BILIARY CYSTADENOMA - COMPUTED TOMOGRAPHY FINDINGS

Ariff AR, Haida Hassan & John G*

Department of Radiology, Hospital Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia.

*Department of Radiology, University Malaya Medical Centre, 50603 Kuala Lumpur, Malaysia

Biliary cystadenoma is a rare neoplasm of the biliary ductal system. Surgical management yields an excellent result. We present a case of recurrent biliary cystadenoma in the left lobe of the liver. The cyst was successfully treated with hepatic segmentectomy. The lobulated smoothly marginated septated cystic lesion noted on computed tomography (CT) were highlighted and the other imaging studies, differential diagnosis and management were reviewed.

Key words: Biliary cystadenoma, Computed tomography.


Introduction

Biliary cystadenomas are large, usually multiloculated benign tumours. They arise from intrahepatic or, rarely extrahepatic ducts and represent less than 5% of solitary non parasitic cysts of biliary origin. This multilocular cyst is lined by cuboidal or columnar epithelium. Their cause remains unknown, although presence of aberrant bile ducts in cystadenomas suggest congenital etiology. No association with oral contraceptive use has been documented. If the cystadenomas are of developmental origin, then their late presentation must be ascribed to very slow growth and/or accumulation of fluid (1). This paper will outline the CT findings of a biliary cystadenoma and discuss the other diagnostic imaging modalities, management and the possible differential diagnosis of the cystic lesions.

Case report

A 56 - year - old medium built Chinese woman was admitted to the University Hospital for hepatic segmentectomy for a recurrent biliary cystadenoma which was identified by ultrasound during follow-up assessment at an out patient clinic, where a large thick walled septated cystic mass in the left lobe of the liver was found. A review of her past surgical history revealed that she had undergone an operation to deroof a cystic lesion in the left lobe of the liver 8 months previously. One hundred millilitres of clear yellowish colour fluid was aspirated. The histopathological report of the removed specimen revealed a biliary cystadenoma. She had also undergone hysterectomy for a carcinoma of the cervix 2 years ago. Her post hysterectomy was complicated with deep vein thrombosis which was successfully treated with warfarin. She was also hypertensive on oral propanolol 40 mg b.d for the past 2 years.

On her present admission, she complained of nausea and vomiting for the past two months associated with severe epigastric pain and diarrhoea. She had neither fever nor jaundice.

Physical examination revealed an abdominal mass in the epigastrium measuring 6.0 x 8.0 cm. The haematological and biochemical investigations were normal.
Computed tomography of the liver showed a well defined lobulated septated 9.5 x 6.5 x 6 cm cystic lesion in the left lobe occupying segments II and III (figure 1). The CT values of the cyst were ranging from +10 - +13 HU. In view of past history of cervical carcinoma, the possibility of cystic metastases was put into consideration.

The lesion was completely removed with a left lateral hepatic segmentectomy. Pathologic examination of the excised specimen was a biliary cystadenoma. The multiloculated cyst was well encapsulated and sharply demarcated from the adjacent tissue. The cystic spaces were filled with mucinous fluid.

Patient made an uneventful and complete recovery.

Discussion

The majority of biliary cystadenomas occur in middle aged women presenting with an abdominal swelling or a right upper quadrant mass of weeks to years duration. Pain and biliary obstruction may be present. Our patient had most of these features. Other clinical presentations include weight gain or loss, increased abdominal girth, anorexia, nausea, vomiting, jaundice and biliary colic. CT examination is useful in assessing the extent of involvement. CT and ultrasound are complementary for the evaluation of internal morphology, size and extent of this tumour (2).

The characteristic CT appearance of this tumour is that of a low-density intraheptic mass with internal septae and mural nodules. Typically, such lesions are multiloculated having low attenuation cystic regions ranging from 0 to + 40 HU. However CT attenuation numbers are of little value in separating these from other cystic lesions. Contrast enhancement is seen along the internal septa and wall. However CT failed to display the thick cyst wall (3) as in our patient.

Sonography usually depicts the true internal morphology of cystic hepatic lesions more clearly. Biliary cystadenomas are characterised on sonograms as septated, thick-walled, anechoic lesion with mural nodules and cysts. These findings virtually exclude a simple cyst. Septations are uncommon in abscesses or complicated cyst but occur commonly in cystic metastases. Septations, multiple loculations, internal echoes and fluid filled levels have been described in liver metastases (3).

Percutaneous cyst aspirations should be considered in indeterminate cases. Aspiration biopsy or surgery is usually required to differentiate metastases from primary biliary neoplasm. Cyst

*Figure 1: Intrahepatic Biliary Cystadenoma. Axial contrasted CT - scan of the liver revealed a lobulated, smoothly marginated septated cystic lesion in the left lobe (*)."
aspiration usually allows differentiation of a cystadenoma from a hematoma, abscess or cyst complicated by haemorrhage or infection. Biliary cystadenomas contain mucinous bile-tinged or brownish cloudy fluid without cells. Cholesterol crystals are detected occasionally (4). However, differentiation from cystadenocarcinoma or solitary cystic metastases may remain difficult. The presence of a solid nodular mass or calcifications along the wall or septa in a multilocular cystic mass indicates a more likely diagnosis of biliary cystadenocarcinoma.

Other imaging tests may be useful. Angiographically these tumours are usually hypovascular though abnormal clusters of vessels are frequently seen within their walls. Accumulation of contrast materials within the wall or septum may be seen on delayed imaging (3). Endoscopic retrograde cholangiopancreatography (ERCP) to investigate the cause of jaundice may demonstrate communication of the tumour with the bile ducts. Regardless of the diagnostic modality used, cystadenoma and cystadenocarcinoma cannot be differentiated macroscopically. Resection of cystadenomas is usually curative and since cystadenocarcinomas are believed to arise from benign cystadenoma, complete surgical resection when possible, is warranted to avoid malignant transformation of these rare tumours. Ultrasound, computed tomography and angiography are the imaging modalities required to evaluate these tumours adequately prior to surgery (2).

Correspondence:

Dr. Abdul Rahman Mohd Ariff, M.Med,
Department of Radiology,
Hospital Universiti Sains Malaysia,
16150 Kubang Kerian, Kelantan, Malaysia.

References:

CASE REPORT

EWING’S SARCOMA MIMICKING TUBERCULOSIS –
A CASE REPORT

B. Shalini, S. Wahinuddin, M. Monniaty & S. Rosemi

Department of Medicine, Hospital Kota Bharu,
15586 Kota Bharu, Kelantan, Malaysia

A 13-year-old Malay school girl who had been apparently normal previously, presented with a three-month history of fever, malaise and loss of weight. She had anemia and raised values for ESR, lactic dehydrogenase, C-reactive protein, ferritin and a positive Mantoux test. Her routine chest x-ray showed hilar prominence suggestive of hilar lymph node enlargement. C.T. Scan of the thorax revealed a posterior mediastinal mass, the histopathology of which was suggestive of Ewing’s sarcoma. The rarity of the location of the tumour and its unusual mode of presentation prompted us to report this case.

Key words: tuberculosis, posterior mediastinal mass, extraosseous Ewing’s sarcoma.


Introduction:

Ewing’s Sarcoma (ES) is a malignant neoplasm of the bone and sometimes of soft tissues with characteristic radiological, morphological immuno-histochemical and cytological features. ES has now been shown to belong to a family of tumors with overlapping histopathological features and comprising of Ewing’s sarcoma (ossaceous and extra-osseous), peripheral primitive neuroectodermal tumor (PPNET) and Askin tumor (1). We report a teen-ager with extra-osseous ES who presented with prolonged fever, positive Mantoux test and a posterior mediastinal mass.

Case Report:

A 13 year-old Malay school girl was admitted with a three-month history of fever, loss of weight and generalized myalgia. She did not give a history of cough, chest pain, shortness of breath or night sweats. There was no history of joint pain, hair loss, rash or photosensitivity.

On examination, she was thin, pale, febrile and tachycardic. There were no vasculitic lesions, alopecia or malar rash. The respiratory, cardiovascular and abdominal examinations were normal and no lymph node was palpable.

Laboratory evaluations revealed a hemoglobin value of 9.3 gm/dl, white blood cell count of 9.2 x 10^9/dl and platelet count of 564 x 10^9/l.

The erythrocyte sedimentation rate (ESR) was high (138 mm/1st hour) and the Mantoux reaction was positive (18 mm). The acute phase proteins like lactic dehydrogenase (916u/l), C-reactive protein (19.16 mg/dl) and serum ferritin (1000 ng/dl) were all elevated. The liver and renal functions were normal. Repeated blood cultures for pathogenic bacteria were negative. Sputum examination for acid-fast bacilli was negative. Tests for connective tissue disease were negative. The chest X-ray revealed a left hilar prominence but the lungs were normal (figure 1). The ultrasound examination of abdomen and echocardiogram were normal. Since the clinical features and laboratory reports highly suggest of tuberculosis, the patient was started on empirical anti-tuberculous therapy (isoniazid 300 mg, rifampicin 300 mg and pyrazinamide 1000 mg). However there was no improvement after two weeks of anti-tuberculous therapy. She was then subjected to bronchoscopy which showed that the left upper
Figure 1: Chest X-Ray: (L) hilar prominence

Figure 2: CT Scan Thorax: A Heterogenous mass adjacent to the vertebra
lobe bronchus was slit-like, suggesting the presence of external compression. Bronchial brushings and washings were negative for acid-fast bacilli (AFB) and malignant cells.

C.T. scan of thorax showed a heterogenous, enhancing lesion in the left hemithorax at the level of the carina, adjacent to the vertebral bodies. No calcification was seen within the mass. There was no evidence of parenchymal lesion in the lung or of pleural effusion (figure 2); and the adjoining vertebrae and ribs were intact indicating thereby that the tumor had not arise from bone and extended into the mediastinum.

Meanwhile the patient continued to remain febrile and her hemoglobin level dropped to 8.4 gm/dl. The blood picture was in favour of microcytic hypochromic anaemia. Biopsy of the posterior mediastinal mass was done under C.T. guidance. The histopathology showed sheets and lobules of primitive small ovoid round cells which displayed high nuclear cytoplasmic ratio, fine nuclear chromatin and punched-out clear cytoplasmic vacuoles. The cells also showed periodic-acid-Schiff stain (PAS) positive inclusions.

These features were in favour of an Ewing’s sarcoma. The patient was referred to the oncologist for further management.

Discussion:

The first large series (39 cases) of extraosseous Ewing’s sarcoma (EOES) was published by Angervall and Enzinger (2) in 1975, of which 12 patients had the tumor in the paravertebral region mainly at the lumbar and sacral level. Subsequently it had been shown to occur at various sites in the human body including the soft tissues of the orbit, vagina, kidney and the posterior mediastinum (3,4,5,6,7). In many of the instances, the diagnosis has been made postoperatively after excision of the mass or by biopsy procedures.

The posterior mediastinal presentation of EOES is also uncommon. To our knowledge no similar case of posterior mediastinal EOES has so far been reported from Malaysia.

In view of the posterior mediastinal and paravertebral location of the tumor, it is quite often mistaken for a neurofibroma (3). In our case, the mediastinal mass associated with a raised ESR, positive Mantoux test and other constitutional symptoms in a teen-ager led us to the diagnosis of tuberculosis. The histopathological report of the biopsy specimen of the mass was compatible with that of ES but for which the diagnosis would have eluded us. Elevated ESR, positive Mantoux reaction and raised acute phase proteins have not been reported in the EOES cases described in the current literature. The explanation remain obscure, though it does raises a possibility of an associated tuberculous illness in our case.

Therefore in the differential diagnosis of posterior mediastinal tumors, EOES should also be borne in mind especially in young patients. Appropriate and early histological diagnosis is essential to plan appropriate management.

Correspondence:

Dr. Shalini Bhaskar MBBS, Department of Medicine, Hospital Kota Bharu, 15586, Kota Bharu, Kelantan.

References:

7) Pallavicini EB, Burgio VL. Extraosseous Ewing’s sarcoma (Italian).Minerva med. 1997; 70: 2897 - 901