Clinicopathological Conference

Apr. 12, 2014

Name:
Chart number:
Gender:
Birth:
Occupation: none

Presented by：

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Discussed by：

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Moderator：

病理科	孫建峰醫師

Chief Complain: right neck huge tumor massive bleeding today

Medical History：

This 33y/o male had been diagnosed right neck Dermatofibrosarcoma Protuberans over right neck, stage I (T1N0M0), status post wide excision. He ever received operation three times due to repeated local recurrence before 14 years ago (2 times in NTUH and once in CGMH). The last local recurrent event happened 14 years ago, and he gave up any further treatment after that. The right neck tumor progressively enlarged, but he did not call any help these periods of time. The tumor enlarged to 12*10 cm in size up to now. The tumor was pedunculated with nodular surface. The patient stated he got a traumatic wound over the tumor one week ago (穿衣服撞到). Massive bleeding from the tumor happened on 2012/5/31, and the patient experienced dizziness, cold sweating, shortness of breath and nearly fainting out.

He was sent to local hospital immediately. Hypovolemic shock was noted, and the patient had received fluid resuscitation and blood transfusion with PRBC 2U. Then he was transferred to our ER on 2012/5/31. The bleeding stopped on arrival. Then CT angiography of the Dermatofibrosarcoma over right neck and scalp showed recent intra-tumoral hemorrhage. Multifocal hyper-vascular cutaneous tumors were also noted. The radiologist suggested elective embolization instead of emergent due to no more active bleeding. Plastic surgeon deferred the high risk operation due to hypervascularity of the tumor. Thus, the patient was admitted to our oncology ward for further management.

Past history:

1. Dermatofibrosarcoma Protuberans S/P wide excision 3 times

Personal history:

No other systemic disease. No known food or drug allergy. No hypertension or DM.

Family history: no cancer history

Physical examination: (2012/06/01)

BT:35.3C PR:102/min. R.R.:呼吸:20/min. BP:105/72 mmHg

General appearance: acute on chronic-ill looking; thin status

Conscious: Clear, E 4 V 5 M 6

HEENT: Sclera: not icteric

Conjunctivae: mild pale

**one 1cm nodule, elastic, movable over right orbital angle

NECK: huge mass over right neck, with pedicle, nodular surface,

** no active bleeding, size: 10*12cm

right neck surgical scar

No jugular vein engorgement

Trachea not deviated

CHEST: Breath pattern: smooth, Bilateral symmetric expansion

No use of accessory muscles

Breath sound: bilateral clear, no wheezing; no basal crackles

HEART: Regular heart beat without audible murmur

No audible S3; No audible S4

ABDOMEN: Soft and flat, No superficial vein engorgement

Liver/spleen: impalpable

No tenderness; No rebounding pain, No Murphy`s SIGN

Bowel sound: normoactive

BACK: No knocking pain over bilateral flank area

EXTREMITIES: No joint deformity

Freely movable; No pitting edema

Lab data:

檢驗項目	單位	2012/05/31	2012/06/01	2013/12/19
WBC	1000/uL	13.3		6.6
Hemoglobin	g/dL	6.7	7.9	10.2
Platelets	1000/uL	359		654
Segment	%	88.6		75.0
Lymphocyte	%	7.3		15.8
BUN	mg/dL			4.0
Creatine	mg/dL	0.74		0.55
ALT	U/L	36		11
Na	meq/L	132		135
K	meq/L	4.2		4.0
Cl	meq/L			99

Image studies: to be present:

Pathology: to be present

Course and Treatment:

No more active bleeding was noted after admission. Whole body CT showed diffuse metastasis of the tumor over skin, muscle and lung. The sarcoma tissue taken in 1998 showed poor DNA quality of the sarcoma tissue. Excisional biopsy of the left lower chest wall nodule on 2012/6/18 was performed by GS. PDGFR-A mutation examination showed positive finding, and Imatinib (Glivec) was prescribed for palliative purpose since 2012/6/20. Spontaneous right pneumothorax and blood transfusion reaction complicated the hospitalization course. Under stable clinical condition, he was discharged on 2012/06/25.

The patient came to our Emergency Room due to massive tumor bleeding again on 2012/6/30. Hypovolemic shock was noted with a Hb of 5.4 g/dL on arrival. Blood transfusion, ET intubation and emergent trans-arterial embolism were performed. And the bleeding episode relieved. Sudden onset of aphasia happened on 2012/07/03. No focal weakness nor paresthesia was accompanied. Emergent brain CT scan revealed no positive finding. The neurologic symptom gradually recovered after hydration. Then, he was admitted to oncology ward on 2012/7/8.

After admission, we kept Imatinib use. Frequent tumor bleeding and even massive bleeding resulted in hypovolemic shock happened during wound care. Plastic surgeon deferred the high-risk operation again. Poor prognosis, possible grave outcome and the risk of CVA and tumor necrosis after repeated TAE was explained to the patient and his family. Repeated episodes of massive tumor bleeding with hypovolemic shock happened 7 times between 2012/08/01-10/07, and we arranged TAE 4 times under the patient and family`s agreement. Besides, tumor wound infection with sepsis, fungemia and right pneumothorax complicated during hospitalization. But, all complications resolved after medical treatment.

Unfortunately, upper GI bleeding happened on 2012/11/7, followed by BP drop and respiratory distress. Non-rebreathing mask with O2 supplement, proton pump inhibitor and blood transfusion were given. However, the upper GI bleeding did not stopped. Sudden onset of right neck mass massive bleeding happened again on 2012/11/10. Blood pressure was undetectable and the heart rate was around 160 bpm. We informed the critical condition to his family immediately. They decided to keep conservative treatment. The EKG monitor showed asystole at 13:36 on 2012/11/10. We claimed that this 33-year-old male patient expired in our hospital.

Points of disucssion:

1. Surgical treatment for Dermatofibrosarcoma protuberans
2. Treatment for locally advanced, recurrent, and metastatic Dermatofibrosarcoma protuberans