VOLUME - 9, ISSUE - 12, DECEMBER - 2020 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Surgery

A RARE CASE OF UNDIFFERENTIATED EMBRYONAL SARCOMA OF LIVER IN 8 YEAR FEMALE CHILD

Dr. Asit V Patel	Professor and Head, Department of General Surgery, AMCMET Medical College, Ahmedabad.
Dr. Shreosi P Sarkar	Third Year Resident, Department of General Surgery,AMCMET Medical college, Ahmedabad.
Dr. Ashwin P Godbole	Second Year Resident, Department of General Surgery, AMCMET Medical college, Ahmedabad.
Dr. Dhruv N Shah*	Third Year Resident, Department of General Surgery, AMCMET Medical college, Ahmedabad.*Corresponding Author
Dr. Poorn V Pandya	First Year Resident, Department of General Surgery, AMCMET Medical college, Ahmedabad.

ABSTRACT

Introduction: Undifferentiated embryonal sarcoma of liver (USEL) is a rare disease that 90% occurs in children between age group 6-10 years with no gender discrepancy. Diagnosis of USEL relies on postoperative pathology and immunohistochemistry.

Objective : This is a rare case report of an 8 year female patient who underwent excision of liver mass with left side lobectomy for a liver mass which turned out to be Undifferentiated Embryonal Sarcoma of Liver(USEL) on Histopathological analysis and Immunohistochemical examination.

Results : Patient was diagnosed to be having Gastrointestinal Stromal Tumor of stomach on radiological investigations but intraoperatively turned out to be a liver mass for which patient underwent excision of liver mass with left side lobectomy for a liver mass which turned out to be Undifferentiated Embryonal Sarcoma of Liver(USEL) on Histopathological analysis and Immunohistochemical examination. Patient underwent chemotherapy including ifosfamide, mesna and doxorubicin. Currently, the patient's physical status is normal with no distant metastases shown in PET-CT.

Conclusion : Undifferentiated Embryonal Sarcoma of Liver is a rare entity but with adequate surgical procedure and followup chemotherapy or radiotherapy has excellent prognosis

KEYWORDS : Undifferentiated embryonal sarcoma of liver (USEL), Liver, Immunohistochemistry

INTRODUCTION

Undifferentiated embryonal sarcoma of liver (USEL) is a rare disease that 90% occurs in children between age group 6-10 years with no gender discrepancy [1]Typically presents with pain, fever, abdominal mass and normal serum AFP. USEL is often misdiagnosed as other types of hepatic malignant tumors. Diagnosis of USEL relies on postoperative pathology and immunohistochemistry [2] It has difficult diagnosis because of overlapping of epidemiological, clinical, radiological findings with those of other liver tumors. According to current studies, management of USEL mainly comprises surgery with postoperative chemo/radiotherapy. It has good prognosis but large tumors may rupture and cause death.

CASE REPORT

An 8 year old female child brought to surgical OPD with severe abdominal pain and fullness since 1 month associated with early satiety. Patient had no any other complaint. On examination there was a diffuse lump in epigastrium and extending to left hypochondrium which could not be examined properly due to tenderness over it.

Patient was advised CECT abdomen (Figures 1 & 2) which showed 6x6x6 cm well defined multiloculated heterogeneously peripherally enhancing cystic lesion arising from body of stomach extending into greater omentum and abutting left lobe of liver p/o. myofibroblastic inflammatory pseudotumor of stomach appears more over GIST.

Patients AFP and liver functions were normal.

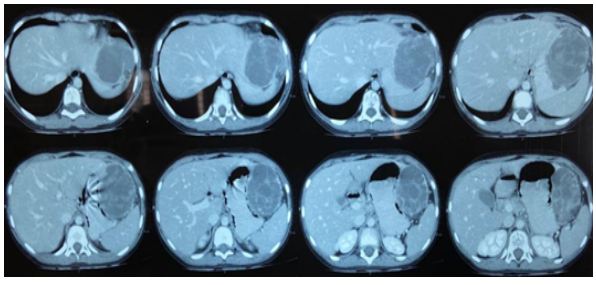

Figure 1

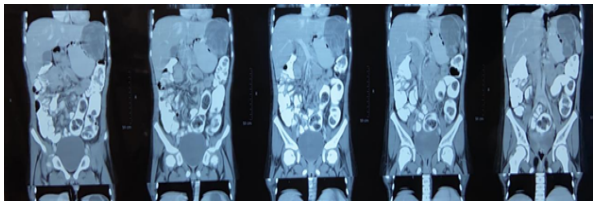

Figure 2

Since patient had severe abdominal pain unable to control with medical management exploratory laparotomy with excision of tumor sos partial gastrectomy was planned for the patient with working diagnosis of strongly tumor of stomach. Left subcostal incision kept with extension to the right. Intraoperatively, on opening peritoneum, a 9x7 cm massfound arising from left lobe of liver (Figure 3 & 4) which was free from stomach and surrounding structures. Rest of the liver was normal. Left Lobectomy performed and Hepatic mass was resected and liver margin was hemostasized. Small bowel, kidneys and spleen found to be normal.

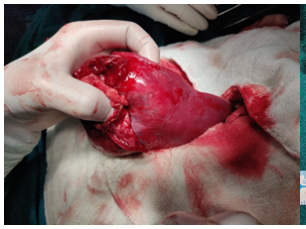

Figure 3

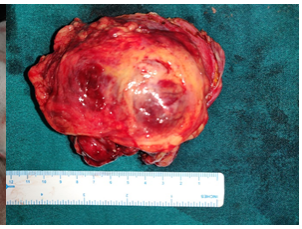

Figure 4

Cut surface found to be brownish, hemorrhagic and few whitish areas.

On microscopy (Figure 5 & 6), liver parenchyma was predominantly replaced by a tumor with a pseudocapsule and displaying fascicles and sheets of spindle to polygonal cells with moderate to marked nuclear atypia. Foci of extramedullary hematopoiesis, myxoidstroma with thin walled and ecstatic vessels along with entrapped hepatocytes and bile ducts seen. In areas there is complex and interconnecting vasculature lined by atypical cells, consistent with angiosarcoma component.

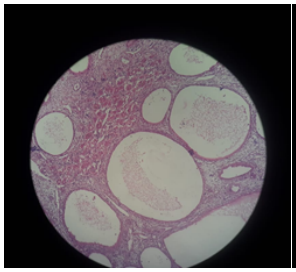

Figure 5

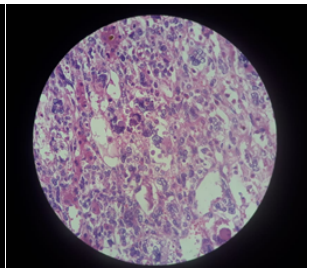

Figure 6

On immunohistochemistry, tumor was CD10, CD 31, CD 56, FLI 1, Desmin and SMA positive. It was negative for CD 117, SALL 4, Myogenin, Heppar 1 and CK. Diagnosis was consistent with undifferentiated embyonal sarcoma with foci of high grade angiosarcoma and a component of mesenchymal hamartoma.

DISCUSSION

Undifferentiated embryonal sarcoma of liver is also known as malignant mesenchymoma as it is considered to be a malignant evolution of mesenchymal hamartoma.it is 9-15% of pediatric hepatic tumors and rare in adults. It is a rare hepatic tumor and first case was reported in 1978 [3] Around 260 cases are found worldwide in pediatric age group by 2015. The tumor is mainly localized or found in the hepatic right lobe (59%), while it rarely develops in the hepatic left lobe (22%) or the bilateral lobe (20%). UESL typically has a diameter of 10-25 cm with a solitary clear boundary. Hemorrhage, necrosis and cystic degeneration are frequently observed while clinical manifestations include abdominal mass, pain, fever and rarely jaundice. [4] [5] A definite diagnosis of UESL is not able to be determined preoperatively; the diagnosis relies on postoperative pathological analysis and immuno histochemical results. UESL ought to be differentiated from hepatoblastoma, embryonalrhabdomyosarcoma, hepatic mesenchymal hamartoma and hepatic echinococcosis.. It is often associated with mesenchymal hamartoma and rarely with vaginal embryonal rhabdomyosarcoma.

Undifferentiated embryonal sarcoma of the liver usually occurs as a single and well-circumscribed lesion grossly. The well-demarcated appearance is created by a fibrous pseudocapsule [4]

It appears to be a primitive mesenchymal neoplasm with possible foci of differentiated sarcoma, such as angiosarcoma. USEL is considered to be highly invasive malignant tumor with primary hepatic mesenchymal tissue with distant metastases. Prognosis is not based on size or differentiation of tumor but is solely depends on invasion, diffusion and metastasis[6]The key management of the disease is total resection followed by postoperative combined therapeutic measures, including chemotherapy, radiotherapy and interventional therapy. In recent years, survival rates have significantly improved due to improvements in therapy, and long-term survival cases have been reported [6]Studies have shown improved survival rates ranging from 70% to 100% in patients who were treated with multimodal therapy. The recurrence rate in UESL is higher during the first 2 years after surgery and the risk is higher with positive resection margins and cases with spontaneous or iatrogenic rupture of the hepatic lesion. [7][8][9]

CONCLUSION

Patient underwent chemotherapy including Ifosfamide, Mesna and Doxorubicin. Currently, the patient's physical status is normal with no distant metastases shown in PET-CT. We conclude that preoperative and/or postoperative interventional therapy, combined with radiotherapy and chemotherapy, may improve survival rates and times in certain cases. The precise timing of the total surgical resection is crucial to prevent invasive growth of the tumor, and a liver transplantation is the most effective therapy for a patient whose tumor cannot be surgically resected.

REFERENCES

1. Primary undifferentiated embryonal sarcoma of the liver mistaken for hydatid disease. Faraj W, Mukherji D, El Majzoub N, Shamseddine A, Shamseddine A, Khalife M. 2010;8, World J Surg Oncol, p. 58.
2. Undifferentiated liver embryonal sarcoma in adults; a case report of four fases and literature review. Li XW, Gong SJ, Song WH, Zhu JJ, Pan CH, Wu MC, Xu AM. 2010;16, World J Gastroenterol, pp. 4725-4732.
3. Undifferentiated embryonal sarcoma of the liver in a child ; a case report and review of the literature. Gao J, Fei L, Li S, Cui K, Zhang J, et al. (2013). 2013, Oncol let 5, pp. 739-742.
4. Undifferentiated embryonal sarcome of the liver: case report and literature survey. Pachera S, Nishio H, Takahashi Y, Yokoyama Y, Oda K, et al. 2008; 15, J HepatobiliaryPancreatSurg, pp. 536-544.
5. Immunohistochemical analysis of embryonal sarcoma of the liver. Kiani B, Ferrel LD, Qualman S, Frankel WL. 2006;14, ApplImmunohistovhem Mol Morphol, pp. 193-197.
6. Undifferentiated embryonal sarcoma of liver in childhood: a curable disease. Bisogno G, Pilz T, Perilongo G, Ferrari A, Harms D, Ninfo V, Treuner J, Carli M. 2002;94, Cancer, pp. 252-257.
7. A single-institution retrospective case series of chilhood undifferentiated embryonal liver sarcoma (UELS):success of combined therapy and the use of orthoptic liver transplant. Plant AS, BussuttiRW, Rana A, Nelson SD, Auerbach M, et al. 2013;35, J PediatrHematoloncol, pp. 451-455.
8. Undifferentiated sarcoma of the liver: a single institution experience using a uniform treatment approach. may LT, Wang M, Albano E, Garrington T, Dishop M, et al. 2013;34, J PediatrHematolOncol, pp. 114-116.
9. Management of primary sarcoma. Weitz J, Klimstra DS, Cymes K, JarnaginWR, D'Angelica M, et al. 2007;109, Cancer, pp. 1391-1396.