DOI: 10.7860/JCDR/2020/43257.13610

Case Report

Surgery Section

Splenic Haemangioendothelioma in an Infant

AMIT KUMAR[1], SAMBEDNA[2], RASHI[3], AMIT KUMAR SINHA[4], BINDEY KUMAR[5]

ABSTRACT

Primary Haemangioendothelioma (HE) of spleen is a rare vascular tumour. These tumours have intermediate biologic behaviour between haemangioma and angiosarcoma. So far, few cases have been reported in literature and most of them are in adults or adolescents. Here the present authors report a 6-month-old infant who presented with mass per abdomen. Contrast Enhanced Computerised Tomography (CECT) abdomen revealed a splenic mass which was treated by partial splenectomy. Histopathology confirmed it to be haemangioendothelioma of spleen.

Keywords: Abdominal lump, Partial splenectomy, Splenic mass

CASE REPORT

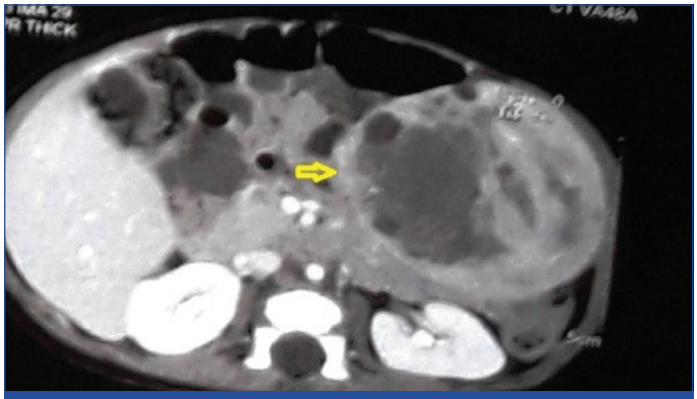
A 6-month-old male infant was presented to the outpatient department of paediatric surgery with complaint of progressively increasing abdominal swelling since one month. The child was apparently normal a month back when his mother noticed an abdominal lump while bathing him. There was no history of pain, fever, any respiratory distress, jaundice or bleeding tendency. His perinatal history was uneventful.

Baby was alert and playful with stable vitals. His weight was 6 kg. On inspection, abdomen was flat, umbilicus was central and all quadrants were moving equally with respiration. On palpation, there was no local rise of temperature and there was no tenderness. There was a solitary lump occupying the left hypochondrium, epigastrium, left lumbar and umbilical regions. The lump was firm in consistency and the margins were smooth. All the borders were well defined except the upper border which merged with the left costal margin. Swelling moved with respiration as well as side to side. Renal angle was free and bilateral hernial orifices were intact. On percussion, liver dullness was felt in right 5th intercostal space. Dull note was heard over the lump and rest of the abdomen is tympanitic. On auscultation, venous hum was heard over the lump.

Patient was admitted and investigated further. On routine blood investigation, Hb was 10.0 gm/dL, total leucocytes count was 14200/thousand/µL, platelet count was 75000 thousand/microL, total serum bilirubin was 0.73 (0.2-1.2 mg/dL), direct serum bilirubin was 0.32 (0.0-0.3 mg/dL). SGOT was 88.40 (05-40 IU/L) and SGPT was 65.42 (0-45 IU/L). Viral markers were negative. Coagulation profile was within normal limit.

Ultrasonography revealed an intraperitoneal hetroechoeic mass lesion measuring 5 cm × 4 cm originating from the liver. Lesion was well marginated with areas of necrosis and calcification. A Contrast Enhanced Computerised Tomography (CECT) was advised for further delineation. CECT abdomen suggested a large heterogeneous soft tissue mass measuring 13 cm × 13 cm × 9 cm in left hypochondriac, left lumbar and epigastric region. Heterogeneous enhancement was seen in post contrast study with few areas of non enhancement likely necrosis. Focus of calcification was also noted within. Mass was seen to be scalloping the upper pole of the spleen and it was displacing and abutting the adjacent bowel loops with maintained fat planes [Table/Fig-1]. The differential diagnoses were Small round cell tumour and Mesenchymal tumour.

A trucut biopsy was planned for histopathological correlation. Histopathological examination of the specimen showed irregular congested vascular channels by plump round to spindled indented



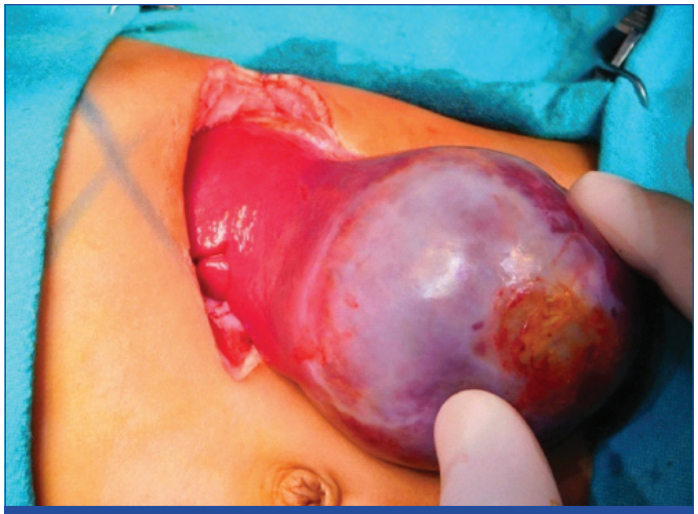
[Table/Fig-1]: CECT showing splenic mass (arrow).

nuclei and moderate amount of cytoplasm some of the cells had vacuolated cytoplasm with presence of intracytoplasmic erythrocytes in few. Areas of haemorrhage were also present. No mitotic activity and necrosis were found. Features were suggestive of vascular tumour and Immunohistochemistry (IHC) was advised to rule out Epithelioid haemangioma and Haemangioendothelioma. On IHC CD31 and CD34 showed strong immunoreactivity in all the vascular channels and some of the endothelial cells. Impression was high grade vascular neoplasm (possibility of haemangioendothelioma and angiosarcoma).

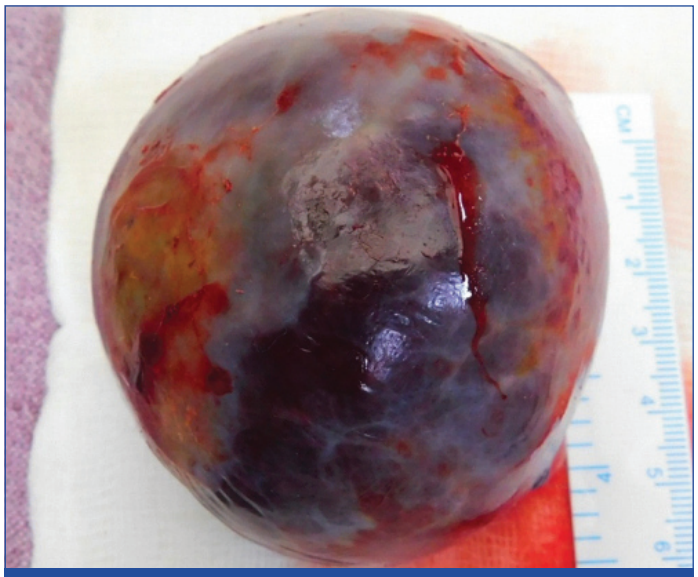
Consultation was done with radiation oncologist but chemotherapy was not possible due to vascular nature of the mass. So the patient was worked up and posted for excision of the tumour and biopsy. On exploration with left sub-costal transverse incision, there was a 6 cm × 6 cm × 5 cm globular mass arising from the lower pole of the spleen [Table/Fig-2,3]. The mass was fleshy and vascular abutting the tail of pancreas. Feeding vessels were ligated and partial splenectomy was done. Specimen was sent for histopathological examination. Histopathology confirmed it to be haemangioendothelioma. Post-operative course was uneventful and patient was discharged on post-operative day seven.

DISCUSSION

The term 'Haemangioendothelioma' includes a heterogeneous group of vascular neoplasms, which are intermediate between benign and malignant tumours [1]. Although haemangioendothelioma is a vascular tumour, it exhibits mild cellular atypia and low mitotic rate; an intermediate appearance between haemangioma and conventional angiosarcoma histopathologically [2]. To the best of knowledge and extensive literature search the present authors could not find splenic haemangioendothelioma in children in two



[Table/Fig-2]: Intraoperative picture of splenic mass.



[Table/Fig-3]: Excised specimen of splenic mass.

reports only, one pediatric patient and another 9-year-old child with splenic haemangioendothelioma that was also treated with partial splenectomy [3,4].

Mallory (1908) defined haemangioendothelioma as all proliferations that originated from endothelial cells of blood vessels [5]. Enzinger and Weiss later categorised haemangioendothelioma into composite epithelioid, hobnail, Kaposiform and epithelioid sarcoma-like haemangioendothelioma [6].

The treatment options for splenic haemangioendothelioma is limited due to very less number of cases reported. In paediatric age group partial splenectomy is preferred in view of their partially developed immunity. In adults, the most successful therapy is considered to be complete splenectomy [7]. Chen CW et al., reported and elderly female of epithelioid haemangioendothelioma of spleen with metastasis. She was treated with oral thalidomide which showed decrease in disease progression [8].

As no specific clinical manifestations are evident in the early stages of development so the diagnosis of splenic HE remains challenging [9]. The common causes of mortality in these cases are infection, consumptive coagulopathy and metastasis. The index case presented with lump abdomen which was diagnosed with computerised tomography and trucut biopsy. Partial splenectomy was done in view of immature immune system of the infant. No infant has been reported in literature so far with this diagnosis. The infant is in close follow-up for last six months and so far doing well.

CONCLUSION(S)

Pediatric haemangioendothelioma is a rarity and diagnosis is a challenge. Due to concerns of potential rupture, thrombocytopenia, anaemia, coagulopathy and intermediate malignant potential, these cases should be managed with splenectomy.

REFERENCES

[1] Nayler SJ, Rubin BP, Calonje E, Chan JK, Fletcher CD. Composite haemangioendothelioma: A complex, low-grade vascular lesion mimicking angiosarcoma. Am J Surg Pathol. 2000;24:352-61.
[2] Fukunaga M, Ushigome S, Nikaido T, Ishikawa E, Nakamori K. Spindle cell haemangioendothelioma: An immunohistochemical and flow cytometric study of six cases. Pathol Int. 1995;45(8):589-95.
[3] Goyal A, Babu SN, Kim V, Kataria S, Rao K. Haemangioendothelioma of liver and spleen: Trauma-induced consumptive coagulopathy. J Pediatr Surg. 2002;37(10):E29.
[4] Wang Z, Zhang L, Zhang B, Mu D, Cui K, Li S. Haemangioendothelioma arising from the spleen: A case report and literature review. Oncology Letters. 2014;9(1):209-12. https://doi.org/10.3892/ol.2014.2693.
[5] Requena L, Kutzner H. Haemangioendothelioma. Semin Diagn Pathol. 2013;30:29-44.
[6] Goldblum JR, Folpe AL, Weiss SW. Haemangioendothelioma: Vascular tumours of intermediate malignancy. In: De Francesco K, editor. Enzinger and Weiss's Soft Tissue Tumours. 6th ed. Philadelphia: Elsevier Saunders. 2014. pp. 681-702.
[7] Yu L, Yang SJ. Kaposiform haemangioendothelioma of the spleen in an adult: An initial case report. Pathol Oncol Res. 2010;17(4):969-72. doi: 10.1007/s12253-010-9331-1.
[8] Chen CW, Hsiao KH, Lu TJ, Lai CW. Retiform and epithelioid haemangioendothelioma arising from the spleen. Formosan Journal of Surgery. 2019;52(4):147-50.
[9] Tjandra JJ, Clunie GJ, Kaye AH, Smith J (eds): Benign and malignant diseases of the hepatobiliary system. Textbook of Surgery. 3rd edition. Wiley-Blackwell, Oxford, pp119, 2006.

PARTICULARS OF CONTRIBUTORS:
1. Senior Resident, Department of Paediatric Surgery, All India Institute of Medical Sciences, Patna, Bihar, India.
2. Senior Resident, Department of Obstetrics and Gynaecology, All India Institute of Medical Sciences, Patna, Bihar, India.
3. Senior Resident, Department of Paediatric Surgery, All India Institute of Medical Sciences, Patna, Bihar, India.
4. Assistant Professor, Department of Paediatric Surgery, All India Institute of Medical Sciences, Patna, Bihar, India.
5. Professor and Head, Department of Paediatric Surgery, All India Institute of Medical Sciences, Patna, Bihar, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Sambedna,
Pitrichhaya, Arpana Bank Colony, Ramjaipal Road, Baily Road, Danapur,
Patna, Bihar, India.
E-mail: sambedna29@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Nov 21, 2019
- Manual Googling: Jan 16, 2020
- iThenticate Software: Mar 02, 2020 (8%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Nov 20, 2019**
Date of Peer Review: **Dec 30, 2019**
Date of Acceptance: **Feb 18, 2020**
Date of Publishing: **Apr 01, 2020**