to 5 cm below the costal margin. Jaundice gradually deepened and he died after an episode of hematemesis.

Case 2: A 2-year-old boy presented with multiple scalp swellings and scalp lesions for 3 months. There was history of discharge from both ears for 3 weeks. He had multiple soft to firm swellings over the scalp, the largest being 4 cm x 3 cm. There was protrusion of the right eye and firm hepatomegaly 8 cm below the costal margin. The rest of the examination was unremarkable.

The blood counts and renal function tests were normal. Liver function and coagulation screening tests were normal, except for serum alkaline phosphatase (110 KA units). X-ray of the skull revealed multiple lytic lesions. The findings on fine-needle aspiration cytology from the scalp swellings were morphologically consistent with a diagnosis of LCH.

He was started on intravenous etoposide (150 mg/m²/day x 3 days, every 3 weeks x 6 courses) and oral prednisolone (2 mg/Kg/day initially with gradual tapering over 15 months). The patient was completely asymptomatic and a firm splenomegaly (3 cm below the costal margin) was observed after 1.5 years. CECT scan revealed a liver with lobulated outline and ill-defined poorly enhancing areas in both lobes. Percutaneous transhepatic biopsy demonstrated disrupted architecture. Wide irregular bands of fibrosis were seen to traverse the hepatic parenchyma, with bile ductular proliferation and minimal inflammation. Hepatocytes were normal. Endoscopy revealed grade I neoplastic varices. On his last visit, at the age of 6.5 years, the LCH was in remission, hepatosplenomegaly was persisting, and he had no history of gastrointestinal bleed.

A definite diagnosis of LCH is based on demonstration of CD1 antigen determinants on the foamy histiocytic cells, characteristic Birbeck granules on electron microscopy. We did not have facilities for these two parameters. The light microscopic findings in our cases, coupled with the typical clinical manifestations and response to therapy with oral prednisolone and/or etoposide, lend support to a diagnosis of LCH.

Hepatomegaly is common in LCH. Histologically, the liver shows portal infiltration with CD1 positive cells that rarely contain Birbeck granules. Other hepatic manifestations include obstructive jaundice with a histological picture resembling sclerosing cholangitis or a solitary nodule in the liver parenchyma. Other changes that have been described include portal triads, bile ductular distortion and proliferation, variable hepatic fibrosis with histiocytic infiltrates, and cirrhosis.

Engelbreth-Holm et al. described four histological stages of LCH: proliferative, granulomatous, xanthomatous, and fibrous. It is postulated that, despite chemotherapy, progressive fibrosis occurs in areas previously infiltrated by histiocytes, ultimately producing cirrhosis, lung fibrosis, diabetes insipidus and growth hormone insufficiency.

To summarize, liver damage in LCH can be independent of local or generalized LCH activity. Treatment plans must keep in mind the possibility of portal hypertension and fatal liver failure despite LCH being in remission.

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Multicystic mesothelioma of the peritoneum

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Multicystic mesothelioma is an uncommon lesion presenting as an abdominal mass. We report a 40-year-old woman presenting with recurrent intra-abdominal lump. She had undergone abdominal surgery thrice before. The lump at this presentation was partially excised surgically; histology and immunohistochemistry confirmed the diagnosis. [Indian J Gastroenterol 2001;20:202-203]

Key words: Mesothelial tumor

Multicystic mesothelioma (syn: cystic mesothelioma) is an uncommon lesion, most frequently described in the pelvic peritoneum of young or middle-aged women. Prior to their description by Mennemeyer and Smith, these were termed as lymphangiomas or as reactive processes. The neoplastic nature of these lesions has now been proved. Recent ultrastructural studies have shown that these tumors are of mesothelial cell origin.

Even though the overall clinical behavior is benign, multiple post-surgery recurrences are known.

A 40-year-old woman presented with a gradually increasing mass in the abdomen since six months. She had a history of three surgical interventions. Hysterectomy with right salpingo-
oophorectomy was done ten years back for menorrhagia due to fibroids. Five years later, she developed a cystic mass in the abdomen, which was excised. The specimen had multiple thin-walled cysts of varying sizes in the omentum and mesentery containing hemorrhagic fluid. Histologically, it was labeled as endometriosis and the patient was treated with danazol. She had a third surgery when a recurrent, large cystic mass in the lesser sac that was adherent to important structures was partially excised. The left ovary was removed at the same time.

On examination, she had huge distension of the abdomen. Multiple large, firm, mobile masses were palpable in the umbilical, left hypogastric and iliac regions. There was no free fluid.

**Investigations:** normal chest X-ray. Hepatic function tests and serum CA-125 level were within normal limits. CT scan of the abdomen and pelvis showed large hypodense, multiloculated, cystic masses filling the pelvis, abdomen and extending up to the diaphragm. Multiple thin septae were seen within the mass. It was infiltrating the mesentery and encasing the bowel loops. It had also invaded the anterior paravertebral space causing scalloping of the anterior cortex of the left kidney and medial surface of the spleen. During surgery, a debulking procedure was carried out. The patient recovered uneventfully.

The excised specimen measured 14 cm x 11 cm x 4 cm and consisted of a cystic structure with smooth external surface. On cutting open, it was multiloculated, with multiple small cysts measuring 0.5 cm to 1.5 cm in diameter, having a thin wall and containing hemorrhagic fluid. Sections from the mass showed multiple cystic spaces separated by delicate fibrovascular stroma. The lining of the cyst was either flattened cuboidal endothelium-like or stratified at places. Small buds, tufts of cells or detached clumps or mesothelial cells were also present within the lumina of the cysts (Fig.). Immunohistochemical study was carried out using a panel of antibodies comprising cytokeratin (Dako A/S, Denmark, dilution 1:100), epithelial membrane antigen (Dako, 1:100) carcinoembryonic antigen (CEA, Dako, 1:400), mesothelioma-associated antigen (Meso-Ag, Dako, 1:50) and factor VIII-associated antigen (factor VIII-Ag, Dako, 1:800) using the avidin-biotin-peroxidase complex (ABC) and the peroxidase-antiperoxidase (PAP) technique with 3,3'-diaminobenzidine as the chromogen. The cells lining the cysts showed positivity for cytokeratin and epithelial membrane antigen, but were negative for CEA, Meso-Ag and factor VIII-Ag. The endothelial lining of the vessels in the fibrovascular septae showed strong positivity with factor VIII-Ag.

**Clinically,** multicystic masses in the abdomen in a woman could be arising from the ovaries, pancreas and rarely the kidneys, liver or soft tissues (lymphangioma). The ovaries in this patient had already been removed. The other abdominal organs were normal on CT scan. Lymphangioma is more localized in its growth and is composed of endothelial-lined spaces that are more uniform in size. The walls of these cysts may show smooth muscle and lymphocytes. The endothelial lining does not show the stratification, budding or detachment seen in the present case. In problematic cases, immunohistochemistry can be of help by demonstrating positivity with antibody against factor VIII-Ag.

**Neoplasms of the peritoneum in women can be classified as metastatic, Müllerian or of mesothelial origin.** Tumors producing pseudomyxoma peritonei are commonly associated with mucinous tumors of the ovary, appendix, gall bladder or colon. Primary peritoneal tumors of Mullerian origin are linked to the concept of the secondary Mullerian system. The lesion reported here is probably not related to the secondary Mullerian system as it lacked the typical histologic features of endometriosis, endosalpingiosis or endocervicosis or corresponding malignant tumors of Mullerian origin such as serous, mucinous or endometrioid carcinomas.

Immunohistochemistry was not of much help in our case as staining with Meso-Ag was negative. More specific antibodies like E-cadherin and calretinin were not used. The histogenesis of this lesion is controversial. Mennemeyer and Smith reported a history of prior abdominal or pelvic surgery or pelvic inflammatory disease in 30% of their patients. Our case also had history of multiple surgeries in the past. Cystic mesotheliomas are best treated by surgical excision. The lesion, although benign, is punctuated by multiple recurrences and an indolent clinical course.

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**Fig:** Photomicrograph showing cystic spaces lined by single-layered or stratified epithelium (H&E, 100 X)