Surgical Pathological Conference

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Case 1

- Name: OOO
- Chart number: OOOOOOO
- Age: 65 year-old
- Gender: female
- Date of admission: 2013/04/01

Chief Complaint

- Epigastric dull pain after lot of intake for 30+ years

Present Illness

- This patient has constipation, hypothyroidism and hyperlipidemia under Meta OPD follow up.
- Abdomen sono revealed gallstones since 2005.
- Abdomen sono on 2013-01-08 showed gallstones, **echogenicity 1.3x1.3 cm at fundus of gall bladder**. DDX: chronic cholecystitis, neoplasia
- She was referred to GI OPD and abdomen CT scan was arranged.

Present Illness

- CT findings on 2013-3-13:
  - A lobular heterogeneous low attenuate lesion (4.7 X 3.9 X 3.7 cm) in the spleen
  - Suspicious small contracted gallbladder with gallstones.
- Due to gallstones with intermittent epigastric dull pain and splenic tumor, she went to our GS OPD for help and was admitted for surgical intervention.

- **Past history:**
  - Hypothyroidism on Eltroxin
  - Constipation
  - Hyperlipidemia
  - s/p hysterectomy
- **Personal history:**
  - Smoking: 1 package/day for 30+ years
  - Alcohol: nil
- Allergy: NKA
- Family: no pertinent family history
- Travel history: nil

Physical Examination

- General appearance:
  - Fair-looking
  - Alert consciousness
- HEENT:
  - Sclera: not icteric
  - Conjunctiva: not anemic
- Chest:
  - Smooth breath pattern
  - Coarse breath sounds
- Abdomen:
  - Soft and flat, no tenderness, no palpable mass, no Murphy's sign
- Extremities:
  - No muscle weakness, no cyanosis, no pitting edema
- Skin:
  - No ecchymosis, no rash

Lab Data

項目名稱	判斷	結果值	單位	參考值範圍
CBC				
WBC		7.4	10^3/uL	4.0 - 10.0
RBC		4.54	10^6/uL	3.70 - 5.50
HGB		13.6	g/dL	11.3 - 15.3
HCT		41.4	%	33.0 - 47.0
MCV		91.2	fL	80.0 - 100.0
MCH		30.0	pg	25.0 - 34.0
MCHC		32.9	g/dL	30.0 - 36.0
PLT		351	10^3/uL	130 - 400
DIFF				
NEUT%		56.4	%	40.0 - 75.0
LYMPH%		35.9	%	20.0 - 45.0
MONO%		3.9	%	2.0 - 10.0
EO%		3.3	%	1.0 - 6.0
BASO%		0.5	%	0 - 1
Prothrombine time				
Prothrombine time		10.7	sec	8.0 - 12.0
MNPT		10.7	sec	
PT INR		1.00		0.85 - 1.15
APTT				
APTT		29.6	sec	23.9 - 35.5
APTT control		28.0	sec	

Lab Data

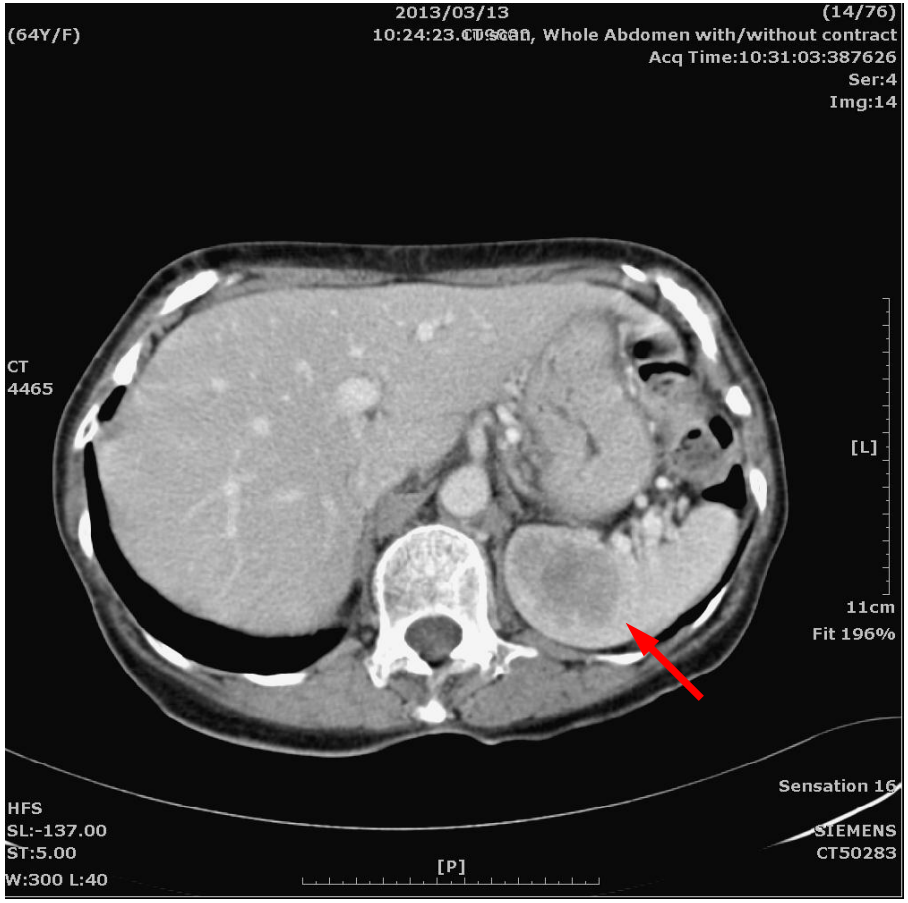
項目名稱	判斷	結果值	單位	參考值範圍
Glucose AC		78	mg/dL	70.0 - 110.0
BUN		18.7	mg/dL	8.0 - 20.0
Creatinine		0.83	mg/dL	0.44 - 1.27
eGFR		69		> 60
AST		29	IU/L	5 - 50
ALT		23	IU/L	5 - 50
Na		142	mmol/L	136 - 144
K	L	3.4	mmol/L	3.6 - 5.1
Cl		103	mmol/L	101 - 111

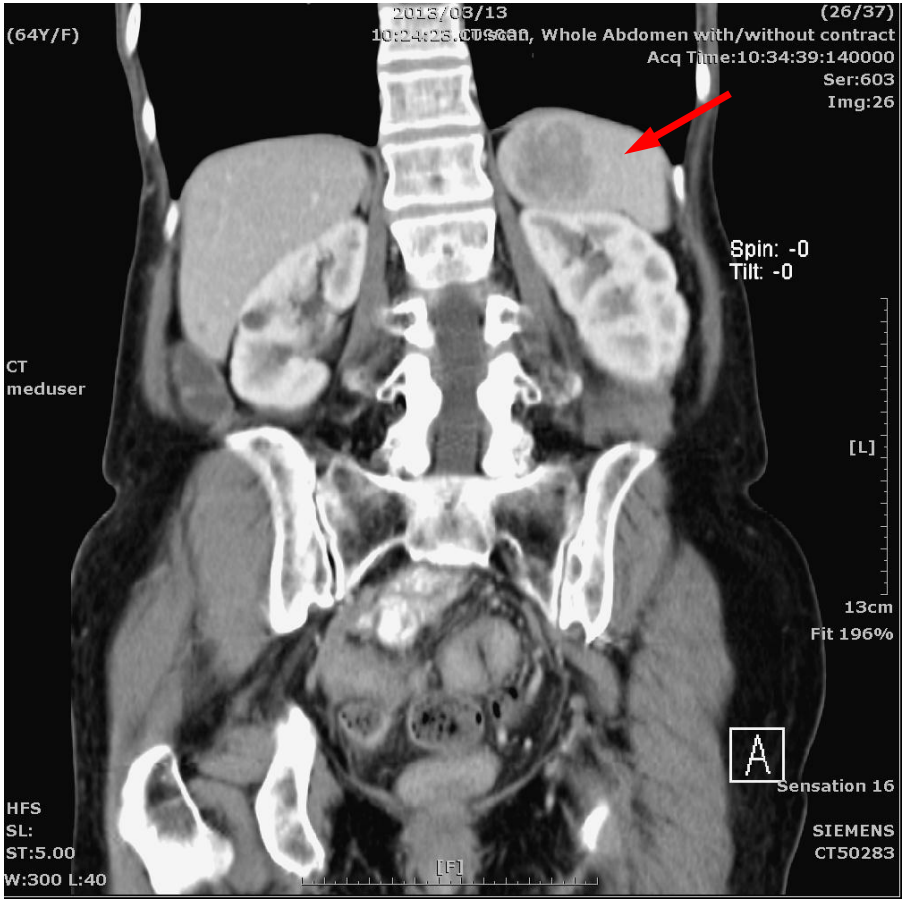
項目名稱	判斷	結果值	單位	參考值範圍
CEA		3.52	ng/mL	
備註 : Nonsmoker(20-69yrs)<=3.8;Nonsmoker(>=40yrs)<=5.0				
AFP		3.36	ng/mL	0.00 - 20.00
CA19-9		12.81	U/mL	0.00 - 27.00

Abdomen CT

2013-03-13

A lobular heterogeneous low attenuate lesion (4.7 X 3.9 X 3.7 cm) in the spleen



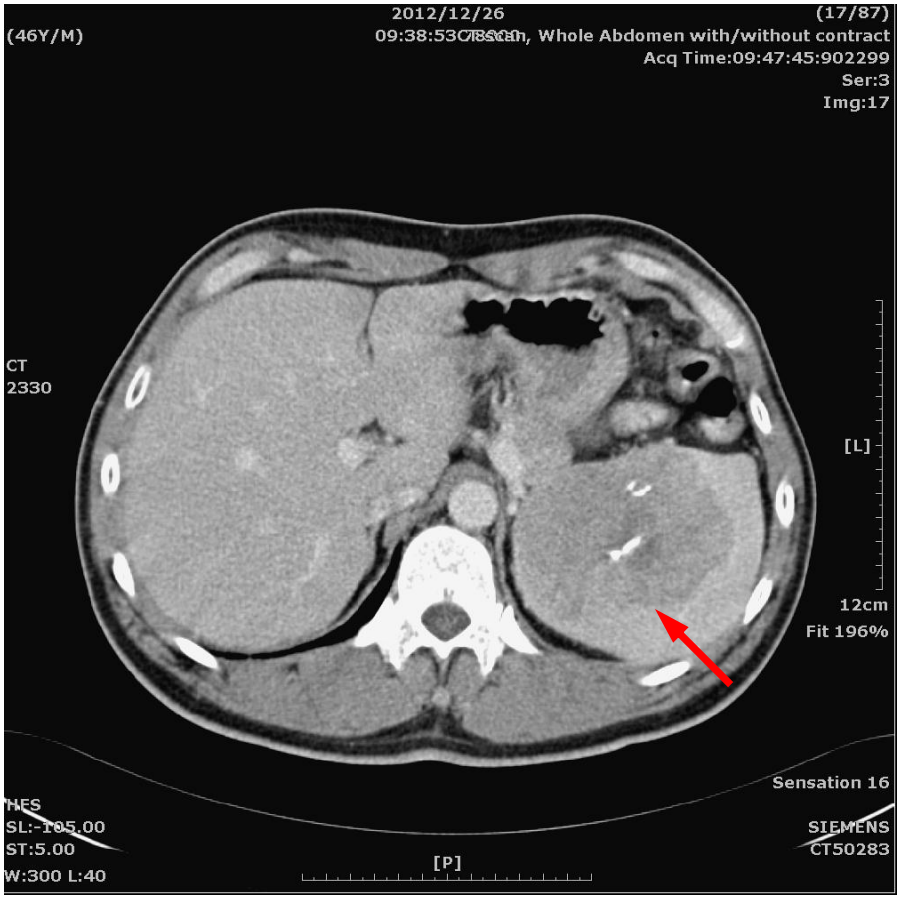


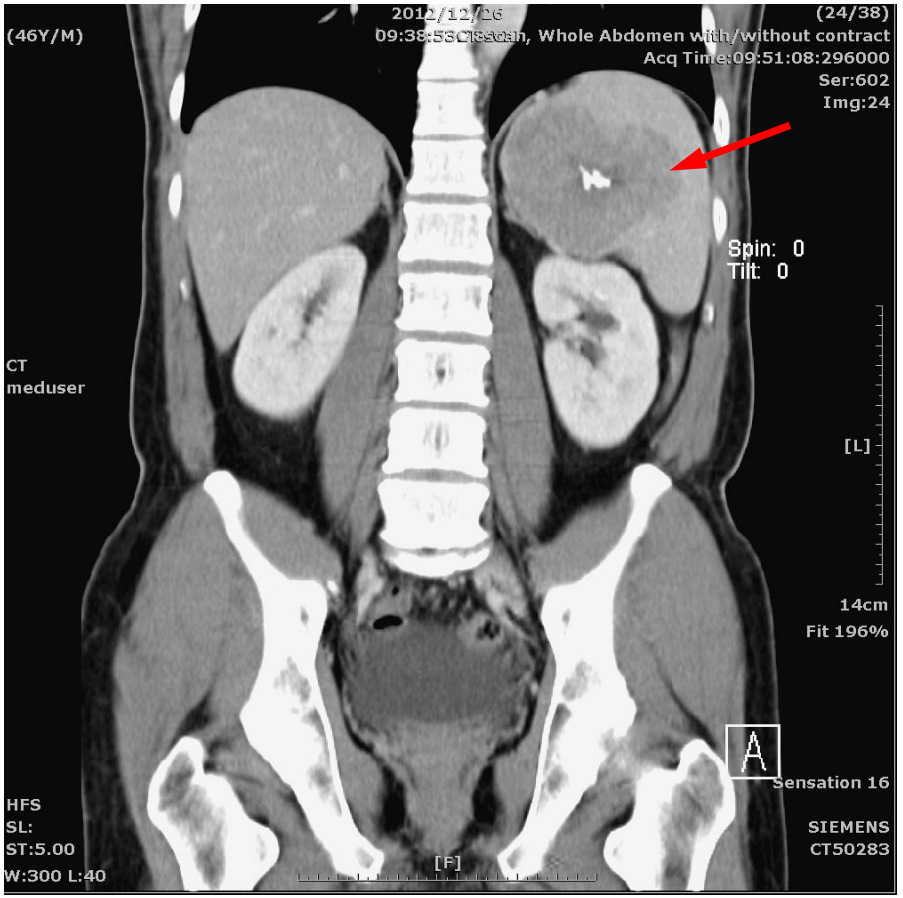
Case 2

- 46-year-old male
- He was well in the past
- A spleen tumor was found accidentally by health check up at LMD

Abdomen CT

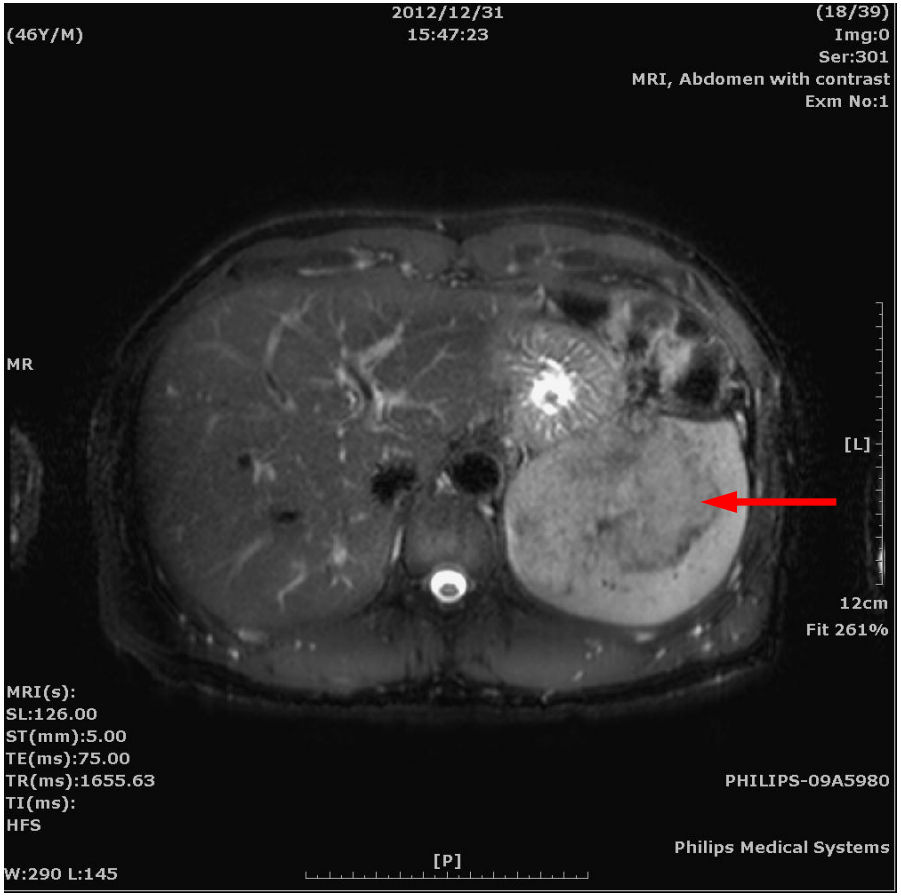
A well-defined lobular contour heterogeneous contrast enhanced lesion (8.7 X 7.1 X 6.1 cm) with internal calcifications in the spleen

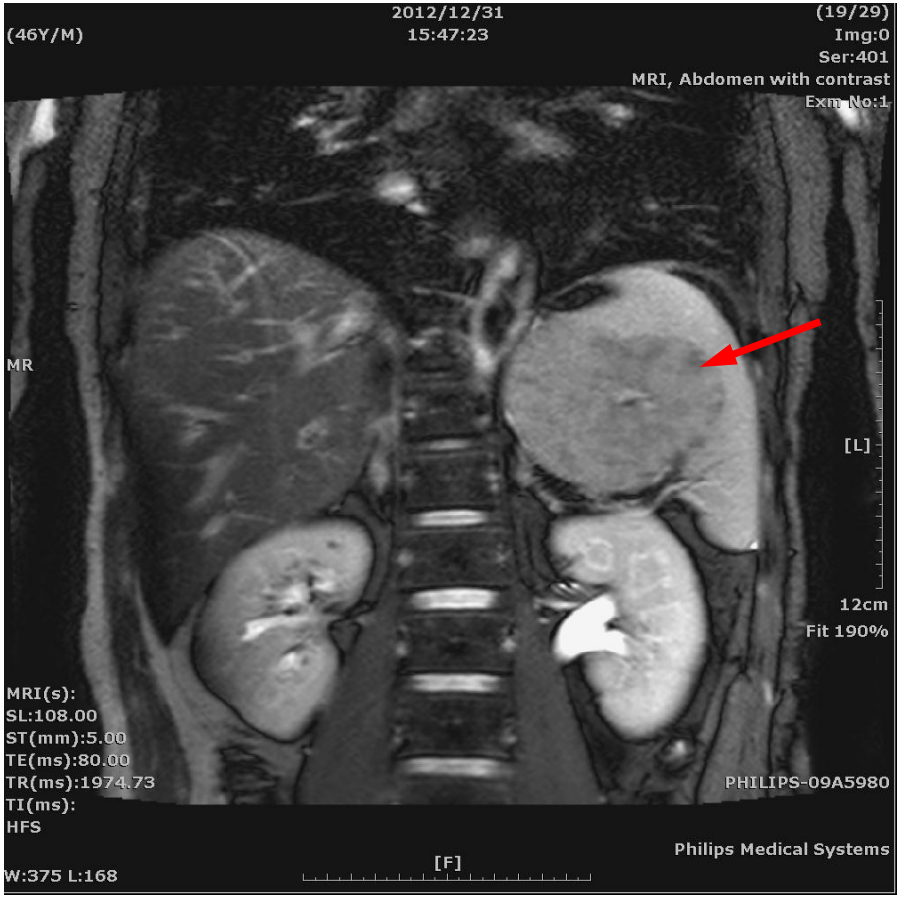




Abdomen MRI

Well-defined, delayed homogeneous enhanced lesion (9x7x6 cm) in the spleen





Splenic Mass

- A **splenic tumor** is a rare form of tumor that may be malignant or benign.
- Benign
  - Cysts
  - Abscess
  - Inflammatory mass
  - Haemangiomas, litteral cell angioma, lymphangioma
  - Hamartoma
- Malignant
  - Lymphoma
  - Sarcoma
  - Metastasis

Splenic cysts

- Primary (true) cysts
  - Parasitic: echinococcal
  - Non-parasitic: congenital (i.e. epithelial) and neoplastic (lymphangioma, metastases, haemangioma) cysts
- False cysts
  - Develop secondary to trauma
  - Haemorrhage, infarction-degeneration and inflammation

Haemangiomas

- The most common benign tumor
- They are usually asymptomatic and found incidentally
- On CT:
  - Capillary haemangiomas appear as small, well-marginated homogeneous iso- or hypodense masses with homogeneous contrast enhancement.
  - Cavernous haemangiomas, usually of larger dimensions, appear more or less cystic with occasional iso- or hypodense areas and after the injection of contrast medium, they demonstrate early peripheral nodular enhancement with progressive fill-in and are homogeneous on delayed images, although they do not exhibit this typical enhancement pattern. Calcifications if present may be either peripheral and curvilinear or scattered centrally.

Hamartoma

- Rare benign tumor of the spleen
- Hamartoma is composed **red pulp components**, but may also contain cystic or necrotic components and **small calcifications**.
- CT:
  - Hamartomas appear as well-demarcated, solid, hypodense masses. It demonstrates inhomogeneous and moderate contrast enhancement.
- MRI:
  - A well-defined homogeneous mass which is isointense on T1-weighted images and slightly hyperintense on T2-weighted images.
  - Dynamic enhanced T1-weighted images depict diffuse heterogeneous enhancement early after injection of contrast medium and more uniform enhancement is seen on delayed images.

Littoral cell angioma

- A distinct new clinico-pathological entity of a very rare benign tumor of the spleen
- It develops from the lining cells of the red-pulp sinuses.
- CT:
  - Multiple hypoattenuating masses in an enlarged spleen which on histopathologic examination, represent blood-filled vascular channels. After injection of contrast medium, minimal delayed enhancement is seen. In contrast to typical haemangioma, the internal morphology of LCA is inhomogeneous, and lesion distribution is also diffuse.
- MRI:
  - Inhomogeneous hyperintense on T2-weighted MR images, with signal similar to that of haemangiomas and slightly hypointense on unenhanced T1-weighted images.
  - Dynamic enhanced T1-weighted images depict delayed contrast enhancement, suggestive of a vascular lesion with contrast media pooling

Lymphangioma

- Lymphangioma is relatively uncommon and frequently presents as a solitary splenic nodule or as a part of systemic lymphangiomatosis in young patients;
  - Both types are grossly cystic in most cases.
  - A solid lymphangioma with sclerotic change and papillary endothelial proliferation are also described.
- Histologically it is classified into three subtypes: simple (capillary), cavernous and cystic.
- **Cystic lymphangioma** is the most frequent type and is characterised by a honeycomb of large and small thin-walled cysts containing lymph-like clear fluid.
- Because the lesions are cyst-like, their appearance on ultrasound, CT, and MRI is similar to and indistinguishable from that of cysts.

Inflammatory pseudotumor

- **Plasma cell granulomas** and **pseudosarcomatous myofibroblastic proliferations**
- Some are **infection-associated** (latent infection by the Epstein–Barr virus), and some are **neoplastic**.
- CT:
  - Inflammatory pseudotumour appears as a hypodense mass with delayed enhancement and there may be a central scar without enhancement corresponding to collagen fibres around vessels.
- MRI:
  - Hypointense on T1-weighted images and hyperintense on T2-weighted images and following contrast medium there is inhomogeneous delayed enhancement similar to that previously described for hamartomas.

Lymphoma

- The most common splenic malignancy
  - non-Hodgkin lymphomas
- Staging of lymphomas on CT can be limited as only 45%–70% of lymphomas show diffuse splenic infiltration or tumor foci less than 1 cm in diameter so that the diagnosis of lymphoma can sometimes only be made microscopically. The focal lesions with diameter from 1 to 10 cm are typically of low attenuation and rarely enhance so may be better demonstrated on post-contrast scans.
- MRI:
  - Non-specific and similar to those of metastases from other primary tumors. Typically, lymphomas are hypointense or nearly isointense on T1-weighted images and hyperintense on T2-weighted images. Injection of contrast medium may improve detection of splenic lymphoma.

Metastases

- It is an infrequent site for metastatic disease (3.4% of metastatic carcinoma)
- Explanations proposed for the **relative paucity of splenic metastases** have included:
  - The sharp angle made by the splenic artery which makes it difficult for tumor emboli to enter the spleen.
  - The rhythmic contractile nature of the spleen which squeezes out the tumor emboli.
  - The absence of afferent lymphatics to carry metastatic tumor to the spleen.
  - Antitumor activity due to a high concentration of lymphoid tissue in the spleen.
- Splenic metastases are most commonly found in **malignant melanoma**, **lung**, **breast** or **ovarian carcinomas**

Metastases

- CT:
  - Splenic metastases typically appear as hypodense lesions which may be solid or cystic and with inhomogeneous contrast enhancement indicating a mixture of vascularisation or necrosis.
- MRI:
  - Metastases are predominantly hypointense on T1-weighted images and hyperintense on T2-weighted images, with occasionally inhomogeneous contrast enhancement.
  - MRI is more accurate for the detection of splenic metastases which are necrotic or haemorrhagic.

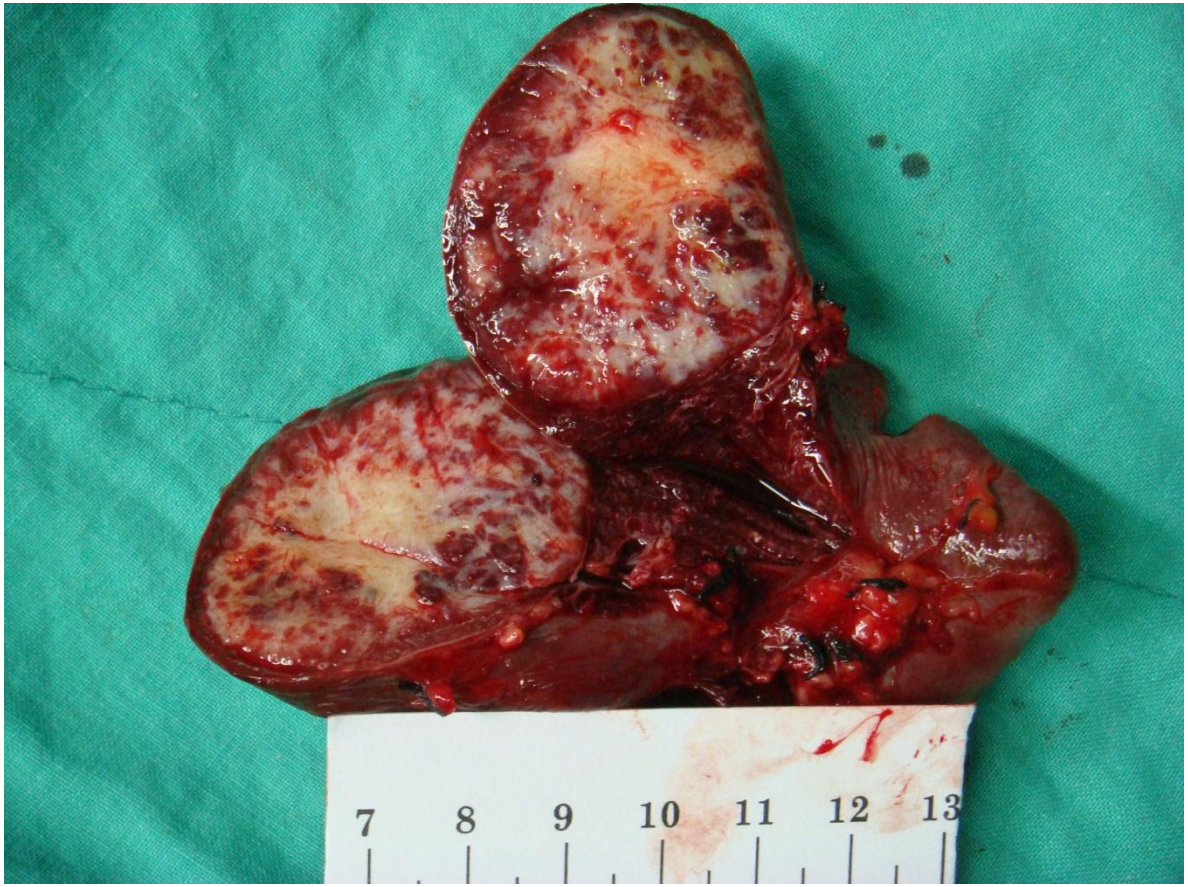
Haemangiosarcoma

- Only 1%–2% of all soft tissue sarcomas, and is highly aggressive with poor prognosis
- **Haemorrhagic areas** that are often present, and which suggest how tumor growth occurred over **a short period of time**.
- CT:
  - Splenomegaly with a poorly circumscribed, inhomogeneous mass of mixed attenuation with poor contrast enhancement on CT. Occasionally intratumoral necrosis and subcapsular or extracapsular blood collection can be seen because haemangiosarcoma may bleed and cause spontaneous rupture of the spleen.
- MRI:
  - MR dynamic images clearly depict heterogeneous enhancement within the tumor, which corresponds to the pathologic findings of solid parenchyma with necrosis.

Differential diagnoasis

- Hamartoma
- Inflammatory pseudotumor
- Lymphoma
- Metastases
- Haemangiosarcoma

Operation on 2013/04/03



A 4.5*3.5*3.0 cm firm tumor over upper pole of spleen

Pathological Daignosis

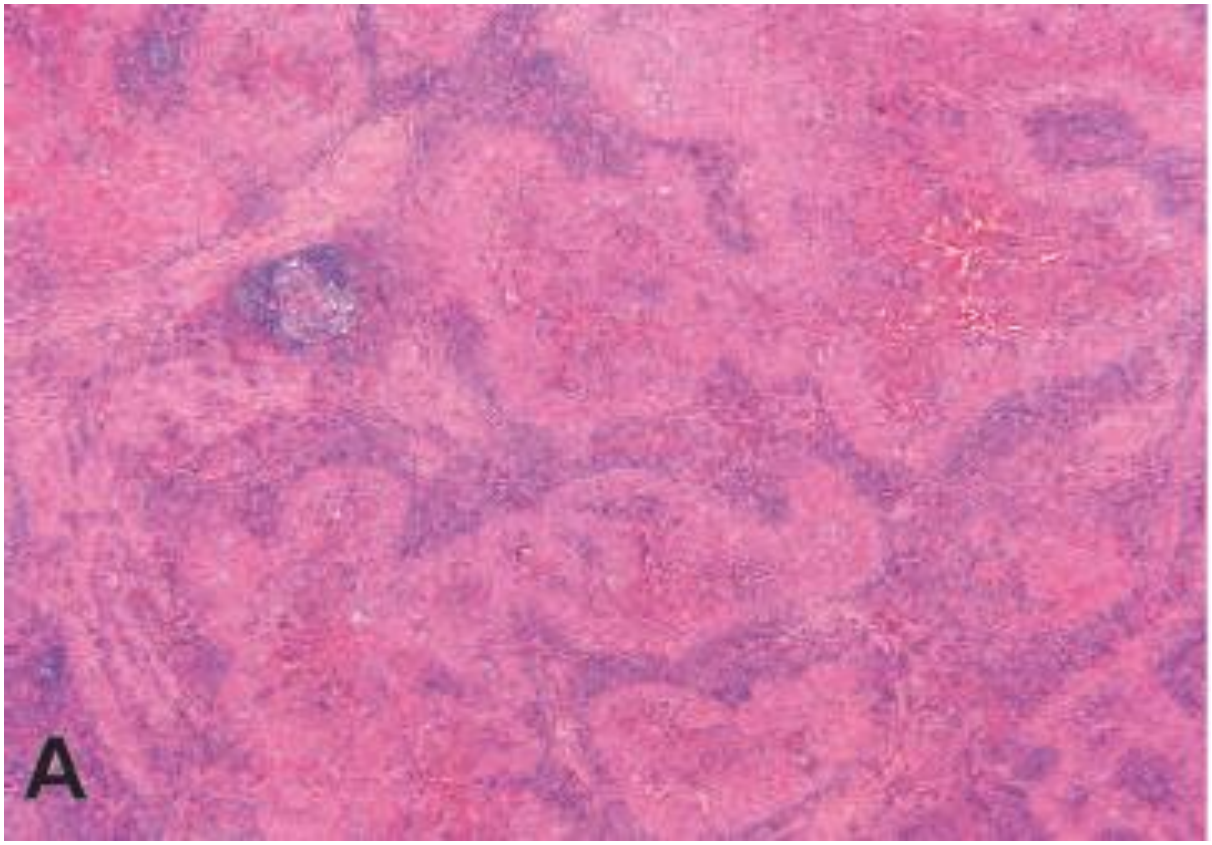
- Spleen ---
  - sclerosing angiomatoid nodular transformation (SANT)

Sclerosing Angiomatoid Nodular Transformation (SANT)

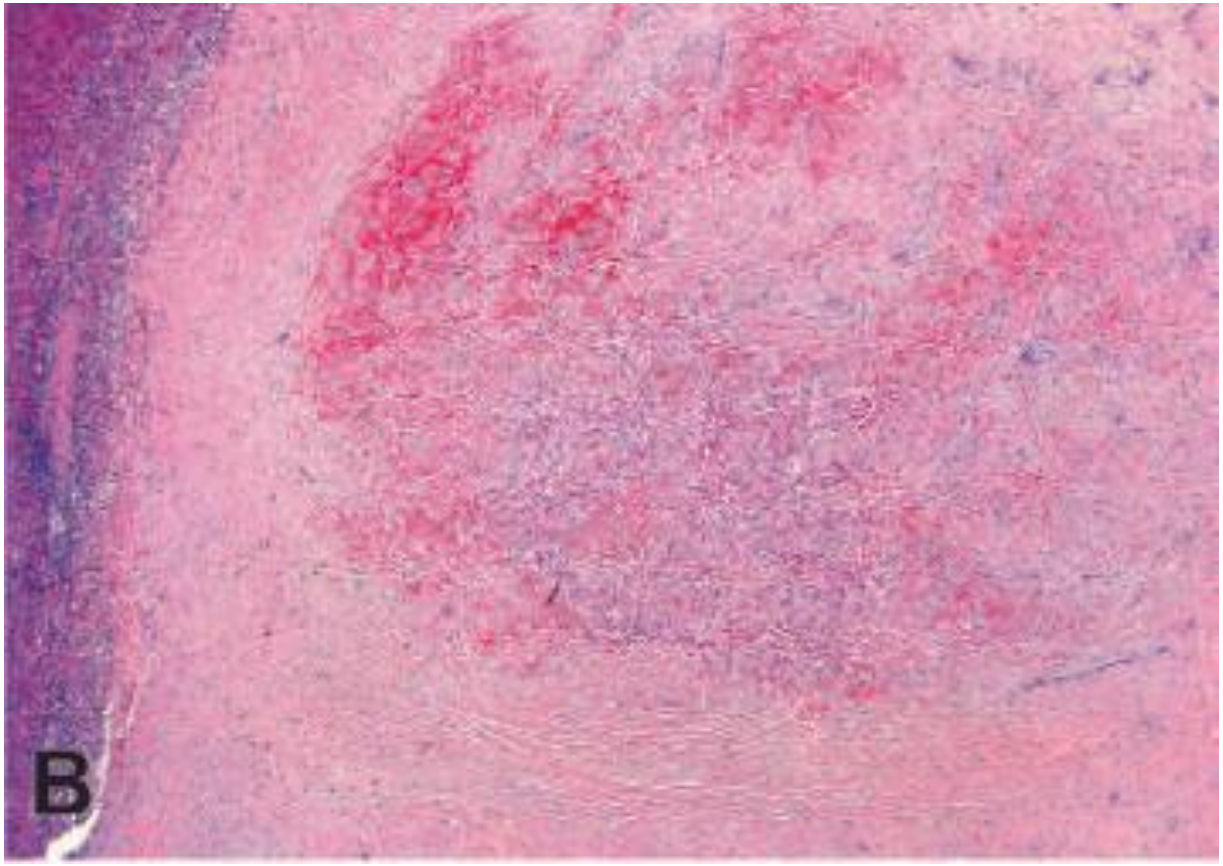
- A recently recognized nonneoplastic vascular lesion of the spleen by Martel et al in 2004
- SANT consists of:
  - Multiple well-circumscribed vascular/angiomatoid nodules showing plump endothelial cell and extravasated erythrocytes.
    - A mixture of sinusoidal, capillary, and veinlike vessels with complex endothelial phenotypes resembling splenic sinusoids
  - The nodules are surrounded by a variable lymphoplasmacytic infiltrate, spindle cells, and collagenous stroma.

Sclerosing Angiomatoid Nodular Transformation (SANT)

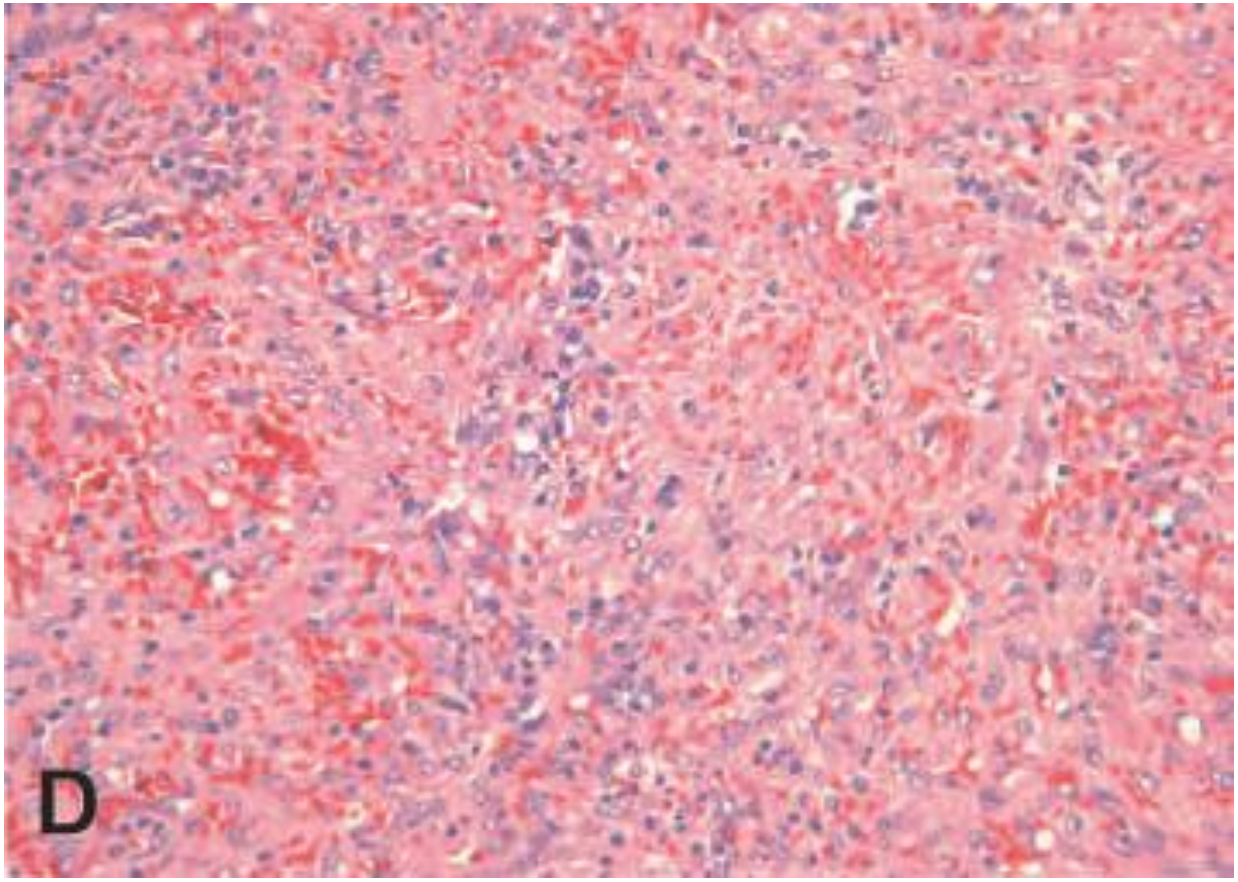
- Sclerosing angiomatoid nodular transformation may also represent a peculiar form of splenic hamartoma because it is composed of **red pulp tissue**.
- SANT had been previously described as ***splenic hamartoma***, ***multinodular hemangioma*** and ***hemangioendothelioma.***
- Most patients were asymptomatic and the splenic mass was an incidental finding during laparotomy or during imaging studies for unrelated conditions.
- Imaging studies usually reveal a hypodense, multinodular splenic mass.



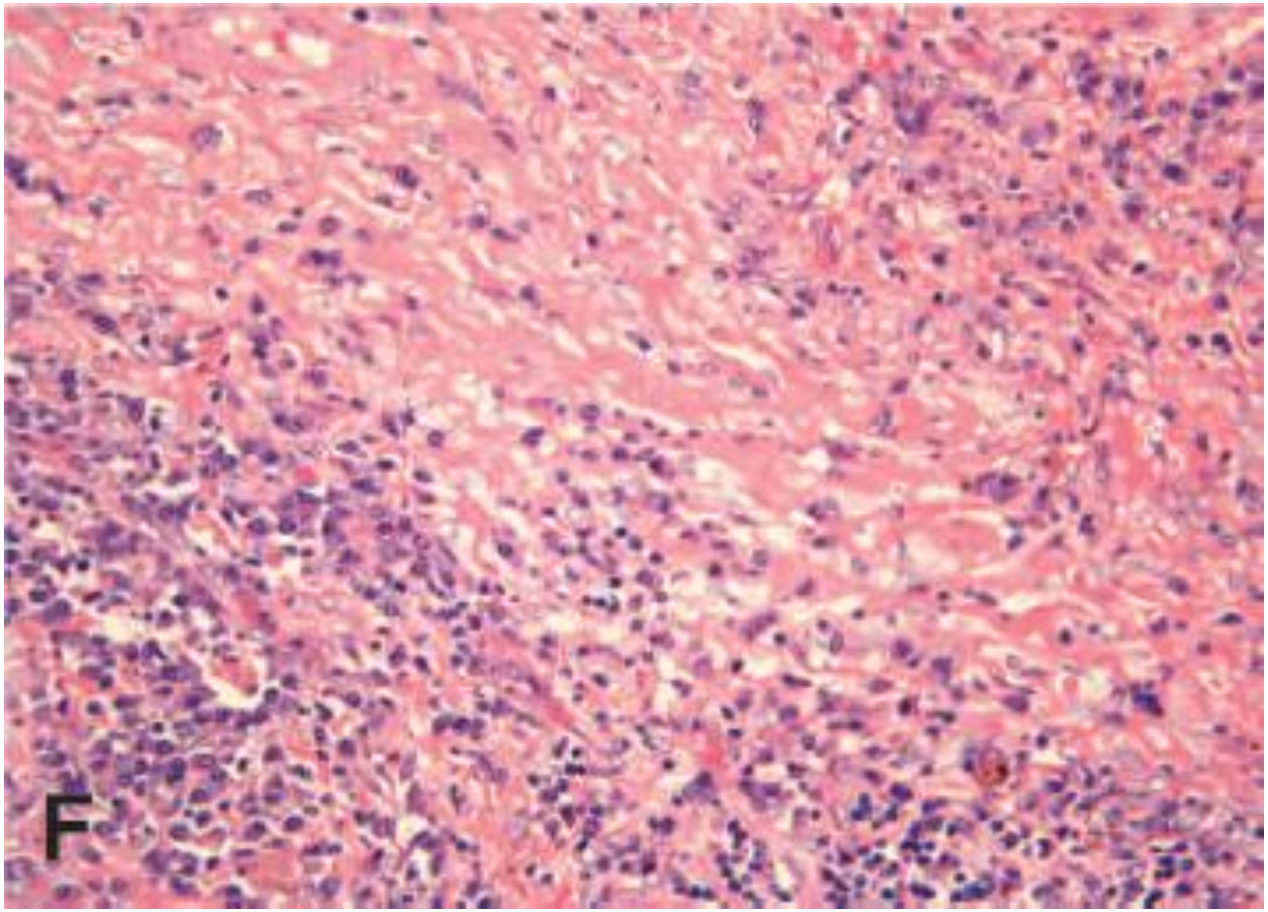
Multiple individual and confluent nodules



The nodules are surrounded by a hypocellular densely sclerotic tissue



The nodules are composed of slitlike vessels with plump endothelial cells with numerous red cells



The internodular areas of SANT show numerous plasma cells, lymphocytes, and siderophages with a variable fibromyxoid or sclerotic stroma

Immunohistochemical analysis

- SANT demonstrates a complex heterogeneous mixture of blood vessels recapitulating normal red pulp with different patterns of expression for:
  - CD8
  - CD21
  - CD31
  - CD34
  - Smooth muscle actin

- **Splenectomy** is usually performed on the discovery of a splenic mass and appears to be curative in all reported cases.

Thanks for your attention!