CASE REPORT

Primary embryonal rhabdomyosarcoma of the liver

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SUMMARY

Rhabdomyosarcomas are malignant neoplasms with striated muscle differentiation. This is the most common type of soft-tissue sarcoma in children, but occurs rarely in adults. Its occurrence in liver is infrequent. We report a case of primary hepatic embryonal rhabdomyosarcoma in a 67-year-old man. The tumour was occupying the left lobe of the liver with large component of lesion seen bulging in left subhepatic space indenting over the stomach, compressing the pancreas and gall bladder. A percutaneous biopsy was performed which revealed embryonal rhabdomyosarcoma. He underwent a successful left lobectomy with complete resection of the tumour, followed by adjuvant chemotherapy. The patient is free of disease at 24 months of follow-up. Hence, an early diagnosis, prompt surgical resection with negative resection margin along with adjuvant chemotherapy can provide complete remission. This case is extremely rare both due to the age of the patient at presentation and location of the tumour.

derived skeletal muscle, or undifferentiated mesoderm. This is most common soft-tissue sarcoma of children and contributes 5% of all childhood

Figure 1 Ultrasound whole abdomen showing large complex mass (~12×11 cm) with hyperechoic heterogeneous texture in left hepatic lobe with extension into left subhepatic area with both cystic and solid components (marked with arrow).

BACKGROUND

Rhabdomyosarcoma (RMS) is a soft-tissue sarcoma with high malignant potential. It is from myotome-

Figure 2 Contrast-enhanced CT (axial section (A and B) and coronal section (C–E)) showing 14.5×12.3×9.1 cm heterogeneous hypodense mass lesion containing cystic and solid components noted involving left hepatic lobe with postcontrast enhancement in solid component. Large component of lesion is seen bulging in left subhepatic space indenting over stomach, compressing the pancreas and gall bladder.

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Figure 3 Intraoperative view of resected mass.

cancers.¹ Its occurrence in adults is rare. Most common sites of RMSs are head and neck, genitourinary organs, retroperitoneum and extremities.²⁻³ RMS in the liver is very rare. Until now only 13 cases of RMS of liver have been reported out of which, 6 cases are of embryonal RMS.²⁻⁷ Embryonal RMS in liver has never been reported in the elderly age group. We present a case of embryonal RMS of the liver in a 67-year-old man. These cases are difficult to manage because of their rarity and absence of standard treatment protocol or guidelines.

CASE PRESENTATION

A 67-year-old man admitted with symptoms of abdominal pain, abdominal distention and respiratory difficulty for past 1 month. General physical examination revealed tachypnoea and tenderness in epigastric quadrant with a mass palpable in the epigastric region.

INVESTIGATIONS

Blood chemistry (haemogram, renal and liver function tests) and chest X-ray were within normal limits. Hydatid, hepatitis B and hepatitis C serology were negative. Serum levels of α -fetoprotein and carcinoembryonic antigen levels were within normal limits. Ultrasound scan of the whole abdomen revealed a large complex mass with hyperechoic heterogeneous texture in left hepatic lobe with extension into the left subhepatic area. The lesion had both cystic and solid components. Biliary channel was not dilated. Portal and hepatic vein were normal (figure 1). Contrast-enhanced CT, abdominal and chest, showed a 14.5×12.3×9.1 cm heterogeneous hypodense lesion involving the left hepatic lobe, containing cystic and solid components with postcontrast enhancement in solid component. A large component of the lesion was seen bulging in the left subhepatic space indenting over the stomach, compressing the pancreas and gall bladder (figure 2).

Figure 4 Gross specimen showing (A) partly encapsulated tumour mass with nodular outer surface and (B) cut surface showing tan brown solid cystic friable tumour mass with areas of haemorrhage and necrosis.

Figure 5 H&E-stained section showing (A) hypercellular tumour areas with cells disposed in sheets; (B) round-to-oval tumour cells with hyperchromatic nuclei, coarse chromatin and scant eosinophilic cytoplasm; (C) sheets of tumour cells with occasional cell showing eccentric nuclei and abundant eosinophilic cytoplasm (rhabdomyoblasts); (D) scattered rhabdomyoblasts on myxoid background.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis based on radiology was primary hepatocellular carcinoma, sarcoma, mesenchymal hamartoma of liver and secondary metastasis to liver. Whole body positron emission tomography scan showed significantly increased uptake of fluorodeoxyglucose in liver only with no focal hypermetabolic lesion identified anywhere else in the body.

TREATMENT

A percutaneous Trucut biopsy was performed and pre-emptive diagnosis of embryonal RMS was made. The patient was planned for open surgery and left hepatic lobectomy was performed (figure 3). Intraoperatively the mass was free from surrounding organs. The gross specimen revealed a well-circumscribed, multilobulated mass with solid cystic spaces (figure 4). Histopathological evaluation demonstrated undifferentiated round to polyhedral and spindle-shaped cells arranged in sheets and clusters with

Figure 6 Immunohistochemical stains showing tumour cells (A) desmin positive; (B) vimentin positive; (C) myogenin positive and (D) negative for pan cytokeratin.

Table 1 Management of embryonal rhabdomyosarcoma in liver in various studies

Authors	Number of cases	Age/sex	Extent of disease	Treatment	Outcome
Horowitz <i>et al</i> ² (1987)	3	1.4 years/M	Right and left lobe of liver	Biopsy followed by chemotherapy Chemotherapy protocol: (a) Cyclophosphamide 400 mg/m ² IV and doxorubicin 25 mg/m ² IV (total dose 300 mg/m ²) alternating with (b) vincristine 1.5 mg/m ² IV and dactinomycin 0.5 mg/m ² . (a) weekly×2 followed by (b) weekly×2. Thereafter, weekly maintenance for a total of 12 months of treatment.	Died at 15 months from the date of diagnosis due to lung and brain involvement
		2.4 years/M	Left lobe with diaphragm rupture	Left lobectomy followed by chemo radiotherapy. Chemotherapy protocol: (a) Cyclophosphamide 400 mg/m ² IV and doxorubicin 25 mg/m ² IV (total dose 300 mg/m ²) alternating with (b) vincristine 1.5 mg/m ² IV and dactinomycin 0.5 mg/m ² (a) weekly×2 followed by (b) weekly×2. Thereafter, weekly maintenance for a total of 18 months of treatment.	Died at 76 months from the date of diagnosis
		7 years/M	Left lobe extending into right lobe	Radiotherapy protocol: 34 Gy in 24 fractions over 33 days Biopsy followed by chemotherapy Chemotherapy protocol: (a) Doxorubicin 60 mg/m ² IV day 1 with DTIC 250 mg/m ² IV daily on days 1–5 (b) Cyclophosphamide 200 mg/m ² IV or PO daily on days 1–5 then vincristine 1.5 mg/m ² IV on day 6 with dactinomycin 1.5 mg/m ² IV on day 6 (a) every 3 weeks×3 followed by (b) every 3 week×3 for a total of 4 months.	Died at 15 months from the date of diagnosis due to abdomen involvement
Tutar <i>et al</i> ³ (2007)	1	6 years/M	8×6 cm cystic mass in the medial portion of the left liver lobe	Surgical resection	NA
Aassab <i>et al</i> ⁴ (2012)	1	25 years/M	136 mm lesion in the right lobe of liver measuring.	Biopsy followed by chemotherapy Chemotherapy protocol: Doxorubicin at 50 mg/m ² j1 (given on day 1 of a 21-day cycle), ifosfamide 2.5 g/m ² j1–3 (given on days 1–3 of a 21-day cycle) and vincristine 1.4 mg/m ² j1, 8 (given on days 1 and 8 of a 21-day cycle). Chemotherapy was repeated every 3 weeks for 3 cycles.	Died after 3 months from completion of chemotherapy due to disease progression.
Haider <i>et al</i> ⁵ (2013)	1	17 years/M	20×13 cm mass arising from left lobe of liver with few satellite lesions in right lobe of liver, largest in segment V measuring 3 cm and small left paravertebral lymph nodes at the level of renal hila	VAC chemotherapy×3 cycles followed by MAID chemotherapy×6 cycles followed by gemcitabine-paclitaxel chemotherapy	Died of rapid progression at 31 months from the date of diagnosis
Rajamahendran <i>et al</i> ⁶ (2014)	1	16 years/M	15×11×9 cm mass lesion involving right lobe of liver involving segments 5–8	Right hepatectomy followed by carboplatin and ifosfamide chemotherapy	Patient expired on 30th postoperative day due to acute respiratory distress
Present case report	1	67 years/M	14.5×12.3×9.1 cm lesion involving left hepatic lobe. Large component of lesion is seen bulging in left sub hepatic space.	Left hepatic lobectomy followed by adjuvant chemotherapy Chemotherapy protocol: 50 mg/m ² doxorubicin (day 1), 2.5 g/m ² ifosfamide (days 1–3), and 1.4 mg/m ² vincristine (days 1 and 8). 3 cycles were given at interval of 3 weeks.	At 24 months of follow-up, patient is free from local recurrence and distant metastasis.

DTIC, Dacarbazine; IV, intravenous; M, male; MAID, mesna, doxorubicin, ifosfamide, dacarbazine; NA, not available; PO, orally; VAC, vincristine, actinomycin D and cyclophosphamide.

Intervening myxoid stroma. Interspersed rhabdomyoblasts were also seen. Stroma showed haemorrhagic areas with foci of necrosis. Resected margins were free of tumour (figure 5). Immunohistochemical analysis revealed vimentin positive, desmin positive, myogenin positive and cytokeratin negative (figure 6). Based on these findings, the diagnosis was established to be embryonal RMS of the liver. Following surgery, adjuvant chemotherapy treatment was given which consisted of 50 mg/m² doxorubicin (day 1), 2.5 g/m² ifosfamide (days 1–3) and 1.4 mg/m² vincristine (days 1 and 8). Three cycles were given at an interval of 3 weeks. It was tolerated well with minimal side effects.

OUTCOME AND FOLLOW-UP

The patient is free from local recurrence and distant metastasis on imaging studies at 24 months of follow-up.

DISCUSSION

RMS is defined as a malignant neoplasm originating from striated muscle. It may arise from undifferentiated mesenchyme which has the ability to differentiate into the muscle. Hence, RMS may occur anywhere in the body.¹ RMS is common in childhood and constitutes around 50% of all soft-tissue sarcomas. It constitutes <1% of all malignancies and 3% of all soft-tissue sarcomas in adults.³ It is most common at ages 2–6 and 15–18 years. Head, neck and genitourinary organs are the most common sites and embryonal RMS is most common subtype of RMS in younger age group.^{2 3} Alveolar is the most common subtype in patients more than 10 years of age and pleomorphic RMS is a disease of the elderly.⁸ Our case, embryonal RMS of the liver is extremely rare both due to the age of the patient at presentation and the location of the tumour. This is probably the first case report of embryonal RMS of liver in 67-year-old man. The previously reported cases of embryonal RMS of liver are summarised in table 1.

Embryonal RMSs in liver has never been reported earlier in elderly patients; hence, there is little information on its management. The standard treatment protocol for children as proposed by the Intergroup Rhabdomyosarcoma Study Group (IRSG) includes gross total resection with negative margins, chemotherapy and radiotherapy. Surgical resection and radiotherapy are used for the management of primary tumour site, while chemotherapy is used to prevent metastasis. Chemotherapy is an essential part of treatment protocol to prevent tumour spread.⁹ Chemotherapeutic drugs include vincristine, actinomycin-D, cyclophosphamide doxorubicin, ifosfamide, etoposide and topotecan.⁹ Our patient received doxorubicin, ifosfamide and vincristine and he responded well without local recurrence and distant metastasis. Although, adults have poor prognosis as compared with children, but when treated aggressively using

paediatric protocol, the prognosis can be similar to children. Poor prognosis is associated with metastatic disease at presentation and poor response to chemotherapy.¹⁰

Learning points

- ▶ Although, embryonal rhabdomyosarcoma is rare in elderly, it should be included in the differential diagnosis for liver mass at any age.
- ▶ These cases pose a great difficulty in treatment because of the absence of standard treatment guidelines.
- ▶ With this case report, we would like to stress that improved survival can be expected only by early detection of the disease, and multidisciplinary approach in managing these patients.

Contributors AA was involved in planning, conduct, reporting, conception and design, acquisition of data and analysis. RJ was involved in conduct, reporting, conception and design, interpretation of data. NA contributed to review of literature and manuscript writing. NH was involved in supervision, review of literature and analysis of data.

Competing interests None declared.

Patient consent Obtained.

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