology of HB has not been yet fully understood. Alpha Feto Protein (AFP) is a sensitive and specific marker of the tumor for early diagnosis and monitoring. The complete surgical resection is the most important modality of treatment. Chemotherapy has been proven beneficial in both adjuvant and neo-adjuvant treatment. However inspite of all these available therapies, the prognosis of hepatoblastoma in adults is yet to be fully understood.

Keywords: - Adults, Alpha feto protein Mesenchymal tumor, Hepatoblastoma.

Introduction:

Hepatoblastoma is the most common form of liver tumor however it is comparatively a rare variety of solid tumor in paediatric age group[1].The disease usually affects children below the age of 3 years[2].This tumor primarily originates from fetal liver cell, mature liver cell and/or bile duct cells and usually present clinically as an abdominal mass. Alpha Feto Protein (AFP) is a sensitive and specific marker of the tumor for early diagnosis and monitoring[3].

Hepatoblastoma (HB) accounts for 79 % of all the liver tumors in children and the annual incidences of HB in infant <1 year of age is 11.2 per million. A significant high rate of HB is observed in infants with low birth weight (LBW) and very low birth weight (VLBW) children. The incidences of HB decrease rapidly after the age of 5 years. Only few cases of HB has been reported in adolescent and middle age persons with only four cases been reported above the age of 70 years worldwide[4]. Hence we are reporting a case of HB who presented at the age of 84

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years.

Case Report:

A 84 year old male patient was admitted to the medical ward of MB Govt Hospital and RNT Medical College, Udaipur, Rajasthan with the history of fever, cough, shortness of breath, chest pain on the left side and abdominal mass with loss of weight and appetite for the last six months. All these symptoms are progressively increasing for which the patient had consulted many physicians and he was prescribed with antibiotic and anti pyretic treatment. But he had no improvement. Patient had no significant past history in the form of any hospitalization, major surgery or major treatment for any major illness. He was a non smoker and had no other addiction in the form of alcohol or tobacco chewing. Then the patient was examined thoroughly and found that he had pallor, severe cachexia without any jaundice, clubbing, lymphadenopathy or edema feet. His weight was 40 kg and BMI was 16.8kg/m2. Respiratory system examination revealed left sided pleural effusion and abdominal examination revealed a mass of approximately 8*7 cm in the epigastric area which was globular and hard in consistency with sharp margins and irregular surface and was probably originating from the left lobe of liver.

He was then investigated and was found that his Hemoglobin was 8.6g/dl while the TLC and DLC were in the normal range. The ESR was 25 mm with normal renal function tests. His liver function tests revealed that total bilirubin was 0.3mg/dl, SGPT and SGOT were marginally high (67 and 47 U/L respectively),ALP was 312 U/L ,total serum protein was 5.9g/dl and albumin was 2.5g/dl with reversal of albumin : Globulin ratio. Chest xray was done showing moderate left sided pleural effusion which was aspirated and was hemorrhagic in nature and cytobiochemical investigation of the same revealed Protein 4.3g/dl, Sugar 107.6mg/dl, Adenosine Deaminase 18.3 IU/L, Total cells 350 cell/cumm with Neutrophils 20%,Lymphocytes 70% ,RBC 10% and no atypical cells seen.

Patient underwent Ultra sonography abdomen which showed a ill defined heterogenous mass of approximately 57*32 mm originating from the left lobe of liver causing mass effect on adjacent parenchyma which was suggestive of ?Hemangioma or ?cholangiocarcinoma (Image 1).

These findings were confirmed by Contrast enhanced triple phased computed tomography (CECT) which revealed a heterogeneously enhancing soft tissue mass of 63*51*53 mm with internal low attenuation areas seen in left lobe of the liver with infiltrating segmental branches spreading into left lower bronchus, left descending pulmonary artery and left descending pulmonary vein suggestive of a primary liver malignancy with multiple metastasis(Image 2) .

These findings were further confirmed by Fine Needle Aspiration Cytology of liver (FNAC) which showed clumps of small cells with high Nucleo-Cytoplasmic ratio and mild nuclear pleomorphism. At some places cells are in trabecular pattern while at other places cells are lying singly. Few cells were showing hepatocellular differentiation with bilirubin formations. These findings were consistent with a diagnosis of Hepatoblastoma (Image 3).This was further confirmed by serum Alpha feto protein which was highly raised 708

IU/ml (Normal <10 IU/ml).CA 125 was 97.06 U/ml and CEA 8.70 ng/ml .All these findings were highly suggestive of a pleuripotent type tumor i.e. Hepatoblastoma (HB).

Image 1: USG Showing ill defined Heterogenous mass originating from the left lobe of liver

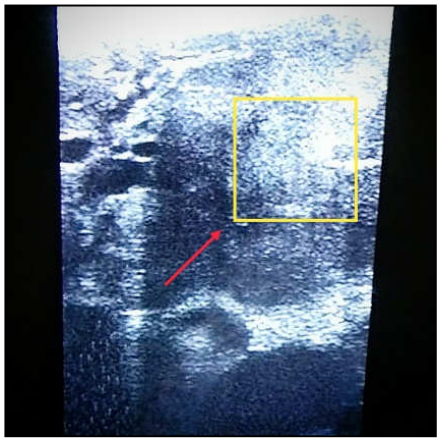


Image 2: CECT revealing a heterogeneously enhancing soft tissue mass in left lobe of the liver

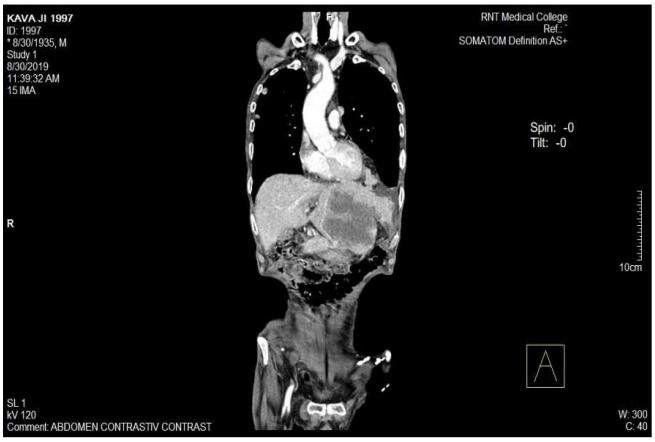
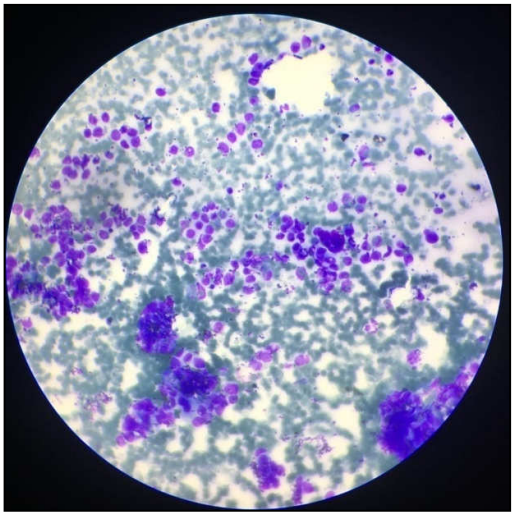


Image 3: Fine Needle Aspiration Cytology of liver showing clumps of small cells with high Nucleo-Cytoplasmic ratio and mild nuclear pleomorphism suggestive of hepatoblastoma



Discussion:

Hepatoblastoma is one of the most common primary malignant neoplasms of the liver in children and around 90 % of the cases are found usually before the age of 5years[5].HB in adolescents ,young adults is a rare disease and is further rarest above the age of 70 years. Some studies suggests the disease is more common in males as compared to females .The male : Female ratio is 1.7:1.The etiology of HB has not been yet fully understood. The present cytogenetic and molecular genetic studies reveals involvement of chromosomal loci on 1q,2q,4q,8q etc and these may have association with HB[6].Based on the embryological theory it is believed that HB arises from hepatoblasts, however this hypothesis is not applicable in HB of adulthood and old age. The persistence of primitive hepatoblasts for such a long period seems unlikely and this would indicate that theses tumors in old age has

different pathogenic pathway as compared to that of children.

Ishak and Glunz classify Hepatoblastoma into two groups namely epithelial type and mixed epithelial and mesenchymal type[7].The epithelial type consists of fetal and embryonic cell. They may present alone or in combination. In the next variety i.e mixed epithelio – mesenchymal type, the mesenchymal elements are seen along with epithelial component. For a long time it has been thought that hepatoblastoma develops during intra uterine life but the same histological pattern seen in adults and old age patients suggest the contrary. Many new interesting theories are now developing and one theory suggests that common hepatocytes could be at the starting point of tumor genesis after they lose differentiation and get transformed into blast cells.

In adult and old age the morbidity of HB is extremely high due to late diagnosis and because of non specific initial presentation[8].The usual clinical presentation of HB are abdominal lump, loss of weight and anorexia etc. The serum AFP are almost always very high ,such as in our case (AFP 708 IU/ml).The initial diagnosis of HB is mainly based on investigation such as USG ,CT scan ,MRI which shows hyperechoic solid intrahepatic mass which can further be confirmed by high or very high AFP along with histopathological diagnosis.The final diagnosis depends on the biopsy of tumor in adults. However there may be several similar varieties of tumor mimicking the diagnosis like hepatic teratoma, carcinoma, sarcoma, malignant mesenchymal tumors and hepatocellular carcinoma with sarcomatous changes[9].

The complete surgical resection is the most important modality of treatment and it has good clinical results. Chemotherapy has been proven beneficial in both adjuvant and neoadjuvant treatment that can shrink the tumor size [10].It makes the tumor less prone to bleed and it delineates the tumor from the surrounding parenchyma. HB is sensitive to chemotherapeutic agents such as Doxorubicin, Cisplatin, Vincristine and cyclophosphamide[11].

Conclusion:

Hepatoblastoma is one of the rarest liver malignancy presenting in adolescent and old age people .The very low incidences of this disease in old age and its non specific presentation makes the diagnosis difficult. These patients may present in very late stage of the disease with spread of the malignancy to different organs of the body. There is no consensus on the treatment of the disease due to rarity of the disease in old age. Most of the patients diagnosed have expired before surgery or soon after surgery. This represents a diagnostic challenge because prognosis could be further improved with early detection.

References:

1. Kasper H.U., Longerich T., Stippel D.L., Kern M.A., Drebber U., Schirmacher P. Mixed hepatoblastoma in an adult. Archives of Pathology and Laboratory Medicine. 2005;129:234–237
2. L. Bortolasi, L. Marchiori, I. Dal Dosso, R. Colombari, and N. Nicoli, “Hepatoblastoma in adult age: a report of two cases,” Hepato-Gastroenterology, vol. 43, no. 10, pp. 1073–1078, 1996.

3. H.-Y. Ke, J.-H. Chen, Y.-M. Jen et al., "Ruptured hepatoblastoma with massive internal bleeding in an adult," World Journal of Gastroenterology, vol. 11, no. 39, pp. 6235–6237, 2005.
4. Kasper HU, Longerich T, Stippel DL, Kern MA, Drebber U, Schirmacher P. Mixed hepatoblastoma in an adult. Arch Pathol Lab Med. 2005;129(2):234–237
5. Lack EE, Neave C, Vawter GF. Hepatoblastoma. A clinical and pathologic study of 54 cases. Am J Surg Pathol. 1982;6(8):693–705
6. Nagata T, Nakamura M, Shichino H, Chin M, Sugito K, Ikeda T, Koshinaga T. et al. Cytogenetic abnormalities in hepatoblastoma: report of two new cases and review of the literature suggesting imbalance of chromosomal regions on chromosomes 1, 4, and 12. Cancer Genet Cytogenet. 2005;156(1):8–13
7. Ishak KG, Glunz PR. Hepatoblastoma and hepatocarcinoma in infancy and childhood. Report of 47 cases. Cancer. 1967;20(3):396–422
8. Ahn HJ, Kwon KW, Choi YJ, Kim HJ, Hong SP, Oh D, Chung JS. Mixed hepatoblastoma in an adult--a case report and literature review. J Korean Med Sci. 1997;12(4):369–373
9. Oda H, Honda K, Hara M, Arase Y, Ikeda K, Kumada H. Hepatoblastoma in an 82-year-old man. An autopsy case report. Acta Pathol Jpn. 1990;40(3):212–218
10. A. P. Pimpalwar, K. Sharif, P. Ramani et al., "Strategy for hepatoblastoma management: transplant versus nontransplant surgery," Journal of Pediatric Surgery, vol. 37, no. 2, pp. 240–245, 2002
11. Reynolds M. Pediatric liver tumors. Semin Surg Oncol. 1999;16(2):159–172