Print ISSN: 2321-6379
Online ISSN: 2321-595X

Case Report

Curious Case of Glittering Mesenteric Mass

Pujitha Gadde[1], Rajoo Ramachandran[2], J D Roy Santhosham[3], G Kiran[4]

[1]Postgraduate Resident, Department of Radiology and Imaging Sciences, Sri Ramachandra Medical College and Research Institute, Chennai, Tamil Nadu, India, [2]Professor, Department of Radiology and Imaging Sciences, Sri Ramachandra Medical College and Research Institute, Chennai, Tamil Nadu, India, [3]Professor, Department of Radiology and Imaging Sciences, Sri Ramachandra Medical College and Research Institute, Chennai, Tamil Nadu, India, [4]Assistant Professor, Department of Radiology and Imaging Sciences, Sri Ramachandra Medical College and Research Institute, Chennai, Tamil Nadu, India

Abstract

The mesentery has a double peritoneal layer that attaches the bowel to the abdominal wall. Computed tomography plays an essential role in the diagnosis of mesenteric disease, which can present as a solid mass, cystic mass, and local or diffuse infiltration depending on the disease pattern. Few mass lesions involving the mesentery present as a hypervascular mass which include carcinoid, malignant fibrous histiocytoma, angiosarcoma, plasmacytoma, and hemangiopericytoma. In this case report, we present an unusual hypervascular mass lesion of the mesentery on imaging which was diagnosed as a pleomorphic hyalinizing angiectatic tumor (PHAT) based on histopathological findings. To the best of our knowledge, there was no report identifiable as PHAT involving the small bowel mesentery in English literature.

Key words: Hemangiopericytoma, Mesentery, Pleomorphic hyalinizing angiectatic tumor, Small bowel

INTRODUCTION

Pleomorphic hyalinizing angiectatic tumor (PHAT) is a rare, locally aggressive mesenchymal tumor with low malignant potential and a high propensity for local recurrence.[1] The tumor has a predilection for the subcutaneous tissues of the lower extremities in adults and affects males and females with a similar proportion. Originally described by Smith *et al.* who stated that the tumor contains sheets of large, pleomorphic spindle cells with prominent vessels in a hyalinized stroma,[3] reminiscent of a schwannoma even if they are not nerve-related. This is the first report in literature with the rare occurrence of this tumor in small bowel mesentery. In this case report, we describe the imaging characteristics of the PHAT in small bowel mesentery which was proven on histopathology and immunohistochemical analysis.

Access this article online

www.ijss-sn.com

Month of Submission : 09-2024
Month of Peer Review : 10-2024
Month of Acceptance : 11-2024
Month of Publishing : 11-2024

CASE REPORT

A 60-year-old female of Indian origin presented to the outpatient department clinic with chief complaints of low-grade intermittent fever and constipation. No history of abdominal pain, reduced appetite, weight loss, or melena. Per abdominal examination revealed a mobile, well-defined mass in the right iliac fossa, with a clearly defined lower border. The vitals were normal. Based on the clinical examination; the patient was advised for routine biochemical blood workup and chest X-ray (in view of fever to rule out underlying infection). The chest X-ray and the blood workup were normal. Vanillylmandelic acid levels were found to be normal. The patient was not willing for an ultrasound (USG) and directly requested for computed tomography (CT) scan as she was anxious. Contrast-enhanced CT (CECT) showed a hypervascular mass lesion in the mesentery in the midline in the pelvis with a superior mesenteric artery supplying the lesion and a superior mesenteric vein draining it [Figures 1 and 2]. Based on the imaging findings, the differentials were hemangiopericytoma of small bowel mesentery or mesenteric neuroendocrine tumor. The patient was advised for excision biopsy and histopathology correlation for further characterization of the lesion considering the hypervascular nature. Laparotomy and excision was done uneventfully and sent for histopathological analysis. Gross

Corresponding Author: Dr.Pujitha Gadde, D.No: 54-19-21A/1, 3rd floor, Srinivasam Apartment, Jayaprakash Nagar Road No:2, Vijayawada, Andhrapradesh - 520008, India.

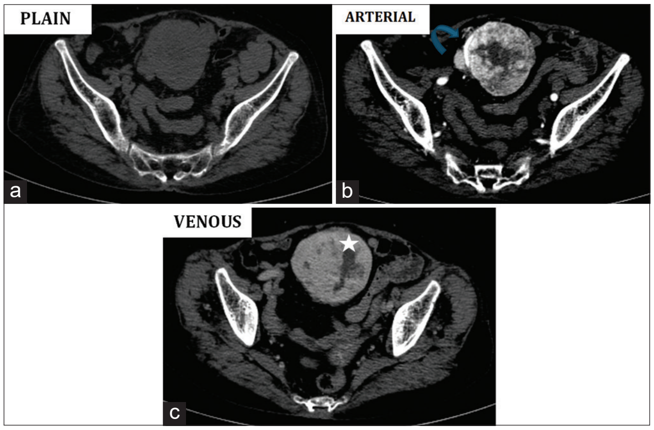

Figure 1: (a-c) Axial sections of non-contrast (a) and post-contrast (b-c) computed tomography abdomen shows a well-defined iso-dense mesenteric lesion in the pelvis with intense heterogeneous enhancement on arterial phase and progressive centripetal filling on venous phase with few necrotic foci within (star). The distal segment of the dilated superior mesenteric artery is seen on the right lateral aspect of the mesenteric lesion (curved arrow)

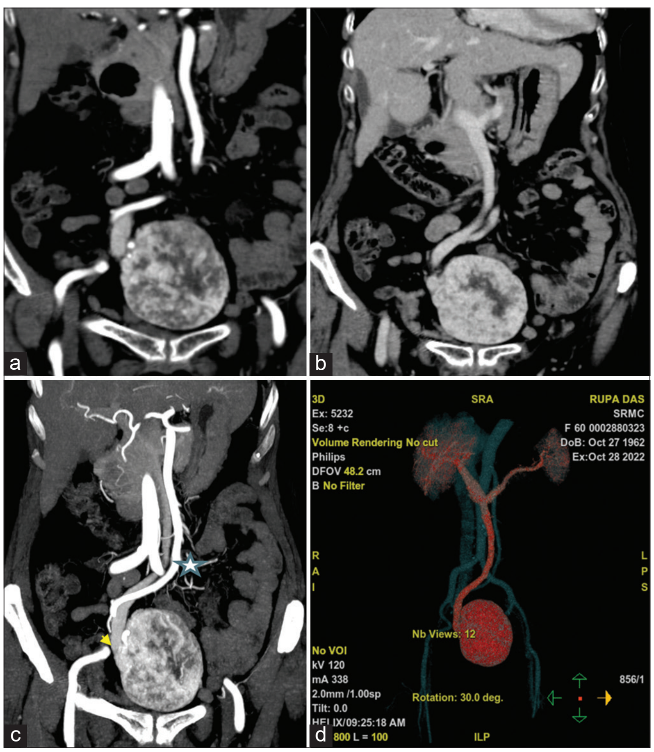

Figure 2: (a and b) Reformatted coronal view of computed tomography abdomen in arterial and venous phase shows heterogeneously enhancing lesion in the pelvis which is seen to receive arterial supply from the superior mesenteric artery (SMA) (star) and venous drainage by the dilated superior mesenteric vein (SMV) (arrow). (c and d) Coronal maximum intensity projection and VR 3D image show the dilated SMA and SMV coursing parallel to the aorta (to pelvis) and terminating into the lesion

specimen showed gray–white globular soft - tissue mass uneventfully and sent for histopathological analysis. Gross specimen showed gray–white globular soft - tissue mass with focal irregularity, which on the cut surface showed focal gray brown areas and extruded hemorrhagic fluid [Figure 3a]. The histopathological sections showed lymphocytes, mast cells, oval spindle cells, admixed with hyalinized material, and dilated blood vessels with hyalinization within [Figure 3b]. On immunohistochemistry it showed positivity for S100 [Figure 4a], vimentin [Figure 4b], and CD34 [Figure 4c]. The final diagnosis based on histopathological analysis was PHAT.

DISCUSSION

PHATs are rare soft-tissue tumors of low malignant potential that usually involve the extremities of adults. It was first described in 1996 by Smith *et al.* in a series of 14 cases as a "low-grade sarcoma of uncertain lineage."[2] In "Soft Tissue and Bone Tumors" by the World Health Organization 2020; PHAT is considered a "neoplasm of uncertain behavior of connective or other soft tissue."[3] In a review study conducted by Folpe and Weiss, about 41 cases of PHAT were analyzed which had a median age of 53 years, and the majority of cases (76%), had lower extremity involvement by the tumor.[4] The rest of the cases showed involvement of upper extremities, axilla, perineum, buttock, and back and he concluded none of his patients with PHAT developed metastases.

Here we report a case of PHAT in the small bowel mesentery with imaging differentials as hemangiopericytoma or mesenteric neuroendocrine tumor due to its enhancement characteristics as these tumors. Excisional biopsy and histopathological correlation was confirmatory which showed PHAT involving the small bowel mesentery. This is an extremely rare location for PHAT, to our knowledge and research, this was the first case in literature of the same.

Histopathologically, while the pleomorphic tumor cells of PHAT resemble those of malignant fibrous histiocytoma (pleomorphic undifferentiated sarcoma), mitotic features and mitotic figures are rare. Microscopically, it shows abundant clusters of ectatic and variably thrombotic blood vessels with damaged walls (hyalinization or fibrinous deposits), sheets, and fascicles of a varying proportion of spindle and pleomorphic cells. Spindle cells show slender to plump cytoplasm and ovoid to wavy nuclei with low pleomorphism. Pleomorphic cells show abundant cytoplasm and large, irregular, vesicular nuclei with prominent eosinophilic nucleoli (resembling virocyte-like cells). Intranuclear pseudoinclusions and hyperchromasia can be present mitotic features and mitotic activity is very low ($<1/mm^2$) and necrosis is rare in PHAT.[5]

On immunohistochemistry stains, PHAT of soft parts usually express CD34 and vimentin and are negative for

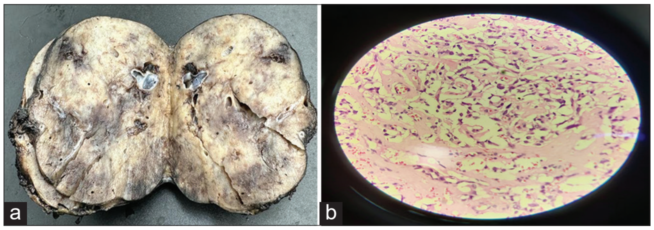

Figure 3: (a) Gross specimen shows gray–white globular soft-tissue mass with focal irregularity, which on the cut surface showed focal gray–brown areas and extruded hemorrhagic fluid. (b) In histopathology, it is composed of oval to spindle cells, admixed with hyalinized material, many dilated blood vessels are also seen with prominent hyalinization around them. The spindled-out cells show moderate cytoplasm with focal clearing and moderate pleomorphism with vesicular nuclei.

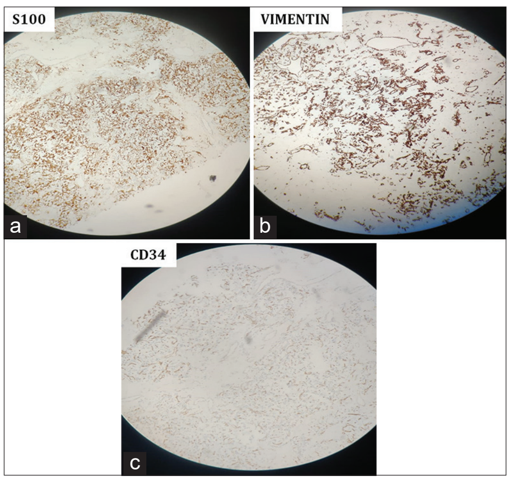

Figure 4: (a-c) Immunohistochemistry showed positivity for S100, Vimentin and CD34.

S-100 protein, actin, desmin, cytokeratin, CD31, factor VIII, and epithelial membrane antigen.[3] In contrast, our case showed positivity for S-100, and more research is required to evaluate this unique pathological finding in the case of PHAT of small bowel mesentery.

Imaging features of the related entity, soft tissue PHAT are limited in literature, all of which share the common features of a subcutaneous lipomatous mass in the extremities.[5,6] These features are non-specific; however, on USG it appears as a heterogeneously hypoechoic mass with a globular intralesional hyperechoic foci and shows heterogeneous enhancement and possible feeding artery on CECT. Soft-tissue PHAT on magnetic resonance imaging is heterogeneously isointense to muscle with multiple hypointense and hyperintense foci on the T1-weighted image, and on the T2-weighted image it appears heterogeneously hyperintense with flow voids. Although there will be a local recurrence, the main treatment of PHATs consists of wide surgical excision.[7] Up to 50% of the resected lesions show local recurrence which can be usually cured by re-excision. In extremely rare situations, PHATs of soft parts have been reported to progress to myxoid sarcoma but the exact relationship is unclear.[7] The imaging findings of mesenteric PHAT have not been reported in the radiology literature. Our case showed a hyperenhancing mass in the small bowel mesentery with a feeding artery and a draining vein with no locoregional and distant metastasis.

CONCLUSION

Pleomorphic hyalinizing angiectatic tumor of the mesentery is extremely rare. Positivity for S-100 on immunochemistry may be one of the diagnostic features; however, more research is required for the same. It should be included in the differential diagnosis of a well-defined, hypervascular soft-tissue mass in the pelvic cavity. This newly recognized, rare neoplasm has imaging features corresponding to its pathological nature and hence a thorough knowledge of this uncommon tumor is essential for clinical practice which alters the course of management.

STATEMENT

The manuscript has been read and approved by all the authors, the requirements for authorship in this document have been met, and each author believes that the manuscript represents honest work and information is not provided in another form.

ACKNOWLEDGMENT

I would like to express my sincere gratitude to all those who contributed to the successful completion of this case report.

First and foremost, I thank Dr. Rajoo Ramachandran, Professor, for his invaluable guidance and support throughout the process. His expertise and insight were instrumental in the development and refinement of this case report.

I am also grateful to my institute, SRIHER, for providing the necessary resources and facilities that enabled us to conduct this study. A special thanks to all of my colleagues and technicians for their assistance.

My heartfelt appreciation goes to the patient for her cooperation and willingness to share her case, which was essential to the completion of this report.

Any errors or omissions are the responsibility of the authors.

REFERENCES

1. Subhawong TK, Subhawong AP, Montgomery EA, Fayad LM. Pleomorphic hyalinizing angiectatic tumor: Imaging findings. Skeletal Radiol 2012;41:1621-6.
2. Smith ME, Fisher C, Weiss SW. Pleomorphic hyalinizing angiectatic tumor of soft parts. A low-grade neoplasm resembling neurilemoma. Am J Surg Pathol 1996;20:21-9.
3. World Health Organization. Soft Tissue and Bone Tumours. WHO Classification of Tumours. 5th ed., Vol. 3. Geneva, Switzerland: World Health Organization; 2020.
4. Folpe AL, Weiss SW. Pleomorphic hyalinizing angiectatic tumor: Analysis of 41 cases supporting evolution from a distinctive precursor lesion. Am J Surg Pathol 2004;28:1417-25.
5. Cazzato G, Colagrande A, Cimmino A, Lettini T, Savino MT, Martella C, *et al.* Pleomorphic Hyalinizing Angiectatic Tumor (PHAT): Review of the literature with case presentation. Dermatopathology (Basel) 2021;8:97-102.
6. Moretti VM, de la Cruz M, Brooks JS, Lackman RD. Early pleomorphic hyalinizing angiectatic tumor: Precursor or distinct lesion? Orthopedics 2010;33:516.
7. Subhawong TK, Subhawong AP, Montgomery EA, Fayad LM. Pleomorphic hyalinizing angiectatic tumor: Imaging findings. Skeletal Radiol 2012;41:1621-6.

How to cite this article: Gadde P, Ramachandran R, Santhosham JDR, Kiran G. Curious Case of Glittering Mesenteric Mass. Int J Sci Stud 2024;12(8):9-12.

Source of Support: Nil, **Conflicts of Interest:** None declared.