

Mesenteric Lymph Node Hamartoma (Castleman's Disease) in Association with Superior Mesenteric Arteriovenous Fistula

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ABSTRACT

We present a case of 21-year-old female patient with history of pain abdomen and abdominal distension. The patient also had oedema of the limbs, puffiness of the face, pallor and palpable mass in the abdomen. Ultrasonography of the abdomen and computed tomographic angiogram was done and it showed presence of vascular mass along with arteriovenous malformation in the mesentery of small gut between distal branches of superior mesenteric artery and vein. Surgical excision of the mass with ligation and division of the arteriovenous malformation was done through midline laparotomy. Histopathological examination was consistent with the diagnosis of Castleman's disease. The Patient recovered well and was discharged after seven days.

Keywords: Castleman's disease, Mesenteric arteriovenous fistula, Portal hypertension

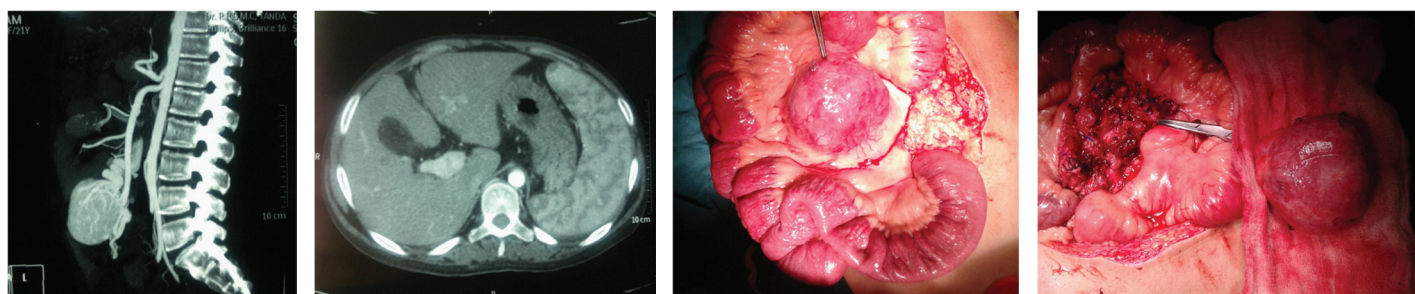
CASE REPORT

A 21-year-old female patient presented with history of pain and abdominal distension for one month without any evidence of axillary, inguinal or neck lymphadenopathy. There was no history of trauma or previous surgery and patient did not have similar symptoms in the past. Patient also had oedema of the limbs and puffiness of the face along with pallor. Positive findings on clinical examination were tachycardia, anemia and palpable mass in the abdomen. Thyroid function tests, cortisol level and adrenocorticotropin levels were within the normal range. Ultrasonography of the abdomen revealed presence of vascular mass with possibility of aneurysm along with features of portal hypertension (splenomegaly + dilated portal vein). Computed Tomographic (CT) Angiogram was done and it showed the presence of a large 7.5 cm x 7 cm vascular tumour in the mesentery of small intestine which was enhancing intensely after contrast administration, moreover the branches of superior mesenteric vein were opacified in the arterial phase of the angiogram showing the presence of an associated arteriovenous malformation (AVM) in the mesentery of small gut between distal branches of superior mesenteric artery and vein [Table/Fig-1a]. CT scan of the abdomen also showed splenomegaly and dilated portal vein as a consequence of portal hypertension due to the AVM [Table/Fig-1b]. Chest radiograph and CT chest were normal and did not reveal any evidence of hilar lymphadenopathy or mediastinal mass and echocardiogram was also normal. Surgical excision of the vascular mass and arteriovenous malformation was planned with or without resection of a part of the small gut. Patient was operated under general anesthesia and abdomen was explored through midline laparotomy. Tumour and AVM was localized in the mesentery of small

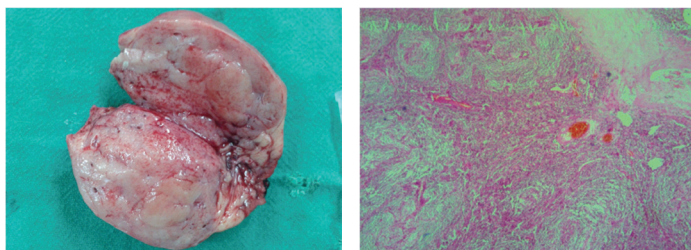
gut [Table/Fig-2a], mesentery was opened on its's anterior aspect and dissection was done close to the tumour to preserve as many branches of the superior mesenteric artery as possible. The tumour was very vascular and the draining branches of superior mesenteric vein were considerably dilated and tortuous and were gently dissected and ligated. The tumour was excised [Table/Fig-2b] and associated arterio venous fistula was closed by ligation and division of the feeding vessels and draining veins, small gut vascularity was preserved and did not require resection and anastomosis. Raw surface in the mesentery was covered with omentum after hemostasis and small gut was observed for any ischemic changes. There were palpable pulsations in the mesentery after excision AVM and there were no ischemic changes in the gut. Excised tumour [Table/Fig-3a] was sent for histopathology examination and abdomen was closed after inserting a drain. Histopathologic examination [Table/Fig-3b] of the mass revealed the presence of a lymph node hamartoma/ Castleman's disease (hyaline vascular type). Patient was given blood transfusion intra operatively and postoperatively, recovery was uneventful and patient was discharged after seven days.

DISCUSSION

Lymph node hamartoma also known as giant lymph node hyperplasia or Castleman's disease is a rare disorder [1]. There are two major pathologic variants of the disease 1) hyaline vascular type and 2) plasma cell type. The disease can involve a single node or can be multicentric [2]. Mediastinum is site most commonly involved; other sites reported in literature are neck, axilla, mesentery, retroperitoneum and pelvis. Resection of the tumour is recommended in case of unicentric disease and is associated with favorable outcome. This case had superior mesenteric arteriovenous fistula (AVF)



[Table/Fig-1a]: Pre-operative CT angiogram in oblique view showing superior mesenteric AVF along with mass in the mesentery of small gut **[Table/Fig-1b]:** Pre-operative CT abdomen showing splenomegaly and dilated portal vein in the axial section as a consequence of portal hypertension **[Table/Fig-2a]:** Intra-operative photograph showing large mass with AVF arising from superior mesenteric vessels **[Table/Fig-2b]:** Intra-operative photograph after excision of the mass and ligation and division of AVF



[Table/Fig-3a]: Gross appearance of the mass in cut section

[Table/Fig-3b]: Histopathological appearance of the resected mass

in association with lymph node hamartoma and is reported because of its rarity and to highlight the favorable outcome with surgical treatment. Mesentery is an uncommon site for lymph node hamartoma. Surgical excision is curative for patients with unicentric disease [3] that is involving a single lymph node as was seen in this case, whereas patients with multicentric disease are managed with chemotherapy and steroids, radiotherapy has also been used with some success. Prognosis for multicentric disease is not as good as seen with unicentric disease [4].

Castleman's disease is a lymphoproliferative disorder, the disease is more common in young adults and males and females are equally affected. Differential diagnosis of Castleman's disease includes other more common diseases. 1) inflammatory diseases like tuberculosis. 2) benign tumours like fibromas, leiomyomas. 3) malignant tumours like Hodgkin's disease, non Hodgkin's lymphoma, various sarcomas and carcinoma's. The lesion is hypervascular and usual CT finding in the unicentric type is homogenous contrast enhancement. It is very difficult to pre-operatively diagnose this condition and in majority of the patients the diagnosis is confirmed only on histopathological examination. There are few reports in literature on mesenteric Castleman's disease [5,6].

Superior mesenteric AVF is extremely rare in itself with few reports available in literature [7]. Causes of AVF can be traumatic [8], iatrogenic, infectious and congenital. AVF has also been reported many years after gut resection. Without treatment the size of AVF continues to increase and can lead to complications, therefore, early intervention is recommended for this pathology. Complications associated with superior mesenteric AVF are gastrointestinal bleeding, ischemia distal to site of AVF, compression on surrounding structures, splenomegaly, portal hypertension and its associated complications and cardiac failure in cases of large AVF. Characteristic features of AVF at different sites are portal hypertension in gastrointestinal tract [9], right to left shunt and cyanosis in cases of pulmonary AVF (AVF at other non pulmonary sites leads to left to right shunt without

cyanosis) and hypertrophy of the limbs in cases of AVF involving the limbs. Important investigations in the diagnosis of mesenteric AVF are angiography, endoscopy and intraoperative fluoroscopy [10]. Intraoperative fluoroscopy can be very useful during open surgery in preventing excessive bowel resection. Intervention is required for AVF to prevent complications and it can be done through surgical excision by either open or minimally invasive techniques (Laparoscopy and endoscopy) or by endovascular techniques using coil embolization [11,12].

CONCLUSION

Castleman's disease can be associated with AVF in the gastrointestinal tract. Surgical management of unicentric Castleman's disease and associated mesenteric AVF is associated with favorable outcome and early management is required before complications due to portal hypertension or heart failure supervene.

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