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Case Report

Mesenteric hemangiolymphangioma: A rare entity with unusual location

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ABSTRACT

Hemangiolymphangioma is a rare venolymphatic vascular malformation characterized by networks or proliferations of vascular spaces—such as lymphatics, capillaries, veins, or arteries—lined by bland endothelium and embedded in connective tissue stroma. The mesentery is a specialized tissue structure in the abdomen that supports the intestines and contains blood vessels, lymphatic vessels, and nerves that are essential for gastrointestinal function. Mesenteric hemangiolymphangiomas are exceedingly rare in this anatomical location, as vascular malformations more commonly occur in other parts of the body, particularly in the skin, subcutaneous tissues, or organs like the liver, lungs, or brain. The rarity of mesenteric hemangiolymphangiomas is attributable to their unusual anatomical location, the coexistence of both hemangiomatous and lymphangiomatous vascular components, the limited number of documented cases in medical literature, the diagnostic difficulties they pose, and their infrequent occurrence in pediatric populations.

This case report details an 8-year-old girl who presented with abdominal distension since past 1.5 years. Imaging modalities, including ultrasonography and CECT of the abdomen, suggested a mesenteric cyst. The patient underwent exploratory laparotomy and excision of the mesenteric cyst. Histopathological analysis of the specimen confirmed the diagnosis of hemangiolymphangioma of the mesentery, further validated by immunohistochemistry. This case highlights the key aspects and histopathological features of this rare condition. Mesenteric hemangiolymphangiomas are intriguing because of their rarity, unique dual vascular origin and diagnostic difficulties. These features make them a valuable subject of study for clinicians and researchers.

1. Introduction

Hemangiolymphangioma is an exceptionally rare lymphatic and vascular malformation characterized by cystic, dilated veins and lymphatic vessels.[1] Extensive involvement of the retroperitoneum and gastrointestinal tract is uncommon. These lesions are primarily congenital, accounting for 40-60% of cases, with an incidence of 1-2 per 10,000 newborns, and are more prevalent in females. They often present clinically in adulthood. Identified risk factors include prematurity, a family history of hemangiolymphangioma, and erythropoietin use.

Hemangiolymphangiomas are predominantly found in the head, neck, and axilla (95%), with only a small percentage (5%) appearing in the mediastinum, retroperitoneum, and mesentery.[2] This article reports a case of hemangiolymphangioma located in the mesentery of an 8-year-old girl, highlighting the pathological features of this case.

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2. Case Report

8 year old girl presented with complaints of abdominal distension since 1.5 years which was insidious in onset, not associated with pain abdomen, puffiness of face, lower limb swelling. Abdominal examination revealed distended abdomen with a soft mass felt in the umbilical region. There was no tenderness and no guarding or rigidity. Laboratory tests did not show any significant abnormalities. CECT abdomen and pelvis were done and showed large mesenteric cyst noted measuring 16 cm x 8.4 cm x 4 cm occupying most of the mid and lower abdomen and part of the pelvis with displacement of the bowel loops as well as the mesenteric vessels.

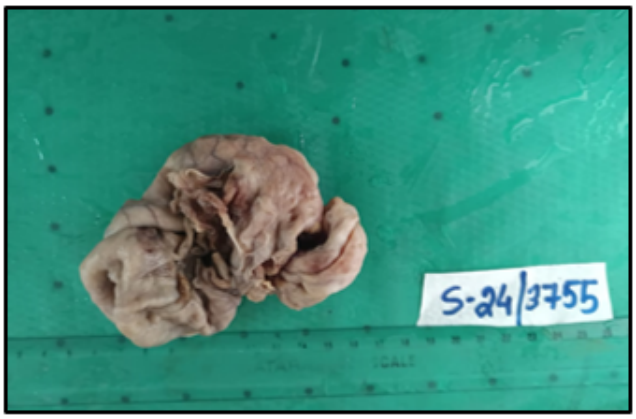

Figure 1: Cystic structure measuring 14x8x2 cm

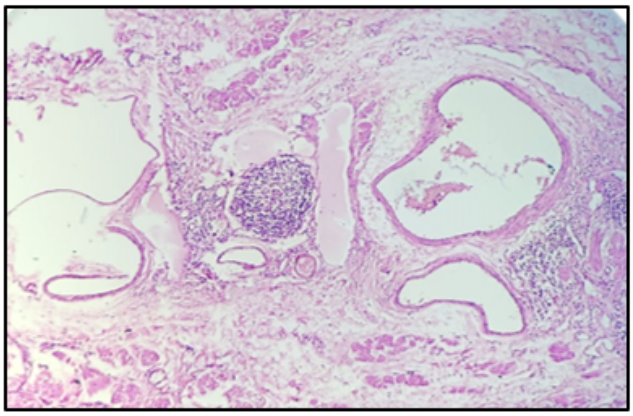

Figure 2: Low power view- Multiple dilated blood vessels and cystic spaces surrounded by lymphocytes. These are surrounded by smooth muscle bundles and fibrous tissue.

Patient underwent exploratory laparotomy with mesenteric cyst excision under GA. Clear fluid of 2 litre was suctioned out during surgical procedure. Post surgical histopathological examination of the specimen was done. Grossly, received a cut open grey white to grey brown cystic structure measuring 14 x 8 x 2 cm. Thin and thick

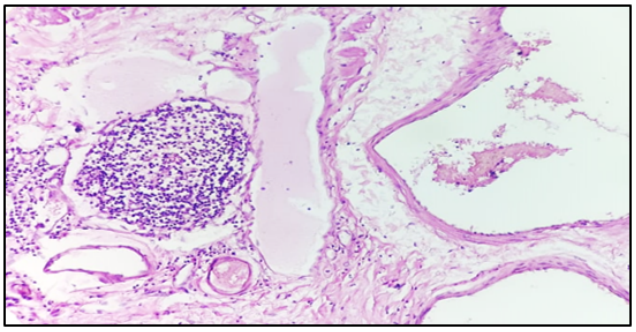

Figure 3: High power view: Thick walled blood vessels filled with RBCs and lymphatic channels.

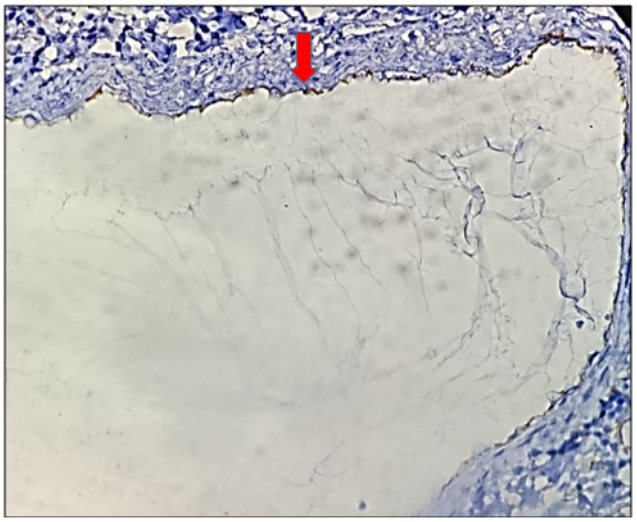

Figure 4: CD 31 positive for vascular endothelial cells. (Red arrow)

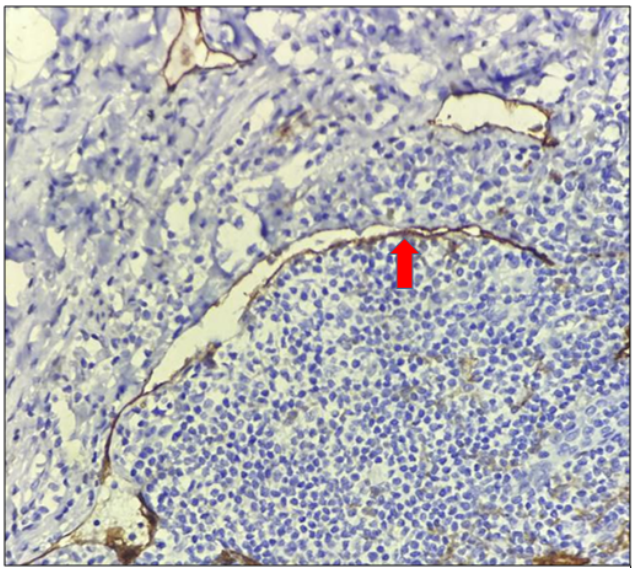

Figure 5: D2-40 positive for lymphatic endothelial cells (red arrow).

walls made out (Figure 1). Histopathological examination from multiple thin and thick cyst wall bits showed multiple dilated thick walled blood vessels which are filled with RBCs and also other cystic spaces which are thin walled lined by single layer of low cuboidal epithelium filled with proteinaceous fluid and surrounded by lymphocytes. These vessels are surrounded by smooth muscle bundles and fibrocollagenous tissue (Figures 2 and 3). Histopathological features were suggestive of Hemangiolymphangioma. On immunohistochemistry, vascular endothelial cells were positive for CD-31 (Figure 4) and lymphatic endothelial cells showed D2-40 positivity (Figure 5).

3. Discussion

Hemangiolymphangioma is a rare benign tumor believed to result from congenital vascular system malformations. It is thought to develop from mesenchymal tissue and contains both venous and lymphatic elements in varying amounts.[3] The tumor's formation may be due to a blockage in the communication between the venolymphatic system and the systemic circulation, which affects the dysembryoplastic vascular tissue.[4]

In the literature, there are various case reports documenting hemangiolymphangiomas occurring in unusual locations such as the spermatic cord, vertebral bodies, oral mucosa, gastrointestinal tract, pancreas, bladder, extremities, paravertebral area, and even the knee. When located in the abdomen, these tumors typically present as space-occupying lesions, causing symptoms such as compression, abdominal pain, or blood in thestool. These symptoms may persist for weeks or months, with the mass growing slowly and progressively. Potential complications include bleeding, perforation, torsion, or rupture of the tumor itself. Differential diagnosis should consider gastrointestinal stromal tumors,pseudomyxoma peritonei, with histopathological analysis being essentialfor accurate diagnosis.[5–7]

The diagnosis begins with a comprehensive clinical history. Due to the rarity of these lesions, preoperative imaging can be difficult; nevertheless, computed tomography (CT) and magnetic resonance imaging (MRI) are useful for evaluating the extent and invasiveness of the lesion, aiding in preoperative surgical planning. Confirmation of the diagnosis is achieved through histopathological examination, which identifies a vascular lesion with both dilated venous and lymphatic vessels. The endothelial cells of these vessels are positive for immunohistochemical markers such as CD31, CD34, and D2-40.[8,9]

The differential diagnosis of mesenteric hemangiolymphangiomas includes several conditions, such as mesenteric cysts, lymphangiomas, mesenteric fibromas, gastrointestinal stromal tumors (GISTs), and vascular malformations. Mesenteric cysts typically present as fluid-filled lesions, whereas hemangiolymphangiomas display a combination of solid and cystic components with distinct vascular structures. Lymphangiomas are characterized by the absence of blood vessels, in contrast to mesenteric hemangiolymphangiomas, which contain both blood and lymphatic vessels. Mesenteric fibromas are solid, fibrous masses, unlike the vascular composition of hemangiolymphangiomas. GISTs appear as solid and homogeneous on imaging, while hemangiolymphangiomas show heterogeneous enhancement. Histopathological analysis demonstrating both blood and lymphatic vessels is essential for distinguishing mesenteric hemangiolymphangiomas from these other conditions.[9]

The main approach to treating these lesions is total surgical resection, which typically yields low recurrence rates. There are limited documented cases of mesenteric hemangiolymphangioma with widespread involvement of the gastrointestinal tract. It is advisable to consider hemangiolymphangioma as a potential diagnosis in patients presenting with compressive abdominal symptoms, especially after excluding other benign and malignant neoplasms.[10,11]

4. Conclusion

Hemangiolymphangioma of the mesentery is a rare vascular malformation characterized by an abnormal proliferation of blood vessels and lymphatics. Understanding the pathophysiology of hemangiolymphangioma is crucial for effective management, as this condition can present similarly to other mesenteric masses.[1,2] Ultimately, a multidisciplinary approach, involving surgeons, radiologists, and pathologists, is essential for addressing this complex vascular anomaly effectively, ensuring timely intervention and reducing the risk of severe complications. Regular follow-up is also important to monitor for recurrence or new symptoms, fostering improved quality of life for affected individuals.[3,4]

5. Sources of Funding

None

6. Conflict of Interest

None.

References

1. Li X, Zhang Y, Sun H, Shao Q, Zhang S, Li F, et al. Hemolymphangioma of the transverse mesocolon: a case report and literature review. *Transl Cancer Res*. 2021;10(8):3849–55.
2. Caviedes MA, Giraldo LC, Pena LA. Mesenteric hemangiolymphangioma: case report. *Medicina Laboratorio*. 2023;27(1):33–8.
3. Pandey S, Fan M, Chang D, Zhu J, Zhu Y, Li Z, et al. Hemolymphangioma of Greater Omentum: A Rare Case Report. *Medicine (Baltimore)*. 2016;95(17):e3508. doi:10.1097/MD.0000000000003508.

4. Li Y, Pang X, Yang H, Gao C, Peng B. Hemolymphangioma of the waist: A case report and review of the literature. *Oncol Lett*. 2015;9(6):2629–32.
5. Rogel-Rodriguez JF, Gil-Garcia JF, Velasco-Garcia P, Romero-Espinoza F, Zaragoza-Salas T, Munoz-Lumbreras G, et al. Hemangiolymphangioma of the spermatic cord in a 17 year-old: A case report. *Cir*. 2016;84(2):164–8.
6. Zhang Y, Li B, Shi H, Cai L, Hou J. Vertebral hemangiolymphangioma mimics bone metastases on 99mTc-MDP SPECT/CT. *Clin Nucl Med*. 2016;41(1):76–8.
7. Manickam S, Sasikumar P, Kishore BN, Joys S. Hemangiolymphangioma of buccal mucosa: A rare case report. *J Oral Maxillofac Pathol*. 2017;21(2):282–5.
8. Murphy T, Ramai D, Lai J, Sullivan K, Grimes C. Adult neck hemangiolymphangioma: a case and review of its etiology, diagnosis and management. *J Surg Case Rep*. 2017;8:rjx168. doi:10.1093/jscr/rjx168.
9. Smith J, Jones A, Wang L. Differential diagnosis of mesenteric hemangiolymphangiomas: a review. *Abdom Imaging*. 2023;48:123–30.
10. Antonino A, Gragnano E, Sangiuliano N, Rosato A, Maglio M, Palma MD, et al. A very rare case of duodenal hemolymphangioma presenting with iron deficiency anemia. *Int J Surg Case Rep*. 2014;5(3):118–21.
11. Kosmidis I, Vlachou M, Koutroufinis A, Filiopoulos K. Hemolymphangioma of the lower extremities in children: two case reports. *J Orthop Surg Res*. 2010;5:56. doi:10.1186/1749-799X-5-56.

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