

Case Presentation

主治醫師：宋文鑫

日期：2015-2-28

General Data

- Name : 000
- Chart Number : 00000000
- Date of Admission : 2014 年 08 月 04 日
- Age : 33 y/o
- Sex : female
- Occupation : 會計

Chief Complaint

- Palpable soft tissue mass on posterior neck for 3 months

Present Illness

- She complained palpable soft tissue mass on posterior neck for 3 months. She came to OPD for help. No obvious neurologic deficit, nor local tenderness.
- A head & neck CT which showed 34*34*50mm soft tissue mass with enhancement is noted in posterior neck from suboccipital to C2-3 interspinous level, compatible with soft tissue tumor.

- Past history:
 - Inflammation of right kidney on 2014-8
- Operation history:
 - Nil
- Personal history:
 - Allergy: NKA
 - Smoking: 2 cigarettes/day for 20 years
 - Alcohol drinking: sometimes
- Family history:
 - Nil
- Travel and contact history:
 - Nil

Physical Exam

- Consciousness: alert. GCS: E4V5M6.
- Sclera: no icterus, Conjunctiva: pinky
- Neck:
 - **soft mass about 8cm in size without tenderness over posterior neck**
 - No lymphadenopathy
- Breast:
 - No palpable mass, no bilateral breast skin or nipple retraction, no bilateral nipple discharge
- Chest:
 - BS: clear
- Abdomen:
 - Soft and flat
 - No hepatosplenomegaly, no palpable mass, no rebounding pain, no Murphy's sign, no shifting dullness
 - Normoactive BS
- Skin:
 - Palpable soft tissue mass on posterior neck.
- Extremities:
 - Free movement, no pitting edema

Lab Data

CBC				
WBC		4.7	10 ³ /uL	4.0 - 10.0
RBC		4.39	10 ⁶ /uL	3.70 - 5.50
HGB		12.6	g/dL	11.3 - 15.3
HCT		38.2	%	33.0 - 47.0
MCV		87.0	fL	80.0 - 100.0
MCH		28.7	pg	25.0 - 34.0
MCHC		33.0	g/dL	30.0 - 36.0
PLT		191	10 ³ /uL	130 - 400
DIFF				
NEUT%		70.9	%	40.0 - 75.0
LYMPH%		24.0	%	20.0 - 45.0
MONO%		4.3	%	2.0 - 10.0
EO%	L	0.4	%	1.0 - 6.0
BASO%		0.4	%	0 - 1
PT				
Prothrombine time		11.0	sec	8.0 - 12.0
MNPT		10.5	sec	
PT INR		1.04		0.85 - 1.15
APTT				
APTT		28.3	sec	23.9 - 35.5
APTT control		28.0	sec	

Lab Data

Glucose AC		87	mg/dL	70 - 110
BUN		10.7	mg/dL	8.0 - 20.0
Creatinine		0.83	mg/dL	0.44 - 1.27
eGFR		79		> 60
Uric acid		4.4	mg/dL	2.6 - 8.0
Total protein		7.3	g/dL	6.1 - 7.9
Albumin		4.5	g/dL	3.5 - 4.8
A/G Ratio		1.6		1.2 - 2.0
Globulin		2.8	g/dL	2.5 - 3.6
Total Bilirubin		1.17	mg/dL	0.4 - 2.0
Direct Bilirubin		0.22	mg/dL	0.10 - 0.50
AST		13	IU/L	5 - 50
ALT		11	IU/L	5-50
LDH		118	IU/L	98 - 192
γ-GT		14	IU/L	7.0 - 50.0
Alkaliphosphohatase		44	IU/L	38-126
Triglyceride		72	mg/dL	50 - 200
Cholesterol, Total		189	mg/dL	< 200
Na		141	mmol/L	136 - 144
K		4.2	mmol/L	3.6 - 5.1
AFP		3.4	ng/mL	≦ 20
CEA		2.2	ng/mL	MRR
B-HCG		<1.2	mIU/mL	≦ 5

Soft tissue mass on posterior
neck

rapid enlargement, painless

Soft tissue mass on neck

- Abscess
- Lymphadenopathy (benign, metastatic)
- Lymphoma
- Salivary gland enlargement
- Thyroid: adenoma, goiter, cyst, carcinoma
- Branchial cleft cyst
- **Soft tissue tumor (lipoma, vascular, neurogenic, sarcoma)**

World Health Organization (WHO) system for classification of soft tissue tumors

- Adipocytic tumors
- Fibroblastic or myofibroblastic tumors
- So-called fibrohistiocytic tumors
- Smooth muscle tumors
- Skeletal muscle tumors
- Vascular tumors
- Perivascular tumors
- Chondro-osseous tumors
- Tumors of uncertain differentiation

Histologic Type	Benign	Intermediate, Locally Aggressive	Intermediate, Rarely Metastasizing	Malignant
Adipocytic	Lipoma and its variants (lipoblastoma, hibernoma, lipomatosis)	Atypical lipomatous tumor, well-differentiated liposarcoma	...	Liposarcoma
Fibroblastic/ myofibroblastic	Fibromatosis colli, myofibroma, giant cell angiofibroma	Desmoid-type fibromatosis	Solitary fibrous tumor, hemangiopericytoma, inflammatory myofibroblastic tumor (inflammatory pseudotumor)	Fibrosarcoma
So-called fibrohistiocytic	Benign fibrous histiocytoma, diffuse-type giant cell tumor (pigmented villonodular synovitis)	...	Giant cell tumor of soft tissues	Malignant fibrous histiocytoma (undifferentiated pleomorphic sarcoma)
Skeletal muscle	Rhabdomyoma	Rhabdomyosarcoma
Smooth muscle	Leiomyoma, angioleiomyoma	Leiomyosarcoma
Vascular	Hemangioma, lymphangioma	Kaposiform hemangioendothelioma	Kaposi sarcoma	Angiosarcoma
Perivascular	Glomus tumor, myopericytoma	Malignant glomus tumor
Chondro-osseous	Soft tissue chondroma	Mesenchymal chondrosarcoma, extraskeletal osteosarcoma
Uncertain differentiation	Myxoma	...	Ossifying fibromyxoid tumor	Synovial sarcoma, alveolar soft part sarcoma, primitive neuroectodermal tumor, Ewing sarcoma

Neuroectodermal lesions

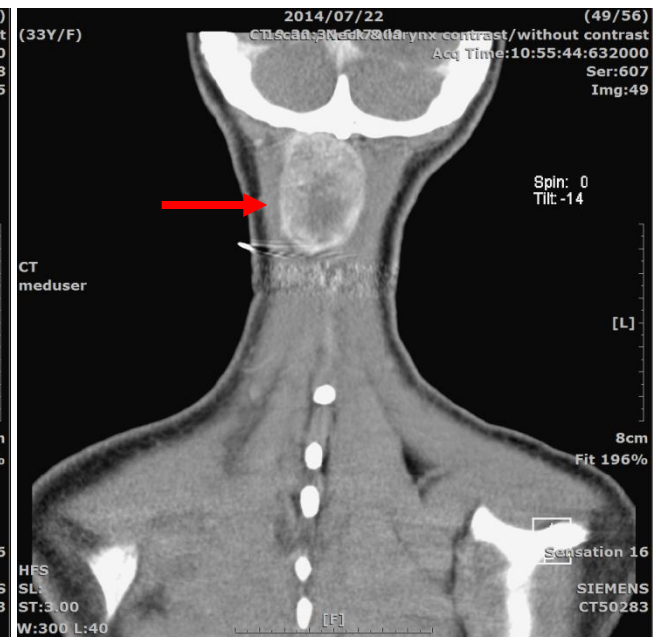
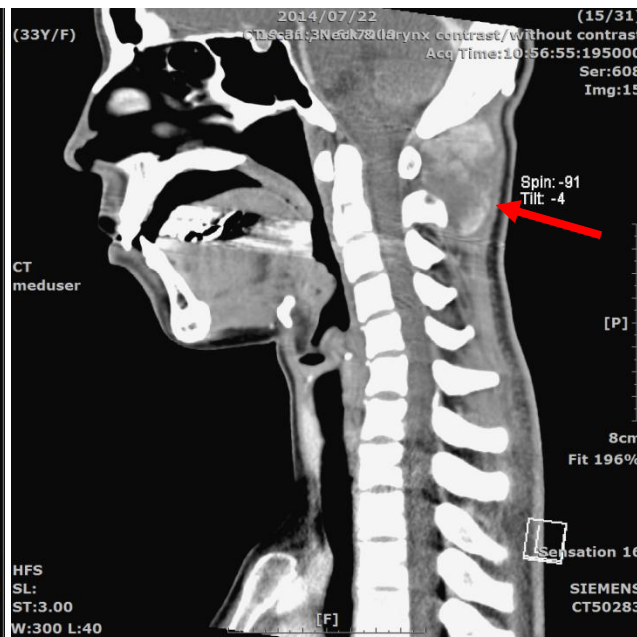
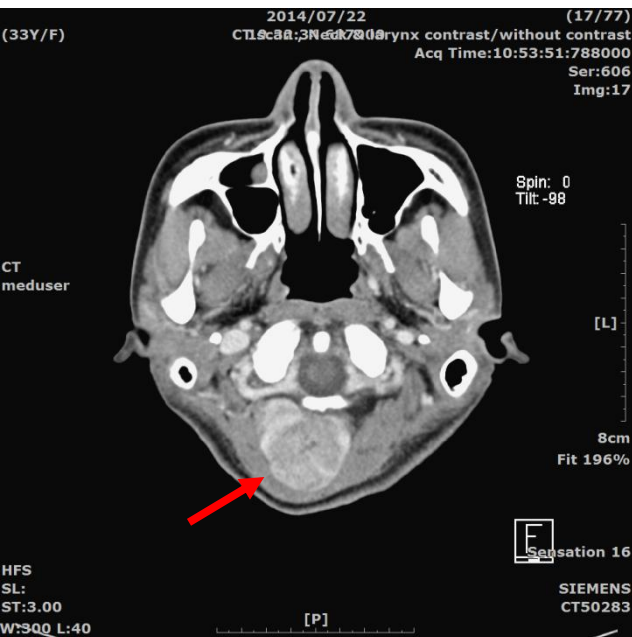
- Schwannomas
- Solitary circumscribed neuromas
- Neurofibromas
- Heterotopic glial (nasal glioma): nasal region
- Ectopic meningotheelial lesions: scalp
 - Ectopic meningotheelial hamartoma
 - Cutaneous meningioma
- Malignant melanoma

Benign or malignancy?

- **Painless, slow-growing, long-standing masses** are likely to be benign
- **Pain, rapid increase in the size** of a mass are worrisome signs of possible malignancy

CT scan

- 34*34*50mm soft tissue mass with **enhancement** is noted in posterior neck from suboccipital to C2-3 interspinous level, compatible with **soft tissue tumor**



Imaging Features Suggestive of Soft Tissue Tumor Malignancy

Large volume

Extracompartmental extension

Poorly defined margins

Broad interface with underlying fascia

Inhomogeneous MR signal intensity

High signal intensity on T2-weighted MR images

Invasion of bone or neurovascular structures

Intratumoral hemorrhage

Intratumoral necrosis

Marked, primarily peripheral enhancement

Soft tissue neck masses have few specific radiographic characteristics

Lipoma

- Approximately **25%** of lipomas occur in the head and neck
 - mostly in subcutaneous locations at the **posterior aspect of the neck.**
- On CT images: attenuation equivalent to that of **subcutaneous fat**

Intense contrast enhancement

- Highly vascularized tumors:
 - Giant cell angiofibroma
 - Hemangiopericytoma
 - Sinonasal glomus tumor
 - Desmoid-type fibromatosis
 - Angioleiomyoma
 - Other vascular soft tissue tumors

In most cases, enhancement characteristics alone cannot be used to reliably distinguish between benign and malignant lesions.

Giant Cell Angiofibroma

- Benign tumor
- **Middle-aged adults** (mean age, 45 years)
- That is most commonly seen in the **orbital region** or **eyelid**
 - may occur in the **buccal mucosa**, **submandibular region**, or **parapharyngeal space**.
- The tumor may **grow rapidly**, simulating malignancy
 - may slow growth over many years
- At CT, giant cell angiofibromas usually appear as **circumscribed, enhancing** masses.

Solitary Fibrous Tumor and Hemangiopericytoma (I)

- Rarely metastasizing tumors with **intermediate malignant potential**.
- Their histopathologic features largely overlap.
- Middle-aged adults
- Slow-growing, **painless**, locally infiltrative masses
- They are occasionally associated with systemic symptoms such as
 - hypoglycemia (because of overproduction of an insulinlike growth factor),
 - arthralgia, osteoarthropathy, digital clubbing.

Solitary Fibrous Tumor and Hemangiopericytoma (II)

- Solitary fibrous tumors arise in the **nasal cavity** or **paranasal sinuses**, **nasopharynx**, **parapharyngeal space**, and **larynx**.
- Hemangiopericytomas most commonly arise in the **pelvic retroperitoneum** and **limbs**
- Both tumors can also arise intracranially from the **meninges**.
- The tumors demonstrate **variable enhancement**, but **enhancement is often intense** because of their **high vascularity**.



Desmoid-Type Fibromatosis (Desmoid Tumor)

- Locally aggressive, fibroblastic lesion with **intermediate malignant potential**
 - poorly circumscribed, with infiltration of the surrounding soft tissues and, often, fixation to underlying muscle or bone.
- There is often an associated history of trauma (30% of cases), and Gardner syndrome (1%–2% of cases).
- The most common sites of involvement being the supraclavicular and neck regions, followed by the face.
- Intense enhancement is a common feature of these tumors.

Angioleiomyoma

- Benign tumors
- Mature smooth muscle bundles surrounding vascular channels
- Women in the 3rd ~ 6th decades of life
- Slow-growing, firm, sometimes painful masses
- Sites of occurrence: oral cavity, lip, auricle, submandibular region, sinonasal cavities, buccal space, larynx, and masticator space
- CT: a well-defined mass containing numerous blood vessels.

Operation on 2014-08-06

- Wide excision of deep intramuscular tumor
- Occipital to C2 longitudinal incision
- Finding :
 1. Huge intramuscular tumor, measured about 10 cm in diameter, located in posterior neck, occiput to C2 level, with **invasion to surrounding muscles** and **to outer cortex of C1,2 laminae**.
 2. The tumor is highly vascular with **abundant blood supply**, and the tumor is **ill-defined without clear margin** with surrounding normal tissues.
 3. The tumor invades into venous plexus on lateral aspect of C1-2, and posterior laminae of C1, C2.
 4. Frozen pathology confirmed intramuscular tumor and could not rule out malignancy.

Pathological Diagnosis:

- Soft tissue, neck, posterior, wide resection
 - **Solitary fibrous tumor**