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Hepatic Resection in Children with Cardio-Pulmonary Bypass

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Abstract
Introduction: *Common indications of hepatic resection in children are primary liver tumours, localized secondaries in the liver, trauma, localized Caroli's disease etc. Refinements in the understanding of surgical anatomy of the liver along with better imaging modalities have accounted for greater percentage of resectable tumours and reduced blood loss resulting in significantly reduced mortality in recent times.*
Materials: *From October 2015 until September 2017, hepatic resections were performed in four children between 23 months to 10 years of age at different surgery unit of Hospitals. Two of these patients had Mesenchymal hamartomas, one had malignant mesenchymoma and the other one patient had type IV Choledochal cyst, the intrahepatic part confined to left lobe of the liver.*
Results: *Right hepatic lobectomy was done in three patients and left hepatic lobectomy in one patient. There was no operative mortality. Mean operating time was 285 minutes (210 to 360 minutes); average bypass time was 105 minutes (90 to 120 minutes); average operating blood loss was 750 ml (500 to 1000ml). Average hospital stay was 12 days (8 to 16 days).The patient with malignant mesenchymoma had died 6 weeks following surgery due to complications of chemotherapy. The other three patients had uneventful recovery. One patient of mesenchymal hamartoma lost to follow up after 3 months of surgery. The other patients are on regular follow up for 24 months, 16 months, 6 months respectively, and all of them are doing well.*
Conclusion: *Hepatic resections in children are challenging problem. Although malignant tumours of the liver are the commonest indications of major liver resections, only one of our patients had malignant tumour. Late presentation with disseminated disease may be the reason for low incidence of malignant tumours needing hepatic resection under cardiopulmonary bypass in our series.*

Introduction

Liver resections were first performed in the late 1800s. Tiffany (1890) performed the first recorded successful resection of Liver & Luke (1891).Elliot (1897) reports his attempted resection of Liver Tumor. Hepatic resection done earlier was plagued by high morbidity and mortality rates, which were largely related to massive intraoperative blood loss. In 1952, Lortat-Jacob was given credit for the first true anatomic right hepatectomy.

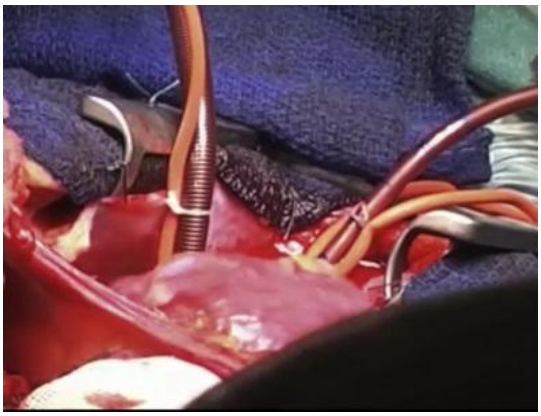
The single most important development in operative technique has been one that has resulted from a better appreciation of the segmental nature of hepatic anatomy .It is resulting in an increasing use of anatomically based resections and, more importantly, an increased use of parenchyma-sparing segmental resections. A precise knowledge of the anatomy of the liver is an absolute prerequisite to performing surgery on the liver or biliary tree-which is most difficult in tiny liver of children. However, using Cardio-pulmonary bypass during surgery not only reduces mortality but also less bleeding and clearer operative field of vision. Role of a trained

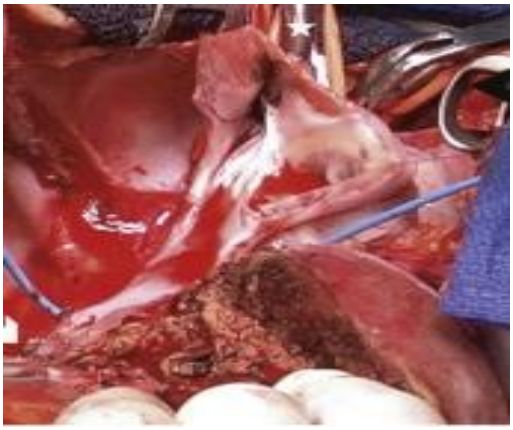
cardiothoracic and vascular surgeon who is also well trained in general surgery is enormous.

Materials and Methods

This report gives details of major hepatic resection of centers from October 2015 to September 2017 of 4 children age ranged from 23 months to 10 years. Among them, three were male & one was female. Two patients had benign hepatic mass, one had primary malignant hepatic tumor and one patient had choledochal cyst with intra-hepatic involvement.

Cardiopulmonary bypass procedure was started by usual methods before hepatic resection. No requirement of deep hypothermic arrest provided an early recovery.





Results

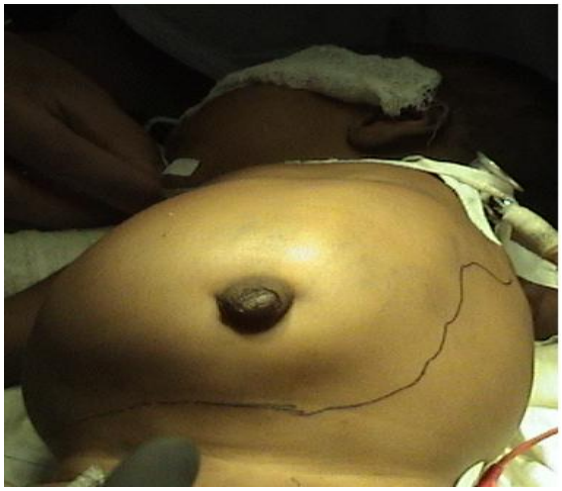
Two patients having benign hepatic mass & one patient having primary hepatic malignant tumor underwent Rt. lobectomy & patient having Todani's type IVA choledochal cyst underwent left lobectomy of liver with mucosectomy of residual cyst with rough-n-y hepatico-jejunostomy. Among them, one patient having primary hepatic malignant tumour died 3 months after surgery because of progressive disease process & complication of Chemotherapy. Others 3 are followed up with uneventful recovery for minimum 6 months to 1.5 years and more.

Case Summery

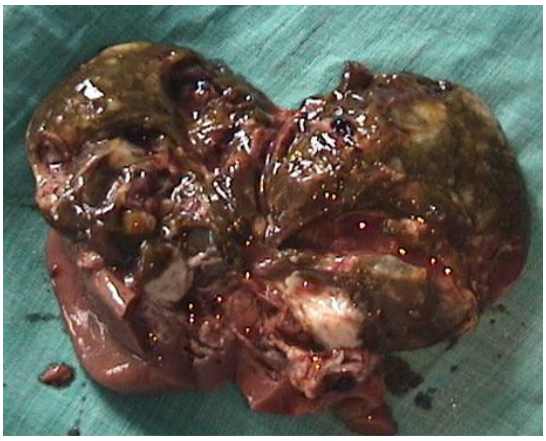
Patient	Presentation	Investigations	Treatment	Result
Master NC, 10y, 29.5kg, Male	Abdominal Lump, Distension, Const. O/E-lump Rt. Abdomen Continuous with Liver Felt rectally	CXR – ↑ Rt. Hemi diaphragm S.AFP-n. CECT- Mass Rt. Hepatic Lobe with Solid & Cystic Component.	Rt. Hepatectomy	Uneventful Recovery. F/U > 1½ Yrs.
Master RB, 2y, 14.2 Kg, Male	Abdominal Distention with Lump - 2m Resp. Distr. X 15d O/E- Liver Up To RIF, firm, Tender Pedal Edema +	CXR-n, S.AFP-n. CECT- Mass Rt. Hepatic Lobe with Solid & Cystic Component.	Rt. Hepatectomy	Uneventful Recovery. F/U: 6 Months

Ms. DS, 8 yrs., 25 Kg, Female	Rec. Colicky Abdominal Pain & Intermit. Jaundice X 1yr. O/E- Tender Rt. Hypochondrium	LFT-N, GGTP-↑ N USG- Choledochal Cyst, Type IV. ? CBD Stricture. MRCP- Choledochal Cyst Confined to Lt. Lobe of Liver.	Exc. of Choledochal Cyst + Lt. Hepatectomy, Mucosectomy + Roux-en-Y Hepatico-jejunostomy.	Uneventful Recovery , Well > 2 Yrs.
Master KN, 6 yrs., 26kg, Male	Abdominal Mass X 2M Gen Weakness, Wt. Loss O/E-liver Mass 4 Cm Below Costal Margin , firm	CXR - N. S.AFP-N. CECT - Hypo vascular Mass Rt. Lobe of Liver with Necrosis	Rt. Hepatectomy. Post-op. Chemothx.	Pt. Died 3 Months post-op due to Progressive Disease & Complication of Chemothx.

Our 2nd patient this 23 month old 14.2 Kg, male child presented with progressive abdominal distension & respiratory distress for 15 days. Liver was enlarged up to RIF, firm, tender.No ascites, and engorged veins were there but minimal pedal edema was present. Routine investigations were within normal limit. CECT revealed mass arising from segment V, VI, VII & VIII of liver.

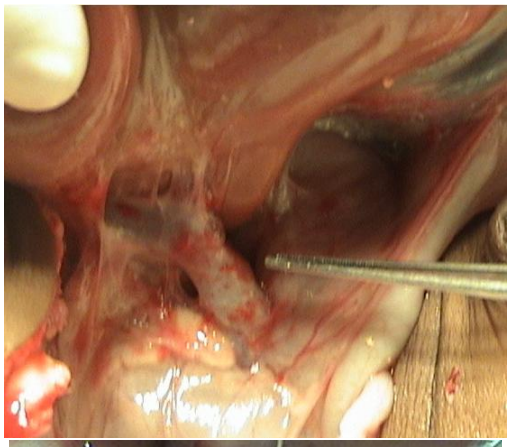


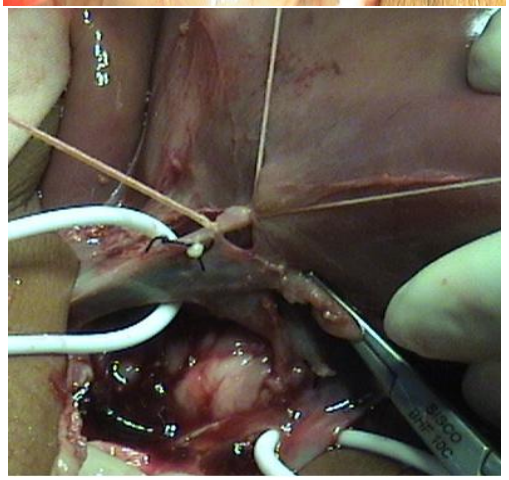
After Rt. Hepatectomy, the cut section shows Rt. Lobe of liver with the mass with areas of hemorrhage & necrosis as shown in the picture here.



Steps of Hepatic Resections

Depending of the procedure & body habitus of the patient incision was selected. Mostly Rt. Subcostal incision with thoracic extension was made, rooftop incision in one case. Preparation of the GI tract was done for surgery with hepatomegaly of known or unknown aetiology. Proper positioning of the patient was done on O.T table. Perihepatic adhesions were cut carefully to avoid injury. Occasionally these adhesions are vascular. Electric cautery was used. Relation of the liver to the diaphragm above and the several anatomic entities was clearly delineated. Right triangular and coronary ligaments were incised. The gallbladder was sacrificed.





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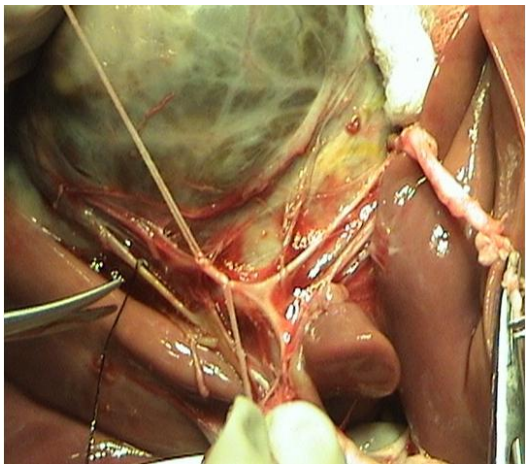


B

Picture A shows infra-hepatic IVC. Picture on the middle shows Rt. Hepatic vein was ligated & divided. Picture B shows portal structures were properly identified & individual branches supplying Rt. Lobe were ligated & divided.

Dissection started at the hilum, branches of the hepatic artery, portal vein, and bile duct of the lobe to be removed were ligated. The line of the interlobar fissure (line of Rex, median fissure) extends from the gallbladder fossa below to the inferior vena cava above - pass to the right of the middle hepatic vein to preserve drainage of segment IV was scored with electro-cautery. For left hepatectomy or left lobectomy the left hepatic artery, portal vein, and bile duct were ligated. Sectioning of the triangular ligament permitted us for mobilization of the left lobe. Transection was done following a line from the left side of the fossa of the gallbladder to the left side of the fossa of the inferior vena cava. Exposure and ligature of the left and middle hepatic veins within the liver or extra hepatically at the vena cava was done after confirmation of the precise anatomy. Extreme care was taken to preserve the left hepatic vein as the middle vein typically joins it prior to its junction with the vena cava. Liver was resected with finger fracture or fracture with artery forceps. Individual bile duct or bleeding vessels was ligated. In our operation of type IVA choledochal cyst mucosectomy of intrahepatic cyst along with Roux-en-Y hepatico jejunostomy was done.

Post Operative Managements

In all these patients, post-operatively we have given IVF with 10% Fructodex & Isolyte-P alternatively. Patient was catheterized. Urine output maintained @ 1-2 ml/kg./hr.FFP 50-100 ml was given daily for 3 to 5 days post operatively. Monitoring of Hb%, Serum Glucose, Serum Bilirubin was done daily for 5 days along with serum electrolytes. PT, LFT was done on day 1, 3 & 5 and when required. Post-operative raised level of bilirubin became normal with 6 days. Post-operative Chest X-ray was done on 7th day. We repeat USG before discharge, at 1 & 3 months of follow-up.

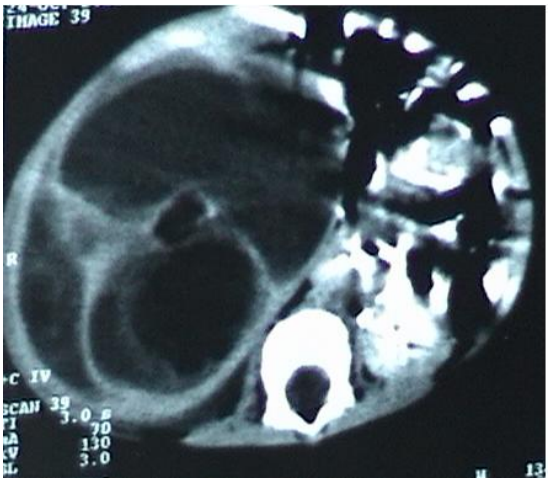
Discussion

Primary liver tumour account for 15% of childhood abdominal tumors. They are usually epithelial or mesenchymal in origin and may be benign or malignant.

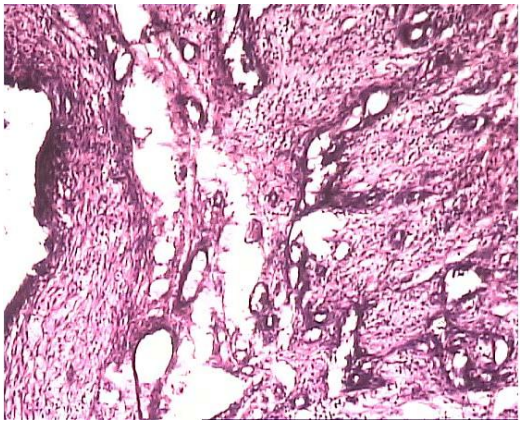
Mesenchymal Hamartoma: It is the 2nd most common benign tumor in children in less than 2 yrs. of age group. Almost ⅔rd. of it occurs below

1 yr. age and approximately 1/5th in neonates. Almost all occur below 5 yrs. of age. Mesenchymal hamartoma usually presents with painless abdominal mass & distension. Occasionally, respiratory depression, vomiting, change of bowel habit, wt. loss.Congestive heart failure may occur secondary to AV shunting. Usually confined to one lobe .Contain both cystic & solid component & may be pedunculated. Cysts are up to 9 cm in diameter & max. wt. 2000 gms. It represents a failure of normal development and arises from the connective tissue tracts within the fetal liver. Edmonton proposed that mesenchymal hamartoma arises from a mesenchymal rest. The tumor grows along bile ducts and may incorporate normal liver tissue. Bile duct, liver tissue & angiomatous components are typically found. Biological behaviour of the tumor depends on the relative predominance of these components. Ultra-structural appearance is well diff. ductal structures surrounded by loose mesenchymes containing fibroblast (Dehner et al, 1975).Okeda proposed a pathogenetic relationship with defective vascularisation. Degeneration and fluid accumulation within the cystic component cause progressive enlargement of this structure (Stocker & Ishak, 1983).Resection with enucleation or lobectomy is almost curative. Unroofing & marsupialization for very large cyst, although the lesion may recur after incomplete resection. The 1st case of Mesenchymal Hamartoma was reported in 1903 [1] .Since then approximately 140 cases have been reported in different world journals [2] . Almost all had shown excellent long-term prognosis [4, 5] .

Our 1st patient 10 Year old male presented to us with abdominal lump for 2 months, progressive abdominal distension, constipation (often-requiring enema) for 15 days. His CECT showed mass in Rt. Hepatic lobe with solid & cystic component involving segments V, VI, VII & VIII as in the picture given here –



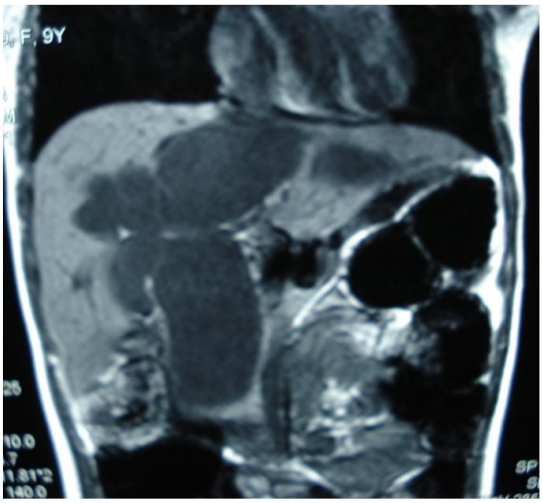
After Rt. Hepatectomy the mass shows histopathologically ducts lined by low cuboidal epithelium, cystic spaces with loose connective tissue stroma suggestive of Mesenchymal hamartomas like the given picture. Patient is followed up for approx. 1.5 yrs. & he is doing well.

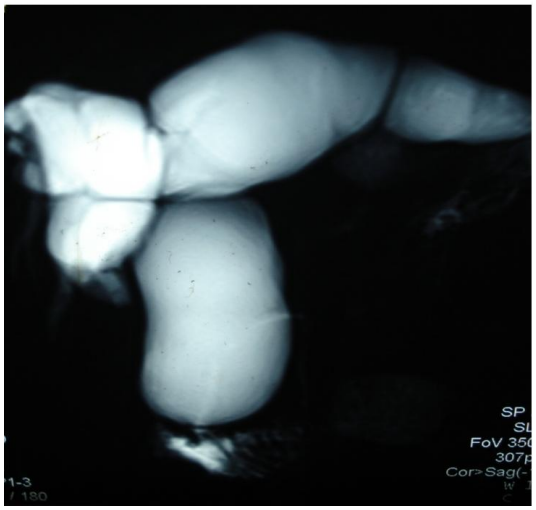


Choledochal Cyst: These are the Focal or diffuse dilatation of the biliary tract.Mostly congenital in origin. Incidence is approximately 1:10000 - 150000.Female predominance is found with M: F= 1:4. Type IVA of Todani's comprise 4% to 28.5% in various reports[6]. Treatment of choice is excision of extrahepatic duct & dilated hepatic ducts and wide biliary anastomoses. Hepatic lobectomy is the most preferable approach if dilatation is confined to one hepatic lobe. Remnant of Intrahepatic cyst may cause-cholangitis, biliary cirrhosis, proximal ductal ectasia, calculus formation & malignancy. Cholangitis (31%-44%)[7]; Stricture (40%)[8]; Malignant degeneration (14% - 18%) [9, 10] are the

few common complications that was reported. Mucosectomy may help alleviate or prevent these complications[11]. Vater and Ezler first described it in 1723. Since then approximately 400 cases has been described world-widely[12]. None of the case from India has reported regarding mucosectomy of residual intra-hepatic cyst.

Our 3rd patient 8 yrs., 25 Kg, female↑ in MRCP showed extrahepatic fusiform choledochal cyst ,intra-hepatic cyst confined to Lt. lobe of liver(choledochal cyst, type IVA of Todani's Classification) as shown in this picture. Excision of extrahepatic choledochal cyst, Lt. Hepatectomy, mucosectomy of residual cyst wall & Roux-en-Y hepatico-jejunostomy was done in this patient. Patient recovered well and her postoperative MRCP showed adequate decompression of residual cystic spaces. Patient on her follow-up never developed cholangitis or jaundice.





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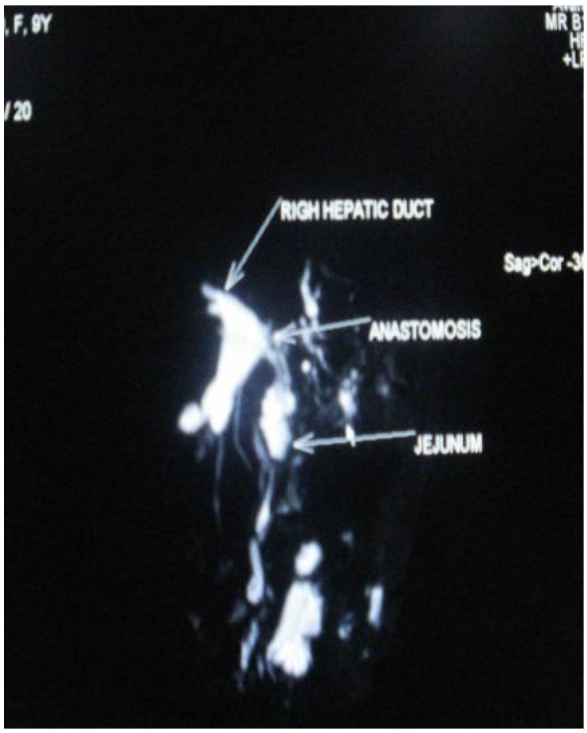


B

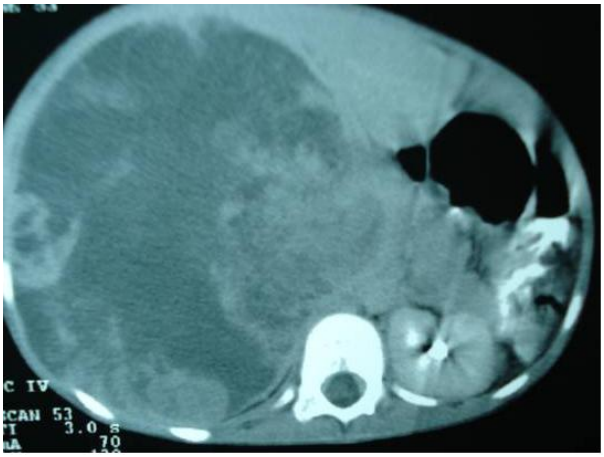
Comparative picture of pre-operative (A) & post-operative MRCP (B) was showing adequate decompression of residual intra-hepatic cyst.

Malignant Mesenchymoma

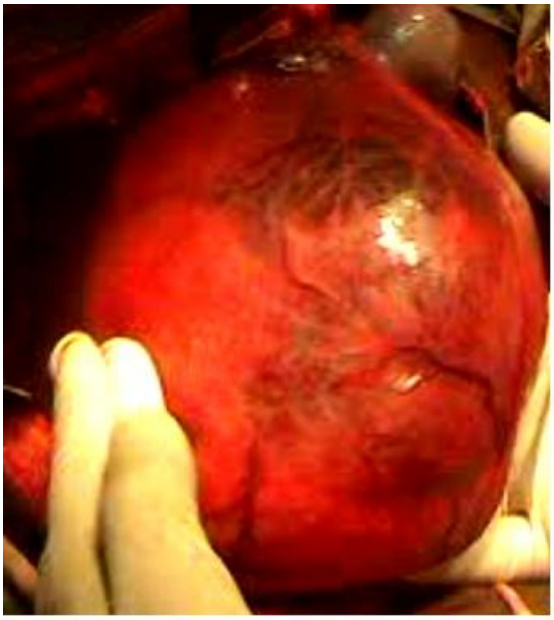
It is a rare soft tissue sarcoma showing two or more distinct types of malignant mesenchymal differentiation. Malignant mesenchymoma is the 3rd most common after hepatocellular carcinoma & hepatoblastoma in children [13] . Three most common components it contains Liposarcoma, Rhabdomyosarcoma and Osteosarcoma. However, it is more common in adults in children found in mainly 5-10 yrs. age group .Frequently located in the trunk and lower extremities. Generally, they are high-grade sarcomas & usually showed poor prognosis. First large series (31 cases) was reported by Stocker & Ishak in 1978 [14] . Less than 100 cases have been reported until today [13] .

Our 4th patient a 6 yrs., male having 26kg weight presented to us with abdominal mass for 2 months and generalized weakness for last 15 days along with associated weight loss. On examination, there was palpable liver mass 4 cm below costal margin, firm & non-tender. During investigation, we found CxR- N. and S.AFP-N. CECT as shown

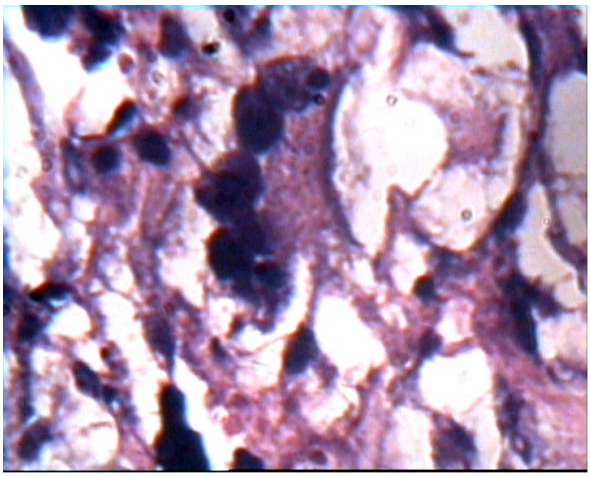
in photograph showed hypovascular mass in right lobe of liver with necrosis.



No evidence of intra-abdominal lymphadenopathy or other organomegaly was found. FANC was inconclusive so we planned for surgery .Intra operatively we found fleshy mass confined to right lobe of liver as we have shown in the photograph, left lobe is normal. No evidence of intra-abdominal metastasis or lymphadenopathy was there.



Biopsy of Rt. Hepatectomy showed in low power HE: Highly pleomorphic cells, some round to oval, others spindle shaped with pleomorphic and bizarre nuclei with occasional multinucleation and giant nuclei suggestive of malignant mesenchymoma. HP as per picture given here showed finding of multinucleated GC characteristic of malignant mesenchymoma and so, diagnosis was confirmed. Patient was treated with Vancomycin, Actinomycin-C & Ifosamide. This patient died 2 months after surgery because of progressive disease & complication of Chemotherapy.



Conclusion

Hepatic resections in children are challenging problem. Although malignant tumours of the liver are the commonest indications of major liver resections, only one of our patients had malignant tumour. Late presentation with disseminated disease may be the reason for low incidence of malignant tumours needing hepatic resection in our series. Using of Cardiopulmonary Bypass procedure using proper techniques and trained anaesthesia reduces much effort of the surgeon and the chief surgeon gets an extra helping hand during surgery. It's also pertinent to mention that inadvertent injury of the any major vessels during operations more easily controlled by the vascular surgeon at bypass. However increased cost for such procedures cannot be kept aside in conclusions.

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