KCR 2014

Case of the Day

October 9(Thu) - 11(Sat), 2014
Hall D2, Convention Hall 3F, Coex
40/F
Trismus & headache for 8 months

**Question**
What is your diagnosis?
1) Lymphoma
2) Meningioma
3) Pachymeningeal metastasis
4) Hypertrophic pachymeningitis
5) Venous infarction with venous sinus thrombosis

**Answer**
4) Hypertrophic pachymeningitis
**Imaging Features**

Peripheral lobulating lesion is noted along the right inferior frontal convexity. The enhancement is predominantly at the meninges on contrast enhanced T1WI. T2WI shows hyperintense meningeal thickening with segmental dark signal in the medial side, and diffuse T2 hyperintense brain parenchyma at the right frontal lobe and corpus callosum genu. Decreased NAA peak and slightly increased choline peak in the right frontal subcortical white matter are noted on MRS with 288 ms TE, and small lactate peak was noted at 1.3 ppm.

**Discussion**

Hypertrophic pachymeningitis is a rare chronic inflammatory disorder which is characterized by marked fibrous thickening of the cerebral or spinal dura mater. Chronic headache and multiple cranial neuropathies are the common clinical manifestation. Its etiology remains unclear but infection, autoimmune disorder, sarcoidosis and neoplasms have been thought as possible causative factors.

This patient did not have any identifiable cause and the diagnosis can be called ‘idiopathic hypertrophic pachymeningitis’. The symptom of chronic headache and trismus due to motor disturbance of the trigeminal nerve was typical.

Contrast-enhanced study is indispensable for the investigation of hypertrophic pachymeningitis. MRI is more sensitive than CT to show the linear or nodular thickened dura matter. The lesion appears hypointense or isointense on T1WI and hypointense on T2WI or FLAIR image.

High dose of steroid therapy usually can induce dramatic improvement and change of symptom and appearance on images. Cerebral edema and venous sinus thrombosis can complicate this disease process.

**References**

What is your diagnosis?
1) Emphysema
2) Lymphangioleiomyomatosis (LAM)
3) Langerhans cell histiocytosis (LCH)
4) Lymphocytic interstitial pneumonia (LIP)
5) Birt-Hogg-Dube syndrome (BHD)
**Answer**

2) Lymphangioleiomyomatosis (LAM)

**Image interpretation**

Numerous discrete, round, thin-walled multiple air cysts are noted on CT images. There is no definite predominant distribution of the air cysts. Abdomen CT also demonstrates fatty masses in the left kidney.

![CT images of LAM](image)

**Discussion**

Lymphangioleiomyomatosis (LAM) is a rare interstitial lung disease that affects women exclusively, typically during their reproductive years. A small percentage of patients, also typically women, have LAM in association with tuberous sclerosis complex (TSC). LAM is characterized by the abnormal proliferation of smooth muscle cells (LAM cells) in the lungs and in the thoracic and retroperitoneal lymphatics. Affected patients are at risk of developing renal hamartomas or angiomyolipomas. Patients with LAM characteristically present with chronic dyspnea and cough and less commonly with spontaneous pneumothorax. Tuberous Sclerosis Complex is an inheritable autosomal dominant disorder characterized by multifocal systemic hamartomas that may affect the central nervous system, skin, heart, kidneys, and other organs. The Vogt triad of epileptic seizures, mental retardation, and facial angiofibromas (formerly misnamed as adenoma sebaceum) is exhibited by only approximately 29% of affected patients. Relationship between TSC and LAM; Although histologic and clinical features may be identical in women with LAM and patients with TSC-associated pulmonary involvement, there are reported differences. Patients with TSC-associated LAM, compared with patients with sporadic LAM, generally experience a longer delay between onset of symptoms and diagnosis of pulmonary disease, exhibit
chylothoraces less frequently, and are more likely to present with gradual onset of dyspnea. Radiographic abnormalities in patients with LAM have been reported as a pattern of generalized, symmetric, reticular, or reticulonodular opacities, seen in approximately 80%-90% of affected patients. Lung volumes have been reported as normal at chest radiography in 55%-78% of affected patients and increased in 22%-45%. Pneumothorax is a common clinical and radiographic presenting manifestation of LAM, reported in 39%-53% of patients. Pleural effusion (secondary to chylothorax) is a common radiographic feature, occurring in 10%-20% of patients, and may be unilateral or bilateral. The CT manifestations of LAM are distinctive, characterized by numerous thin-walled cysts surrounded by normal lung parenchyma and distributed diffusely and bilaterally. Occasionally, a chyloous effusion is suggested at chest CT based on low-attenuation (−17HU) fluid that may relate to the presence of fat. Renal angiomyolipomas have been observed on abdominal CT scans of 20%-54% of patients with LAM. These tumors are characterized by CT evidence of soft tissue, fat, and enhancing vessels in variable proportions within a renal mass. A variety of intracranial manifestations of TS are known. Four common CNS abnormalities are cortical tubers, subependymal nodules, subependymal giant cell astrocytomas (SGCAs), and white matter abnormalities. Except for SGCAs, these abnormalities can be seen in almost all patients with TS.

References

32/M
Right flank pain.

Fig. 1. Axial CT
Fig. 2. Coronal CT

Fig. 3. Coronal HASTE
Fig. 4. Coronal HASTE
Fig. 5. Coronal HASTE

Fig. 6. Coronal 20-min hepatobiliary phase
**Question**

What is your diagnosis?

1) Choledocal cyst
2) Hydatid cyst
3) Intraductal papillary neoplasm with invasive adenocarcinoma
4) Biliary cystadenocarcinoma
5) Cystic metastasis

**Answer**

3) Intraductal papillary neoplasm with invasive adenocarcinoma

**Imaging Features**

Contrast enhanced CT scans (Figs. 1 and 2) show an approximately 17-cm lobulating multilocular cystic mass at the right liver and left medial segment. On coronal HASTE MR images (Figs. 3–5), some cysts show either tubular shaped or cystic shaped. The some portions appear to be connected to other cystic portions. There are small nodular portions within the cystic mass. On coronal 20-min hepatobiliary phase, the nodular portions show mild enhancement.

**Brief Review**

Intraductal papillary neoplasm of bile duct (IPNB) has been recently recognized as a new entity of biliary neoplasm which includes previous categories of biliary papilloma and papillomatosis. IPNB is considered as a premalignant lesion in the dysplasia-carcinoma sequence, and if there is an invasive component, it would be referred to as IPN with an associated invasive carcinoma, and can be progressed to intraductal growing cholangiocarcinoma. Histologically, IPNB is characterized by the intraductal papillary or villous growth of biliary epithelium covering fine fibrovascular stalks. IPNB is known as a biliary counterpart of intraductal papillary mucinous neoplasm (IPMN) of the pancreas based on their pathologic similarities, although macroscopic mucin production is less frequent in the IPNB than in the IPMN of the pancreas. IPNB can appear as cyst-forming masses because cystic or aneurysmal dilatation of the bile duct occurs as a result of excessive mucin production by papillary tumor cells lining the inner surface of the cyst. Because cystic IPNBs usually manifest as unilocular or multilocular cystic lesions in the liver, imaging differentiation from mucinous cystic neoplasms can be challenging. Prominent mural nodules in cystic tumors usually suggest the possibility of biliary cystadenocarcinoma or cystic IPNB, whereas mural nodules are rare in cases of biliary cystadenoma. In addition, the presence of dilated bile ducts distal to the cystic tumor, which indicates communication between the bile ducts and cystic mass, favors a diagnosis of cystic IPNBs.
References

Case 4.

Urology by Deuk Jae Sung

M/74
Gross hematuria and dysuria

Fig. 1. Initial contrast-enhanced CT.

Fig. 2. Three-month follow-up MRI.
**Question**

what is your diagnosis?

1) Renal cell carcinoma
2) Transitional cell carcinoma
3) Membranous glomerulonephritis
4) Xanthogranulomatous pyelonephritis
5) Renal tuberculosis

**Answer**

2) Transitional cell carcinoma

**Imaging Features**

Initial contrast-enhanced CT images show thrombus in right renal vein and poor excretion of contrast material in the collecting system (Fig. 1). Follow-up MR images demonstrate enhancing, ill-defined mass and wall thickening along the collecting system of right kidney with caliectasis in lower pole. In addition, MR images show an enlarged thrombus in the right renal vein with extension into the IVC (Figs. 2 and 3).
Brief Review

The clinical manifestations of renal vein thrombosis depend on the age of the patient, the specific disease process, and the speed with which it occurs. The causes of renal vein thrombosis are either neoplastic or nonneoplastic. Nonneoplastic renal vein thrombosis is usually caused by an underlying abnormality of the clotting system or the kidney itself or, in infants, dehydration. In adults, the most common underlying abnormality is membranous glomerulonephritis. Renal vein thrombosis is also a complication of the hypercoagulable state in patients with SLE. But, renal vein thrombosis can be associated with tumor and is frequently caused by direct tumor extension. Neoplastic renal vein thrombosis occurs most commonly in cases of renal cell carcinoma and occasionally in cases of transitional cell carcinoma. In renal vein thrombosis associated tumors, CT or MRI can show an enhancing soft-tissue lesion in the renal parenchyma or renal pelvis, and inhomogeneous enhancement within the thrombus. Tumor thrombus in the renal vein may also result from a left adrenal tumor.

References

74/M
Chief complaint: recently aggravated dyspnea

Question
What is your diagnosis?
1) Myocardial infarction
2) Viral myocarditis
3) Hypertrophy cardiomyopathy
4) Hypertensive cardiomyopathy
5) Cardiac amyloidosis
Cardiac MRI shows concentric left ventricular hypertrophy and minimal pericardial effusion. T2 weighted MRI shows no myocardial edema. But, Delayed enhancement MRI demonstrates global subendocardial enhancements in the left ventricle.

Cardiac amyloidosis is a type of secondary nonischemic cardiomyopathy that results from extracellular accumulation of insoluble fibrillary amyloid proteins. Several different forms of cardiac amyloidosis are recognized, including light chain (amyloid Light-chain: AL), familial, and senile amyloidosis, which can have widely varying clinical presentations. AL amyloidosis is the most common form, associated with plasma cell dyscrasias and the production of light chain protein. Cardiac involvement is present in half of plasma cell dyscrasias, and has a particularly poor prognosis with a median survival of less than 6 months in untreated patients with symptoms of heart failure.

Cardiac magnetic resonance (MR) imaging in suspected amyloidosis should include delayed enhancement MR sequence. Functional MR images show concentric ventricular hypertrophy, enlarged atria, restricted diastolic filling, and normal to decreased ejection fraction. Expansion of the extracellular space by the amyloid proteins results in gadolinium retention, which manifests as delayed enhancement. In addition, the absence of contrast material within the blood pool on delayed enhancement MR image results in the characteristic dark blood pool. As the distinct pattern of delayed enhancement, the global transmural or global subendocardial enhancements correlate with the greatest amount of amyloid deposition. In general, incomplete nulling of the myocardial signal on the inversion recovery scout sequence can be seen. So, if difficulty with myocardial suppression on the delayed contrast-enhanced image is noted, then, cardiac amyloidosis should strongly be considered as the cause.

References

**Question**

An otherwise healthy 37-year-old woman admitted to ENT clinic due to her headache, facial pain, and bloody-purulent rhinorrhea. Her symptoms got aggravated 2 weeks ago. Her physical examination revealed a large polypoid mass completely obstructing the left nasal airway, without evidence of increased vascularity. What is the most likely diagnosis?

1) Mucocele
2) Inverted papilloma
3) Malignant lymphoma
4) Organized hematoma
5) Squamous cell carcinoma
Answer

4) Organized hematoma

Her non-contrast computed tomogram showed an expansile lesion centered in the left maxillary
sinus, and extending to the ipsilateral nasal cavity. On her T1-, T2-weighted axial images, it appears
as a round soft tissue lesion with mixed signal intensity, surrounded by very low signal rim, especially
on T2-weighted image. After Gd-administration, it shows markedly heterogeneous enhancement. A
biopsy specimen via endoscopic approach showed only chronic inflammation, hemorrhage and fibro-
sis, however, there was no evidence of malignancy. Because the clinical biopsy findings were unre-
vealing, sinonasal organized hematoma was considered as final diagnosis.

Organized hematoma of sinonasal cavity is a rare, non-neoplastic condition that can be locally
aggressive. Sometimes it is associated with history of facial trauma, surgery, or hemorrhagic
sinonasal lesion. About 70% of patients with sinonasal organized hematoma complain of his/her
recurrent epistaxis. The clinical presentation and radiographic appearance can mimic malignancy, but
definitive diagnosis is confirmed only after histopathologic evaluation.

On computed tomograms, the lesion typically appears as a primarily expansile soft tissue mass
smoothly eroding the bony wall. Magnetic resonance images usually demonstrate a inhomogeneous
mass on both T1- and T2-weighted images. Mostly, they show marked heterogenous enhancement
(ex: papillary or frond-like enhancement) representing sites of neovascularity that occur within an
organizing hematoma. Moreover, peripheral hypointense rim, as characteristic feature to suggest
fibrous pseudocapsule may be obvious on of T2-weighted image.

There are multiple hypotheses regarding the pathogenesis of an organized hematoma. One theory
suggests that intranasal bleeding causes blood accumulation in the maxillary sinus cavity. Another
involves rupture of the vascular branch supplying the maxillary sinus or submucosal bleeding in the maxillary sinus due to loss of mechanical integrity of an arterial branch. Regardless of the inciting event, this condition evolves in several stages. Once blood accumulates in the maxillary sinus, a hematoma develops there. Because of poor ventilation and drainage, the hematoma undergoes neovascularization and fibrosis, thus leading to an organized hematoma. The mass tends to grow slowly and eventually can even cause pain owing to compression of surrounding structures.

The differential diagnosis is extensive and includes infectious, inflammatory, and neoplastic lesions. In cases in which there is no radiographic bone erosion, the differential diagnosis includes sinusitis, extensive polyposis and cysts, inverting papilloma, hemangioma, and mucocele. In cases involving radiographic bone expansion and destruction, the differential diagnosis includes both infectious and neoplastic processes. Fungal sinusitis is an important infectious disease process that needs to be taken into consideration. Malignant conditions include adenoid cystic carcinoma, squamous cell carcinoma, embryonal rhabdomyosarcoma, lymphoma, and tumors of dental origin. Pathologic confirmation of the diagnosis is essential. Histologic findings include hematoma, fibrosis, neovascularization, and no evidence of malignancy.

References

Case 7.
Abdomen by Suk Keu Yeom

91y/M
Right inguinal pain

Fig. 1

Fig. 2

Fig. 3
**Question**
What is your diagnosis?
1) Dermoid cyst
2) Foreign body abscess
3) Amyand’s hernia
4) Obturator hernia
5) Deep vein thrombosis

**Answer**
3) Amyand’s hernia (Inguinal hernia with acute appendicitis)

**Imaging features**
A 91-year-old woman presented with a 3-day history of swelling and pain in the right inguinal region. Computed tomographic imaging of the abdomen revealed a dilated appendix within a right-sided indirect inguinal hernia. The edematous appendix measured 1.1 cm in diameter, narrowing at orifice of inguinal canal, and was accompanied by a fluid collection.

**Discussion**
An inguinal hernia sac containing a vermiform appendix is known as Amyand’s hernia. Claudius Amyand, sergeant-surgeon to King George II of England, is credited by some with performing the first documented successful appendectomy, in which he removed a perforated appendix from a right inguinal hernia sac in 1735 (1).

Amyand’s hernia is classically described to account for 1% of inguinal hernias and 0.1% of cases of appendicitis. In the pediatric population, Amyand’s hernia is about 3 times more common, and its prevalence can actually reach 1%, a difference probably based on certain anatomic characteristics. Amyand’s hernia is, as expected, more common in male patients and presents a bimodal age distribution in neonates and in patients aged 70 years (2).

Preoperative diagnosis of Amyand’s hernia is feasible with US and CT. CT allows direct visualization of the appendix inside the inguinal canal (3).

Classical treatment of Amyand’s hernia includes appendectomy, reduction of hernia, and hernioplasty through the same incision (2).

**References**
18/M
Chief complaint: Left posterior ankle pain

Fig. 1. Left foot lateral view  
Fig. 2. Sagittal T1-weighted image  
Fig. 3. Fat suppressed sagittal T2-weighted image
Question
What is the most like diagnosis?
1) Achilles tendon tear
2) Haglund syndrome
3) Os trigonum syndrome
4) Sever disease
5) Achilles insertional tendinopathy

Answer
2) Haglund syndrome

Imaging Feature
Foot lateral radiograph shows osseous prominence of posterosuperior margin of calcaneus, soft tissue fullness in pre-Achilles bursa region. In sagittal T1-, T2-weighted MR images, fluid in pre-Achilles bursa, inflammatory change in pre-Achilles and retro-Achilles fat, calcaneal bone marrow edema. Insertional Achilles tendinopathy shows streaks of increased signal intensity not as bright as fluid.

Brief Reviews
Definition
* Hanglund deformity: Enlarged superior margin of calcaneal posterior process
* Haglund triad (Haglund syndrome): Haglund deformity + pre-Achilles (retrocalcaneal) bursitis + Insertional Achilles tendinosis

Haglund syndrome is a common cause of posterior heel pain, characterized clinically by a painful soft tissue swelling at the level of Achilles tendon insertion. Generally, this condition is secondary to chronic bony deformity, along with the shoes in question, may lead to a mechanically induced inflammation of the superficial bursa, Achilles tendinosis, and retrocalcaneal bursitis. These inflammatory changes in the presence of the bony deformity are together known as Haglund syndrome. Hindfoot varus and pes cavus are predisposing factors. Most important is the presence of chronic stress. On lateral radiograph, the Haglund deformity is manifested by a bony projection of posterosuperior aspect of calcaneal tuberosity (the radiographic equivalent of the “pump bump”). Soft tissue fullness in pre-Achilles bursa region, soft tissue infiltration in Kager’s fat pad, Achilles tendon swelling with ill-defined margin, and retro-Achilles soft tissue swelling may be seen in lateral radiograph. In MRI, bone marrow edema on posterior calcaneus including osseous bump area, pre-Achilles or retro-Achilles bursitis, edema in Kager’s fat pad, Achilles peritendinitis, and Achilles insertional tendinopathy (streaky areas of increased signal intensity within thickened tendon) may be seen.
References

2. Lawrence DA, Rolen MF, Morshed KA, Moukaddam H. MRI of Heel Pain. AJR 2013;200:845-855
M/1 day
CC: abnormal finding on fetal US

Question
What is your diagnosis?
1) Bronchial atresia
2) Pulmonary Sequestration
3) Pleuropulmonary blastoma
4) Congenital Lobar Overinflation (CLO)
5) Congenital Pulmonary Airway Malformation (CPAM)

Answer
5) Congenital Pulmonary Airway Malformation (CPAM)
**Imaging Features**

Fetal US (Fig. 1) shows multiple hypoechoic cysts and intervening areas of increased homogeneous echogenicity. Postnatal chest AP radiography (Fig. 2) shows heterogeneous radiolucency in the right lung. Chest CT scans in a lung window setting (Fig. 3) demonstrates the multiple cystic lesions in the right lower lobe. The larger cysts are measured more than 2 cm, surrounded by smaller cysts, suggesting type I CPAM.

**Brief Review**

CPAM is characterized by a heterogeneous group of congenital cystic and noncystic lung masses that communicate with an abnormal bronchial tree lacking supporting cartilage. Type I CPAM is most common with one or more cysts larger than 2 cm in diameter, usually lined by ciliated columnar epithelium. Type II CPAM consists of multiple thin-walled cysts, smaller than 2 cm and lined with ciliated cuboidal or columnar epithelium. Type III CPAM manifests as a bulky, firm mass of glandular-like tissue. Type IV CPAM consists of thin walled large cyst with non-ciliated flattened, alveolar lining cells. The generalized consensus is that symptomatic CPAMs should be removed by lobectomy, but in case of prenatally diagnosed, asymptomatic, small CPAMs, there is a controversy about the timing of resection. There is a potential risk of associated complications, such as infection, pneumothorax, and the small risk of malignant transformation, particularly in the case of CPAM type 1.

**References**

Case 10.

Breast by Kyu Ran Cho

F/45
Chief Complaint: Palpable lump in the right breast

Fig. 1. Mammography (MG).
Fig. 2. Ultrasonography (US).

Fig. 3. MRI (T2 Weight image).
Fig. 4. MRI (Early subtraction MPR image).
Question
What is the most probable diagnosis?
1) Phyllodes tumor
2) Pseudoangiomatous stromal hyperplasia
3) Metaplastic carcinoma
4) Mucinous carcinoma
5) Tubular carcinoma

Answer
4) Mucinous carcinoma

Imaging features
Mammography (Fig. 1) shows heterogeneously dense parenchymal pattern. A 2 cm sized lobular circumscribed and partially microlobulated isodense mass is seen at the inner portion of right breast, which is area where the patient complained of palpable lump. No associated calcifications is noted within the mass. Ultrasonography (Fig. 2) demonstrates a relatively circumscribed and partially microlobulated isoechoic mass with posterior acoustic enhancement. MRI of T2-W1 (Fig. 3) depicts well-defined smooth margined round mass with bright high signal intensity and this shows peripheral rim enhancement on early subtraction MPR image (Fig. 4). On time-intensity curve (Fig. 5), this is a gradually enhancing mass (type I curve). Ultrasound-guided biopsy and subsequent surgery revealed mucinous carcinoma.

Brief review
1. Clinical characteristics
   1) About 2% of all invasive breast cancer
   2) Slow growth rate, a favorable prognosis
3) Age-related: 7% (> 75 yrs) / 1% (< 35 years)
4) If palpable, tend to manifest as soft masses

2. Radiologic findings
   1) Mammography
      -Manifests as a low-density, relatively well-defined or microlobulated oval or lobular mass
      -Ill-defined rather than sharply defined on spot compression views
   2) Ultrasoundography
      -Circumscribed or microlobulated margins / Isoechoic or hypoechoic
      -Presence of posterior enhancement on US (46%)
      -Vascularity (33%)
   3) MRI Morphology
      -Lobular, oval or round shaped, well-defined and smooth margined
      -Very high SI on T2WI compared with other histologic types of invasive ductal carcinoma due to the watery nature of mucin

3. Histology
   1) Pure mucinous carcinoma (> 90% of mucin contents)
      better prognosis, lower incidence of axillary LN metastasis
   2) Mixed mucinous carcinoma (< 90% of mucin contents),
      75% of mucinous carcinoma of the breast

References
2. Lam WW, Chu WC, Tse GM, Ma TK. Sonographic appearance of mucinous carcinoma of the breast. AJR 2004;182:1069-1074