ABDOMINAL MESENTERIC CYSTIC LYMPHANGIOMA, A CASE REPORT WITH REVISION OF LITERATURES.

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ABSTRACT

Abdominal mesenteric cystic lymphangioma is a rare congenital benign tumor, that can be misdiagnosed with others cystic intra-abdominal tumors. The etiology is thought to be related with the congenital malformation of lymphatics.

We presented a case of a 4 year old male child with a cystic lymphangioma, arising from lymphatics of small bowel in the mesentery and occupying nearly the entire abdominal cavity. Focal bulging of anterior abdominal wall was observed. Plain film and computed tomography confirmed the intrabdominal septated cystic tumor. Radical surgical treatment was done, according to international guide-lines. The cystic tumor, attached mesentery and 45 cm. in length of the engulfed small intestine were completely excised and then primary end to end anastomosis was done. Histological diagnosis was chylous cystic mesenteric lymphangioma. The patient had an uneventful post operative course and had no evidence of recent post operative complication. No early relapse was recorded.

INTRODUCTION

Lymphangiomas are benign congenital lymphatic malformations with a proliferation of dilated lymphatic spaces lined by thin attenuated endothelial cells. They are frequently found in children, and comprised 5.6% of all benign lesions of infancy and childhood.1 The acquired form may be detected in middle aged adults. They have no predilection for either sex or any race.[1]

Lymphangioma has been classified into three groups: (1) lymphangioma simplex; (2) cavernous lymphangioma; and (3) cystic lymphangioma.

Cystic lymphangioma is an uncommon congenital benign neoplasm, which can occur throughout the body, but are most common in the neck of children and are called cystic hygroma. They usually appears in the axillary region, and rarely occur in the mediastinum. The remainders are found in the omentum, mesentery, retroperitoneum and bone.

Intraabdominal lymphangiomas are rare. Most of them are known to be cystic lymphangioma in pathology. The majorities are presented in patients younger than 5 years of age, but occasionally these tumors do not produce symptoms until adult life. Within the abdomen, the most common site for lymphangioma is the bowel mesentery, following by omentum, mesocolon and retroperitoneum.

The clinical symptoms of mesenteric lymphangioma seem to be related with the size and site of tumor, including an abdominal mass or an acute abdomen. Usually, the cases are asymptomatic but may present with acute or chronic intestinal obstruction.

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Most common complications are volvulus, intestinal obstruction, infarction, perforation, and hemorrhage.

The diagnosis of abdominal mesenteric lymphangioma can be made by multiple radiologic studies, including plain radiograph, ultrasonography, MRI and arteriography.

The aim of our study was to show the rarely huge chylous type, mesenteric cystic lymphangioma, which found in children who had presenting symptom as abdominal pain, increasing abdominal girth, abdominal distension and chronic intermittent intestinal obstruction.

CASE REPORT

On February 02, 2007, a 4 year old boy first presented to our hospital with chronic abdominal fullness, vague abdominal pain and nonbilious vomiting. He had been healthy until then with an unremarkable medical and surgical history. The initial clinical diagnosis at the first presentation was acute gastritis.

Next over 4 months later, on June 26, 2007, he came back to our emergency services again with a history of nonbilious vomiting of increasing frequency, increase abdominal girth, progressive abdominal distension and acute abdominal pain. On examination, the patient was in stable condition, but a small right abdominal movable mass was palpated with firm consistency. It was painful on palpation. He had mild tenderness in the right lower abdominal region. There was no hepatomegaly, splenomegaly or clinical lymphadenopathy. Auscultation of the abdomen revealed normal bowel sounds. Laboratory data were within normal limits.

Preoperative radiographic studies were performed, including plain radiograph and computed tomography (CT). The plain abdominal radiograph (Figure 1) showed a large radiopaque right abdominal soft tissue mass with mass effect to the nearby structures and displacement of bowel loops.

An abdominal CT (Figure 2 A-D) confirmed a large thin walled, bilobed cystic mass and few internal septations which producing multiloculated cysts. It occupied nearly the entire abdomen, right side predominance. The mass extended from the level of left subhepatic region to the pelvic cavity, just above the bladder. The mass caused pressure effect to the intraabdominal structures, strikinly compressed and stretched the adjacent bowel loops. There is no complicated hemorrhage, volvulus or leakage.

Notably, some dilated gastric cavity caused mass effect to the gastric outlet, seen in both plain radiograph and CT. In conjunction with clinical nonbilious vomiting, so partial gastric outlet obstruction should be considered.

Due to progressive worsening and increasing frequency of vomiting, the child was underwent standard preoperative preparation for a laparotomy. This included inserting a nasogastric tube, retained Foley's catheter, initiating intravenous fluid therapy, and starting prophylactic antibiotics. Then patient was taken to the operating room and an urgency surgical treatment was performed, 2 days after admission on June 28, 2007.

The provisional diagnosis was intrabdominal cystic mass with partial gut obstruction. Cystic teratoma, lymphangioma, omental / mesenteric cyst, duplication cyst, pseudocyst, loculated ascites were listed in the differential diagnosis.

At operation, midline vertical incision was done. After opening the peritoneum, the cyst was seen popping out of the abdominal incision (Figure 3). A giant gray-yellowish spongy mutiloculated cystic tumor and relative soft consistency within the root of small bowel mesentery was exposed. It occupied the majority of abdominal cavity. In order to mobilize the tumor, it was necessary to extend the incision cavity

in both directions. It was then noticed that the tumor was apparently involved the adjacent small bowel and infiltrated its mesentery. The rest of bowel and other abdominal structures appeared normal; there was no any dilatation, ischemia or inflammatory change of the intestine and mesentery.

Due to part of small intestine plastered the lateral border of the mass, in a way that the mass could not be enucleated. Extensive resection of the mass, including the mesentery and the part of the engulf small intestine was done. Therefore about 22-23 cm. long segment of gut on either sides of the tumor were radically resected with the mass and then a primary end to end anastomosis was done. With careful dissection of the mesenteric arteries, the tumor was removed completely with accompanying adjacent small bowel and mesenteric resection. No ascites was seen in the peritoneal cavity.

Post operative details, he was maintained NPO with intravenous fluids and nasogastric suction until bowel function returns. Prophylactic antibiotics can be discontinued after 7 postoperative days. He had an uneventful post operative course, and had no evidence of recent post operative complication.

The patient was discharged at the 9th postoperative day. He was followed up 2-3 weeks after being discharged from the hospital and found out to have no problem. At the present time, 2 months after operation the patient is healthy, his condition is normal.

The mass with the attached small intestine was sent to the pathology department for evaluation.

The gross specimen consisted of a large lobulated gray-yellowish multicystic mass in the small bowel mesentery with soft consistency, measured approximately 20 x 20 x 17 cm., contiguous long segment of small intestine plastered to its surfaces about 22-23 cm. of each sided (Figure 4). Serial sections show numerous small and large cysts which were filled with turbid and milky odorless fluid soluble (chylous cysts).

Microscopic description shows numerous dilated lymphatic channels with varying in sizes within loose fibroconnective tissue and a few disorganized bundles of smooth muscle presented in the wall of the larger channels, these findings were consistent with cystic lymphangioma.

Finally, the diagnosis was mesenteric chylous cyst, histologically cystic mesenteric lymphangioma.

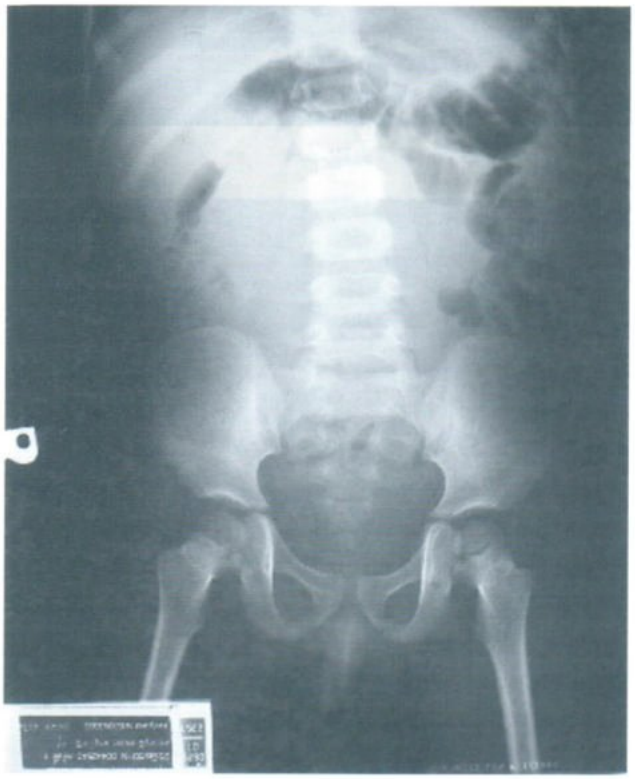

Fig.1 Supine radiographs of the abdomen show a soft-tissue mass within the right hemiabdomen that displacement of bowel loops, upward and laterally. Some dilated stomach and transverse colon due to mass effect.

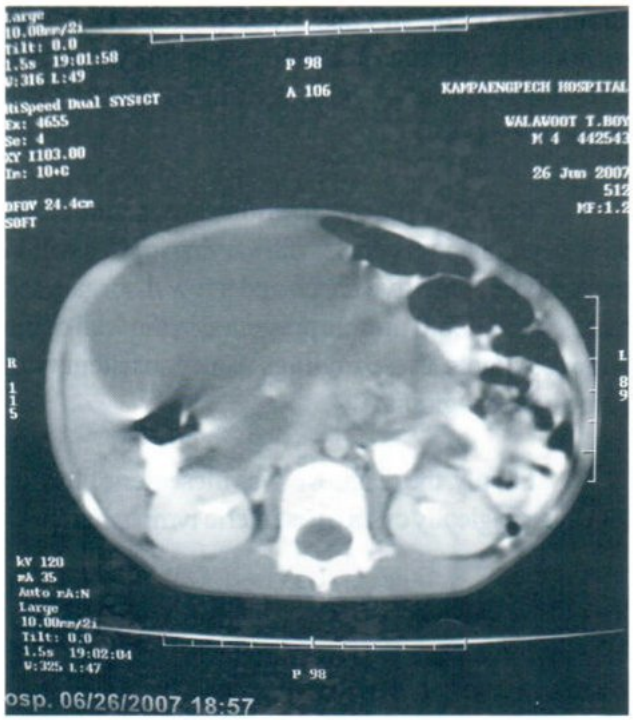

Fig.2A

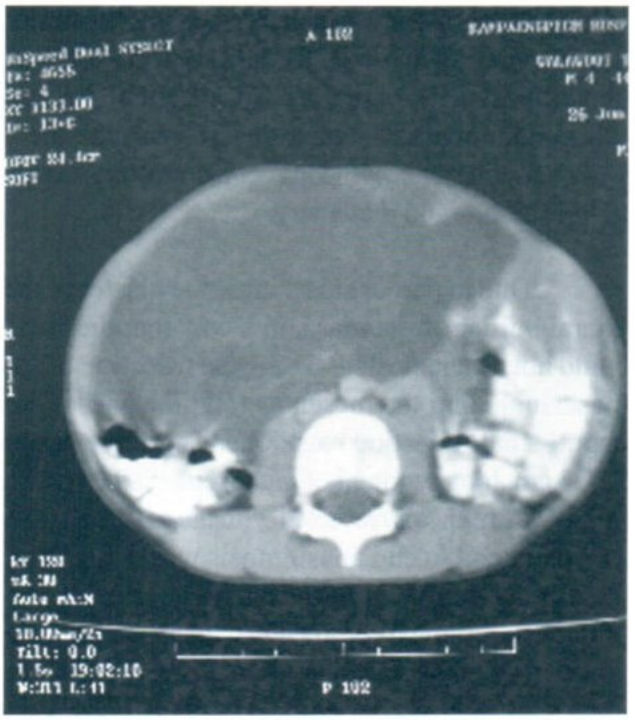

Fig.2B

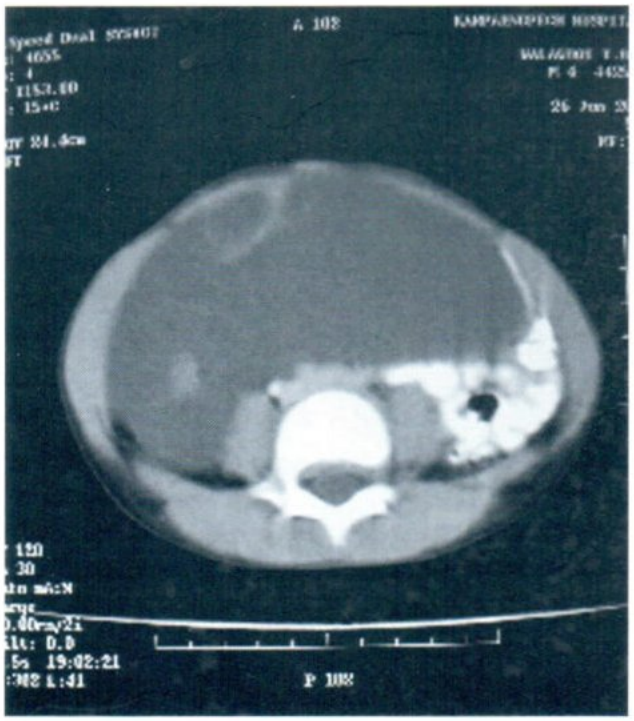

Fig.2C

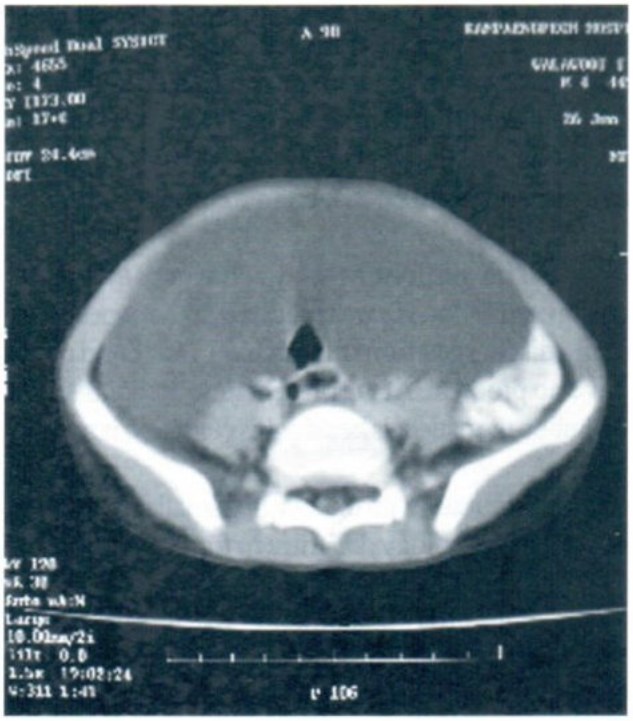

Fig.2D

Fig.2A-D CT images obtained with intravenously administered contrast material show huge bilobed nonenhancing cystic mass occupying nearly entired abdomen and pelvic cavity, with associated thin wall, well encapsulation, and poor enhancing faint internal septations.

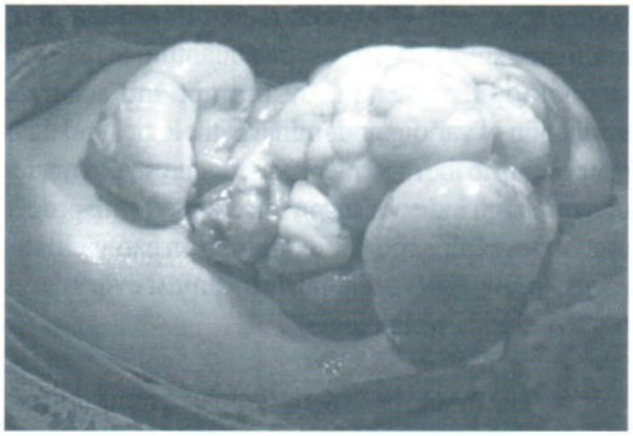

Fig.3 Operative photograph shows bunches of lymphangioma in the mesentery of small bowel, which was seen popping out of abdominal incision.

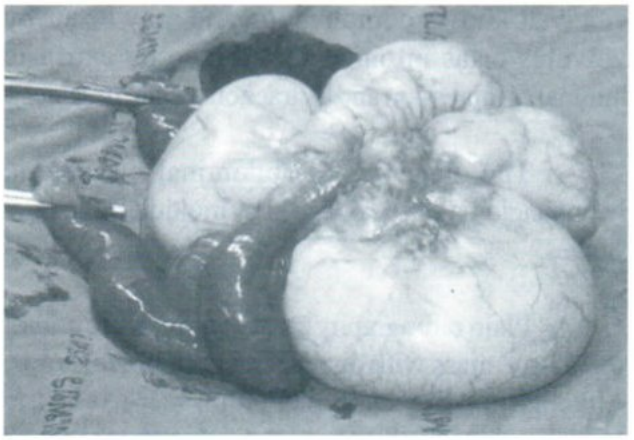

Fig.4 Photograph of the gross specimen shows that the cyst is multiloculated with a long segment of small intestine plastered to its surface. There is no communication between the intestine and the cyst. This picture of mass shows good correlation with CT appearances.

DISCUSSION

Lymphangioma has been classified into three groups: (1) lymphangioma simplex; (2) cavernous lymphangioma; and (3) most frequency, cystic lymphangioma, or cystic hygroma.

Histologically, a lymphangioma can be well circumscribed lesion composed of one or multiple large cysts which can interconnect. These are typically called cavernous lymphangioma. A lymphangioma can also be composed of microscopic cysts, producing an ill defined, compressible, spongelike lesion, known as a cystic lymphangioma. The walls of lymphatic spaces are thin and contain fibrous tissue, smooth muscles and aggregates lymphoid tissue. The lymphatic spaces and channels are lined by one row of the flat endothelial cells. There is no discrete capsule. In the small bowel mesentery, the spaces may be filled with chyle and are called chylous lymphangioma or may be occasionally filled with hemorrhage.

Intrabdominal cystic lymphangioma of the mesentery is a rare congenital lesion with a relative low growth potential. It typically found in young adult and present with chronic features.10 In younger children as in our patient, the lymphatic malformations usually are aggressive with rapid growth. Actually mesenteric cystic lymphangioma is a rare cause of bowel obstruction and preoperative diagnosis is difficult due to silent clinical course.

Most common presenting symptoms are abdominal distention with a palpable mass, followed by abdominal pain. However, the lesion may not be palpable due to its flaccid and mobile nature. Symptoms were presented in all patients including abdominal pain, bilious or nonbilious vomiting and diarrhea.

Our reported case was a chylous cystic mesenteric lymphangoma in a young child which presented as a diagnostic dilemma. Our patient's symptoms include abdominal distention, palpable mass and small bowel obstruction.

Complications are volvulus, intestinal obstruction, infarction, perforation and intracystic hemorrhage. Perforation and hemorrhage may be spontaneous and may be caused by trauma or minor trauma. Partial or complete small-bowel obstruction

is a well known complication and may occur by volvulus or as in our reported case by extrinsic luminal compression and by traction on the mesentery.

The diagnosis of abdominal mesenteric lymphangioma can be deduced by multiple radiologic studies.

Plain radiography most commonly reveals a soft-tissue mass with displacement of bowel loops. Although not diagnostic, plain films help to identify complications such as bowel displacement and/or intestinal obstruction.

The typical US appearance is a large well -circumscribed anechoic cystic or multicystic mass, often with thin wall, multiple thin septations and posterior acoustic enhancement. They also showed variable internal echogenicity, which is accounted for by the various contents that are possible. Occasionally, some solid echogenicity with a honeycomb pattern have also been described. However, the exact origin of the mass may not be ascertained on ultrasonography.

The CT findings demonstrated multiloculated or bilobed fluid-filled masses. The attenuation of the fluid ranged from that of fluid to that of fat, again according to the different content. Lymphangiomas have also been characterized as being well defined with a thin wall and occasionally with septa. The mass usually show no enhancement on IV contrast administration, but the thin septae inside the mass may enhanced faintly. The lesion may be distinguished from ascites by the absence of bowel loop separation or fluid in the typical sites, such as the cul-de-sac, or the presence of focal septa.

The magnetic resonance (MR) imaging can also be used to demonstrate the relationship of the mass and surrounding structures. Actually MR has been considered superior to CT in determining both the exact origin of a cyst and in assessing its exact extent due to the ability of viewing the lesion in multiple planes. MR helps in characterizing its contents as well. However, recent introduction of multislice spiral CT represent a quantum leap in CT technology that has ensure prompt and accurate evaluation of various abdominal pathologies and their relationship to the mesenteric vasculatures. As a matter of fact, a high index of suspicion coupled with proper ultrasonography and CT examination can yield the correct preoperative diagnosis in almost every cases using multislice technology.

The cystic mesenteric lymphangioma is a rare benign tumor and its diagnosis should be included in the differential diagnosis of cystic intra-abdominal neoplasias. The most common type of mesenteric or omental cystic tumor is the lymphangioma, followed by a nonpancreatic pseudocyst, duplication cyst, mesothelial cyst, and enteric cyst. The differential diagnosis includes the other histologic types of mesenteric and omental cysts mentioned earlier, and there are no specific radiologic features. Another consideration is a cystic leiomyoma or leiomyosarcoma, and again differentiation requires histologic evaluation. A cystic teratoma should also be included, but one would expect fat content with one or more clusters of calcifications. Nevertheless, a mesenteric lymphangioma with peripherally calcified caseous material has been documented. Mesotheliomas may also appear cystic, thus requiring a histologic diagnosis, but these usually occur in middle-aged women.

Due to the tumor's location, as in the our patient, the surgeon should plan the appropriate surgery as to avoid morbidity, since the objective of such treatment is curative. The goal of surgical therapy is complete excision of the mass. Mesenteric cystic lymphangioma can be removed without endangering the adjacent bowel. The preferred treatment of benign well encapsulated mesenteric lyphangioma is (1) first option, enucleation, although intestinal resection is frequently required to ensure that the remaining bowel is viable. (2) Second option, involved mesentery and bowel resection may be required in cases of infiltrative lymphangioma. Any resulting mesenteric defect must be closed to prevent an

internal hernia. If enucleation or resection is not possible because of the size of the cyst or because of its location deep within the root of the mesentery, (3) the third option is partial excision with marsupialization of the remaining cyst into the abdominal cavity. Approximately 10% of patients require this form of therapy. If marsupialization is performed, the cyst lining should be sclerosed with 10% glucose solution, electrocautery or tincture of iodine to minimize recurrence. Partial excision alone with or without drainage is not indicated because of the high recurrence rate associated with this procedure.[11] Lymphangioma has an unsatisfactory therapeutic outcome due to its invasive nature.

Other modalities of treatment for unresectable intrabdominal lymphangioma include sclerotherapy with doxycicline, alcohol, bleomycin, Picibanil (OK -432). More recently, new attempts to sclerose these lesions, scerotherapy is now being performed with increasing frequency, and have met with more success than in the past.[6,8]

In our patient, a small-bowel loop engulfed to the mesenteric lymphangioma and had to be excised with the the mass lesion (second option). After surgery, our patient was alright and well-tolerated, and he was discharged without complications or further incidents.

CONCLUSION

In conclusion, mesenteric cystic lymphangioma is a rare congenital benign tumor. The most common presenting symptoms are abdominal pain, abdominal distension, palpable mass, followed by intestinal obstruction and increasing abdominal girth.

Although mesenteric lymphangiomas are rare tumor, they should be considered as a possible cause of acute abdomen especially in children.

Mesenteric cysts must be considered when evaluating children with abdominal pain and mass, associated clinical findings of small-bowel obstruction should prompt suspicion of complicated midgut volvulus.

Cystic teratoma, cavernous hemangioma, pancreatic/nonpancreatic pseudocyst, duplication cyst, mesothelial cyst, mesenteric cyst and enteric cyst, all should be kept in mind in the differential diagnosis of mesenteric cystic lymphagioma.

We thought that proper abdominal ultrasonography complemented by multislice spiral CT can yield the correct preoperative diagnosis in almost every case, but histological examination will confirm the diagnosis.

Complete resection is the treatment of choice and has an excellent prognosis. Although an abdominal lymphangioma is considered benign, it may become locally invasive. Therefore any involved organ must also be resected. Incomplete resection may lead to recurrence. Hence, a rapid and accurate preoperative localization of the lesion is critical and important for facilitating proper management.

Follow-up imaging, therefore, is advised, with ultrasound as the modality of choice.[12]

The effectiveness of radical surgical excision is higher than that of sclerosing therapy, but the risk of complication with surgery was greater, may be three times in paper of Hiki Saori et al.[6,8]

As tumor was diffuse, total excision may require resection of involved abdominal organ and bowel to prevent recurrence.

Early recognition and appropriate treatment of these lesions are associated with a good outcome, provided that, if the tumor is completely excised, a good prognosis could be archived.

Although the lesions tend to surrounds and sometime invades normal anatomic structures it

presumably have no malignant potential, but it has been described in few literatures that malignant degeneration is usually to a low-grade sarcoma, but this very rarely occurs.

ACKNOWLEDGEMENT

I would like to thank Mrs. Jiraporn Maneeprai for her help with this project about reference papers, patient's profiles and manuscript preparation.

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