

Advanced Care Planning: A Case Study

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Case Presentation

27 year old female with PMHx of metastatic axillary synovial sarcoma

CC: SOB

Dyspnea on exertion present for past week, progressively worsening.

Moved in the past month to San Diego from Chicago to “breathe better air”

More recently had dyspnea with ambulation progressing to worsening with even sitting up

Recently started home O2

No fevers/chills/night sweats

ROS + for palpitations, weight loss;

- for cough, wheezing, lower extremity edema, abdominal pain, diarrhea/constipation, nausea/vomiting, dysuria, confusion, headaches

Past Medical History (as per admission H&P)

Stage IV axillary synovial sarcoma with metastases

- s/p 6 cycles of ifosfamide with partial response at MD Anderson as well as RUSH in Chicago. Also had 19 tumor nodules removed from the right lung and 32 nodules removed from the left lung

4.6cm mass in the right atrium

Let's Talk About Synovial Sarcoma!

Primary tumor has good prognosis with resection

Metastatic disease less so: 75% mortality at 2 years. Median survival 11-15 months

Unresectable disease is fatal, chemotherapy for this is palliative

Metastatic pulmonary disease has survival benefit from resection in select cases:

- Age >50, MFH tumors, ≥ 2 metastases
- Metastasis size >2cm
- Metastasis free period ≤ 18 months
- 0 risk factors = 60% 5-year survival with metastatic disease resection
- 1, 2, or 3 risk factors = 30%, 20%, and 0% survival respectively at 5 years

Medications (as per H&P)

- Enoxaparin 60mg SQ q12h
- Naltrexone 3mL PO

Allergies/Social/Family Hx

NKDA

Married, husband at bedside, from Chicago but relocating to San Diego, staying with a friend. Retired from the Marine Corps. No hx of EtOH, tobacco, or drug use.

Mother with ovarian cancer at age 39 in remission

Father without medical problems

Maternal aunt had ovarian cancer

Brother died of brain cancer

Objective Data on Admission

Temp 97.7, HR 127, BP 119/61, RR 18, O2 92% on **15L non-rebreather**

Gen: Ill appearing, NAD

HEENT: No palpable LN

CV: Tachycardic, regular rhythm, normal S1+S2, no m/r/g

Resp: CTAB

Abd: Soft, ntnd, normoactive bowel sounds

Ext: No cyanosis nor edema

Labs in ED

134 | 100 | 9
4.0 | 17 | 0.62 <109

Ca⁺⁺ 9.8

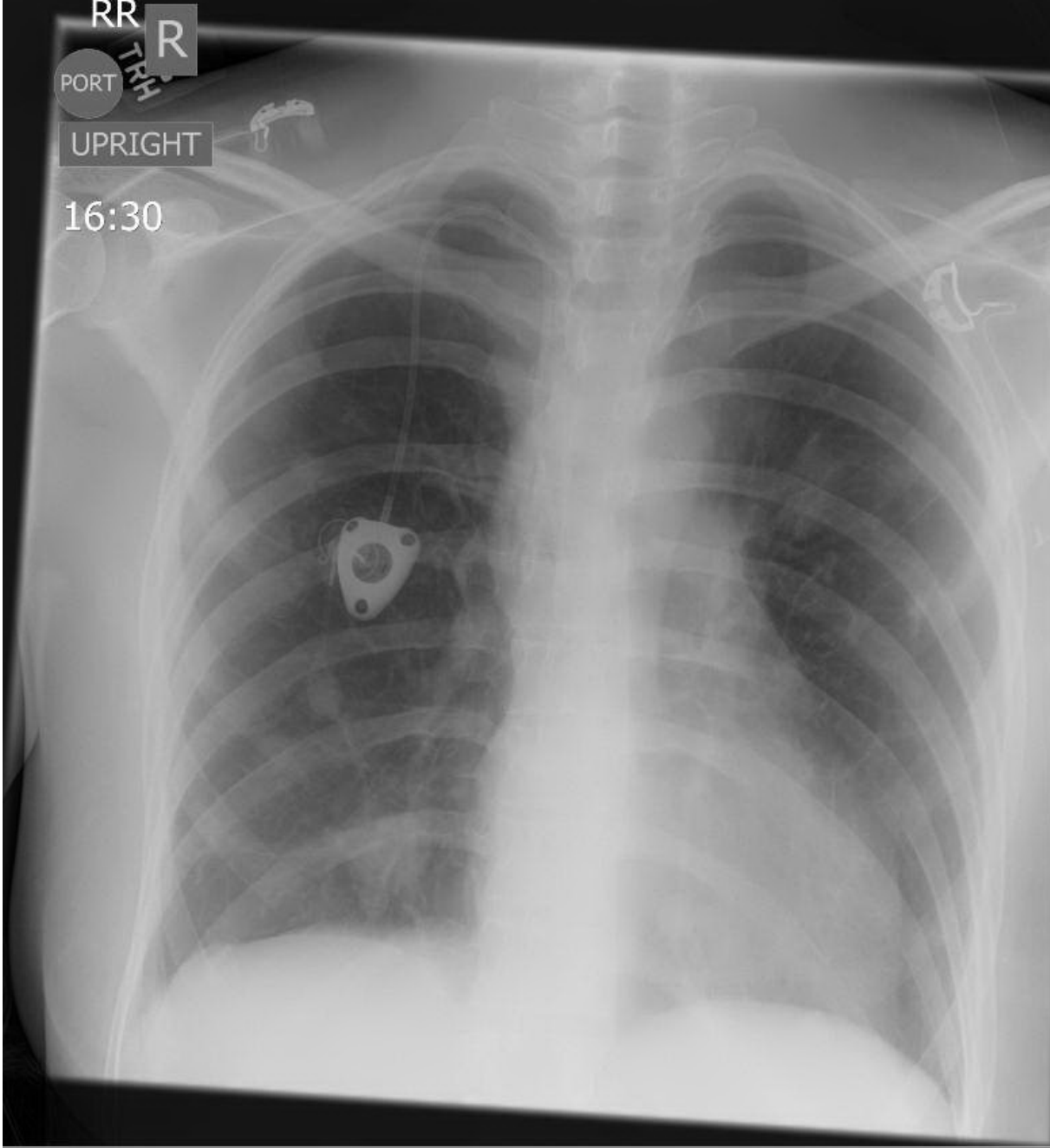
ABG: Not ordered until hospital day 2

BNPP 1057

CPK 84 CKMB-I 1.5 TnT <0.01

8.7 > 15.2 < 296

MCV 84.7
Seg 83%



CXR Read

Right-sided Port-A-Cath followed to the mid superior vena cava.

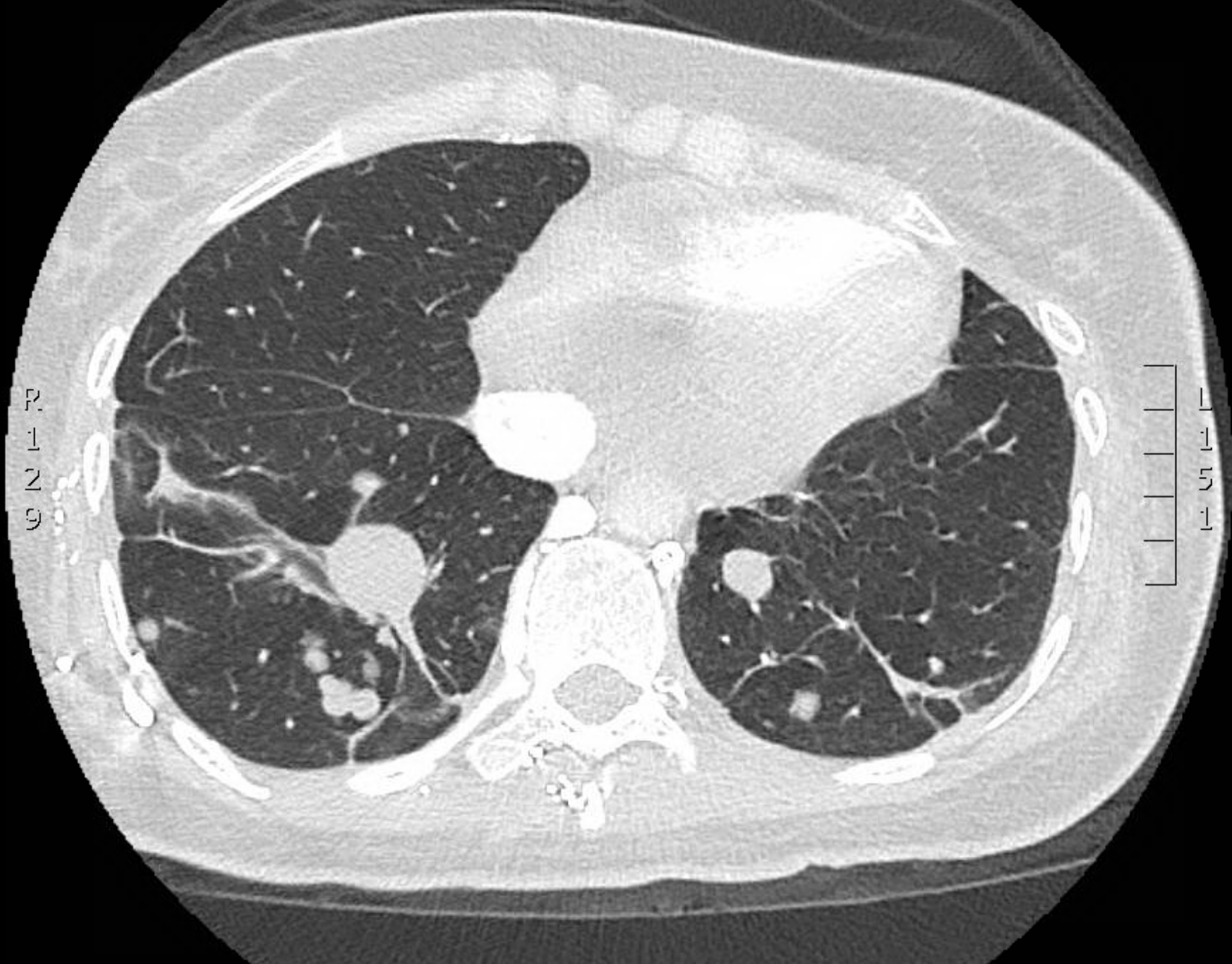
No pleural effusion or pneumothorax demonstrated.

Multifocal bilateral lung opacities, presumably metastatic in nature. A superimposed acute process would be difficult to exclude. Outside priors would be helpful.

Unremarkable cardiac silhouette. Paratracheal/mediastinal fullness --enlarged lymph nodes not excluded.

No acute osseous abnormality identified.

Left axillary surgical clips.







S1879



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6

S1569

CT Chest Read

1. Extensive thrombotic disease within the right atrium extending into the superior vena cava, right internal jugular vein, and innominate vein resulting in vascular collateralization. Cannot exclude bland versus tumor thrombus due to inadequate contrast phase.
2. No evidence of pulmonary embolism.
3. Mass effect of the large right atrial clot on the tricuspid valve, interatrial septum, right pulmonary vein and left atrium. Consider further evaluation with echo to assess for cardiac function.
4. Numerous pulmonary nodules consistent with metastatic disease in this patient with history of cancer.
5. Multiple large masslike lesions in the left axilla consistent with known history of sarcoma. Cannot distinguish lymph nodes from mass due to inadequate contrast timing.

Hospitalization Commence

Step 1: Consult ICU

- ICU consults with attending and fellow, states “**On 15 liters non-rebreather** in ED with SpO₂ >95%. Patient did not desat with trial of positional changes during evaluation. Etiology of shortness of breath and increased oxygen requirements is unclear. CT chest without evidence of pulmonary embolism and there is no evidence of pneumonia on imaging. As noted on CT thorax, **noted to have extensive thrombotic disease within right atrium extending into SVC, right IJ and innominate vein and mass effect of large right atrial clot on tricuspid valve, interatrial septum, right pulmonary vein and left atrium.** Query hypoxia and shortness of breath possibly secondary to right-to-left shunting. Does not appear to be volume overloaded. As patient appears to be stable from a respiratory standpoint despite high oxygen requirements, does not require ICU level of care at this time.”

Hospitalization, Try #2

Step 2: Call medicine for admission

- Medicine admits
- Medicine night admitting team plan “Had a long discussion with patient and her husband. She would like to meet with Oncology here at UCSD to see what kind of other options she may have besides chemotherapy. We discussed hospice. **Pt and husband are not ready for hospice.** Also we discussed code status- she does want CPR, defibrillation and intubation and therefore will be full code. Pt states **‘I know I may die but I want to keep fighting.’ Will consult palliative care as well.**”
- Continued lovenox as stated exam consistent with PE despite lack of such on CT chest
- Patient told it is unclear if CT findings represented blood clot or tumor

Hospitalization Continues

Step 3: Transfer from night admitting team to the day ward team.

Step 4: Day ward team re-thinks the plan.

“# RA Thrombus: The most immediately life-threatening problem. Since HC does not have CT Surgery team, **am trying to arrange a transfer to Thorton for CT surgery evaluation**

-CT surgery at Thorton paged today, aware of patient, will likely have attending to attending

Synovial Sarcoma Stage IV: S/p 6 rounds of ifosfamide therapy and surgery. **Currently does not want anymore chemotherapy**

-Heme-Onc consult appreciated, they agree the RA thrombus is the most urgent matter and agrees surgical intervention may be appropriate, awaiting final consult recs”

Onto the consults!

Step 5: Heme/onc recs: Patient “clear she is not interested in more cytotoxic chemotherapy or radiation therapy options for her disease, but **would be interested in targeted therapies** based on mutation analysis or in tyrosine kinase inhibitors and other targeted agents. Before any systemic therapy options can be discussed, will need to address the atrial mass. Have discussed with CT surgery who will evaluate patient tomorrow, but preliminary discussion suggests that if the tumor mass is a thrombus (sic), she may be eligible for atri-ectomy. However if the mass is tumor, then surgery would not be an option. **Further imaging with cardiac MRI may be needed** to help distinguish.”

Consults Part Deux

Step 6: Await CT surgery official recommendations. These are relayed 9/5/14. “27 year old female with h/o stage IV synovial sarcoma now presenting with SOB possibly from atrial mass. Unclear whether mass is thrombus or sarcoma. **Please obtain MRI.** Will need to transfer to CVC for further workup and possibly operative intervention. “

Step 7: Heme/onc addresses goals of care: “Had more discussions today with patient, who **seems surprisingly stable mentally and accepting of her situation.** Not sure if this is real, denial, or just guards up. In any event she is prepared to undergo surgery if necessary.”

Step 8: MRI ordered 9/5/14 at 4:51PM, ordered as routine, and Thornton contacted for transfer request from IMU to IMU. **Transfer denied** by Thornton IMU charge nurse given patient is still on 15L non-rebreather, saturating 91-98%

Things progress

Step 9: Call ICU team at Thornton to transfer instead, ICU team at Thornton asks the MICU service to evaluate for upgrade in level of care, primary team calls MICU attending to discuss need for upgrade to ICU to transfer to the Thornton ICU for surgery evaluation.

Step 10: ICU consult #2, attending felt status was not changed since admission. “**SOB: Stable since admission on 15L non-rebreather.** No changes in clinical status since 7/4/14 consult. Agree with CT surgical interventions. **Does not necessitate ICU level of care** at this time. Continue current management.”

Things Worsen

Step 11: Patient **desaturates to 79-84%** range at 7pm on 9/5/14. ICU transfer accepted.

“1) RA Thrombus: Given acute hypoxic event concerning for embolic phenomenon. BiPAP has improved oxygenation for the moment, but this is a temporizing measure. The patient's **prognosis appears grim**, but she wishes to be full code/full care and have every effort done including intubation and surgery **to attempt to save her**. CT surgery made aware of the acute decompensation. Currently coordinating with Thornton for ICU transfer given CT surgery.

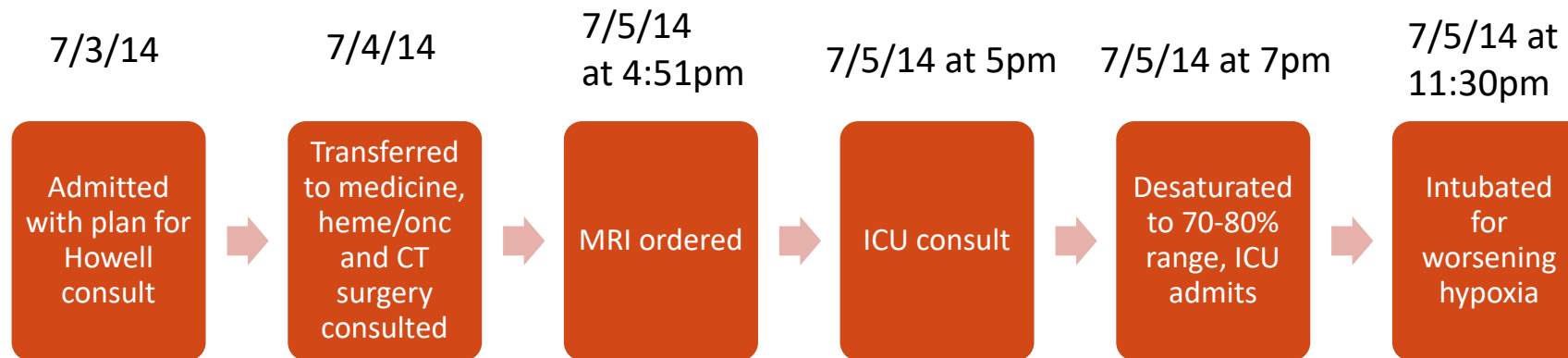
- Will need to intubate prior to MRI, anesthesia paged

- Per CT surg a STAT MRI is needed prior to operative management and hence will attempt to perform tonight to assess whether thrombus is clot vs. tumor.

2) Synovial Sarcoma: Per heme/onc unclear as to further chemo treatments, **to be determined should patient survive** post-operatively.”

Step 12: First blood gas obtained: 15L face mask, 7.49/15/44/12

Let's Recap



Worse Still

Step 13: Start pressors, Increase oxygenation. Blood gas post-intubation on PEEP 10, FiO2 100% was 7.32/22/44/11

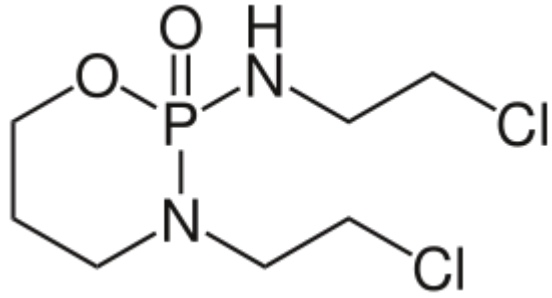
“the patient has had worsening hypotension requiring arterial line monitoring and increasing pressor requirements. **Currently on neo @35, vaso @0.04, and levo @30.** This is in the setting of multiple sedating drips (versed, fentanyl, and propofol) as well positive pressure ventilation in setting of poor right heart function from near complete atrial occlusion by thrombus. Currently awaiting MRI to evaluate for clot vs. tumor. Further management to be determined by this imaging. ABG has not responded to vent despite being on VTPC 100% FiO2, rate 20, PEEP 10, TV 500. Likely that patient has shunt physiology from embolic shower from right atria resulting in this difficulty oxygenating. **Prognosis at this point is quite grim,** and should MRI show tumor invasion, discussion with family will need to be had regarding **futile care** and compassionate extubation. Should the right atria appear to contain clot, will address further with CT surgery. This being said, at this point the patient is too unstable for aggressive interventions given severe hypoxia and profound hypotension. **Will add epinephrine gtt,** initiate bicarb 150mEq gtt @100ml/hr, and D50 + CaGluconate for hyperkalemia.”

Step 14: Discuss futility with family

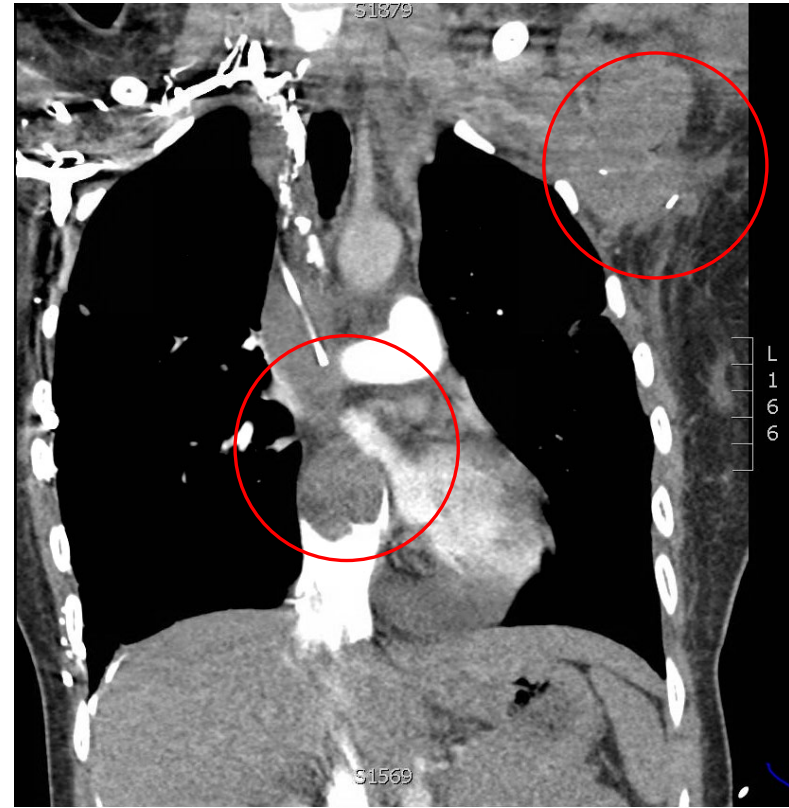
“Patient's clinical condition has continued to worsen, **now maxed on fourth pressor** with persistent hypotension and **intermittent lost pulses**. At time of exam patient was cool in the extremities with palpable dp pulses. Discussed with the family that at this time patient is at the limits of medical care **and it is likely that the patient will suffer a cardiac arrest today**. The family understood and stated that despite chest compressions being a futile effort given the irreversibility of the underlying cause, they feel the patient would want to **proceed with chest compressions**. Hence when a code situation arises we will proceed with a full code scenario as per family wishes.”

Step 15: Call code blue after PEA arrest at 2:22PM. 30 minutes of CPR, time of death 2:52PM.

Break to summarize



Ifosfamide



What was missing?

- 1) Howell service never consulted despite night admitting service planning to proceed down that path
- 2) ICU admission blocked initially
- 3) MRI not ordered the day CT surgery recommended it, but at 5pm the following day
- 4) MRI ordered routine instead of STAT
- 5) Was the MRI really needed? When should goals of care and discussion of futility have been had? First mention of futility was not made until the patient was intubated.
- 6) Should she have been coded? Could the goals of care discussion have taken a different path?
- 7) Where were outside records?

Past Medical History (as obtained per records uploaded to Epic the day after her death)

Stage IV Axillary synovial sarcoma with metastases to lungs:

5/2013: Lump noted in left axilla, growing

5/31/2013: Left axillary lump excision, path showing biphasic synovial sarcoma

6/26/2013: CT chest with bilateral pulmonary nodules c/w metastatic disease and a PE, also found to have DVT in bilateral upper extremities, started on lovenox

Fall 2013: Ifosfamide, 6 cycles, partial response

December 2013: Pulmonary surgery in Germany, removal of 19 tumor nodules from the left lung

February 2014: 34 nodules removed from the right lung in Germany. Path + for metastatic synovial sarcoma

March 2014: CT chest with worsening lung mets, received a seventh cycle of ifosfamide

Course Continued

June 2014: Seen at MD Anderson, with CT chest performed.

“Heterogenously enhancing mass in the left axillary region...appears larger than on previous study (June 2013). Measures 3.1cm in AP vs. 2.4cm prior. Mass invades the vasculature in the region and extends into the left brachiocephalic vein, into the SVC, and there is a large mass in the right atrium 4.6cm in diameter...signs of metastatic disease are noted in the lung parenchyma with numerous bilateral pulmonary nodules, the largest measuring 2.3cm.”

At this time (6/10/14) denied shortness of breath, was on room air, saturating 95%.

Heme/onc recommendations at this time were to start Adriamycin and dacarbazine ASAP. The patient wanted to wait and have chemotherapy sensitivity testing, which was discouraged.

Subsequently the patient moved to San Diego and opted out of further chemotherapy.

Prognosis and gravity of the tumor burden and atrial clot were not addressed. Rather patient was requesting repeat ultrasound to ensure her prior upper extremity DVTs had resolved after lovenox therapy.

Medications (as per H&P)

- Enoxaparin 60mg SQ q12h
- Naltrexone 3mL PO

All Prior to Admission Medications

- Alpha lipoic acid 600mg BID
- Antioxidant complex 1 tab TID
- Vitamin B17 1 tab TID
- Beta carotene 25000 IU BID
- Colostrum 500mg BID
- Dendritic cell shots 1 injection weekly
- Digestive enzymes 2 tablets BID
- Naltrexone 3mg BID
- NK-stim 2 tablets BID
- Pancreatic enzyme 2 tabs BID
- Probiotic 1 tab TID
- Vitamin C 1000mg 4x/day
- IV Vitamin C infusion monthly
- Vitamin E 400 IU BID
- Wellness booster 1 tablet BID
- Alive 3 tablets daily
- Selenium 200mcg daily
- Vitamin D3 50000 IU daily
- Enoxaparin 60mg BID

Complicated Bereavement



Complicated Bereavement

DSM V Diagnosis, Persistent Complicated Bereavement Disorder

- 12 months elapsed since death of someone close to the bereaved (6 months in children)
- Persistent yearning/longing for the deceased
- Intense sorrow, frequent crying, preoccupation with the deceased/manner in which the death occurred
- Also must meet 6/12 additional criteria as outlined in the DSM V

Reactive distress to the death

1. Marked difficulty accepting the death. In children, this is dependent on the child's capacity to comprehend the meaning and permanence of death.
2. Experiencing disbelief or emotional numbness over the loss.
3. Difficulty with positive reminiscing about the deceased.
4. Bitterness or anger related to the loss.
5. Maladaptive appraisals about oneself in relation to the deceased or the death (e.g., self-blame).
6. Excessive avoidance of reminders of the loss (e.g., avoidance of individuals, places, or situations associated with the deceased; in children, this may include avoidance of thoughts and feelings regarding the deceased).

Social/identity disruption

7. A desire to die in order to be with the deceased.
8. Difficulty trusting other individuals since the death.
9. Feeling alone or detached from other individuals since the death.
10. Feeling that life is meaningless or empty without the deceased, or the belief that one cannot function without the deceased.
11. Confusion about one's role in life, or a diminished sense of one's identity (e.g., feeling that a part of oneself died with the deceased).
12. Difficulty or reluctance to pursue interests since the loss or to plan for the future (e.g., friendships, activities).

D. The disturbance causes clinically significant distress or impairment in social, occupational, or other important areas of functioning.

E. The bereavement reaction is out of proportion to or inconsistent with cultural, religious, or age-appropriate norms.

Specify if:

With traumatic bereavement: Bereavement due to homicide or suicide with persistent distressing preoccupations regarding the traumatic nature of the death (often in response to loss reminders), including the deceased's last moments, degree of suffering and mutilating injury, or the malicious or intentional nature of the death.

Complicated Bereavement

Numerous studies have shown a high prevalence in family members of patients in the ICU.

- One study looking at 50 participants found a 46% incidence of complicated bereavement.

Prevalence nationwide is 2.4-4.8%

High concordant rates of depression, anxiety, and PTSD in family members

Complicated Bereavement

Risk factors for development of this disorder include:

- Female gender
- Spouse or parent-child relationship
- Lack of religious belief
- Unavailable family support
- History of co-morbid mood disorder

Protective factors include:

- Duration of caring for the patients (months vs. days)
- Caregivers with history of medical disease
- Patients being on Hospice

Autonomy vs. Beneficence

Palliative Paternalism – Identify Maladaptive Coping

TABLE 1. RISK FACTORS FOR PROTRACTED EMOTIONAL RECOVERY OR MALADAPTIVE COPING

Cognitive

- Cognitively delayed—inability to consider two opposing options at the same time or unable to conceptualize possible outcomes
- Medically naïve—may view the human body similar to a car; discrete parts that work together but have no real interaction
- Extremes of age—young or old (dementia)

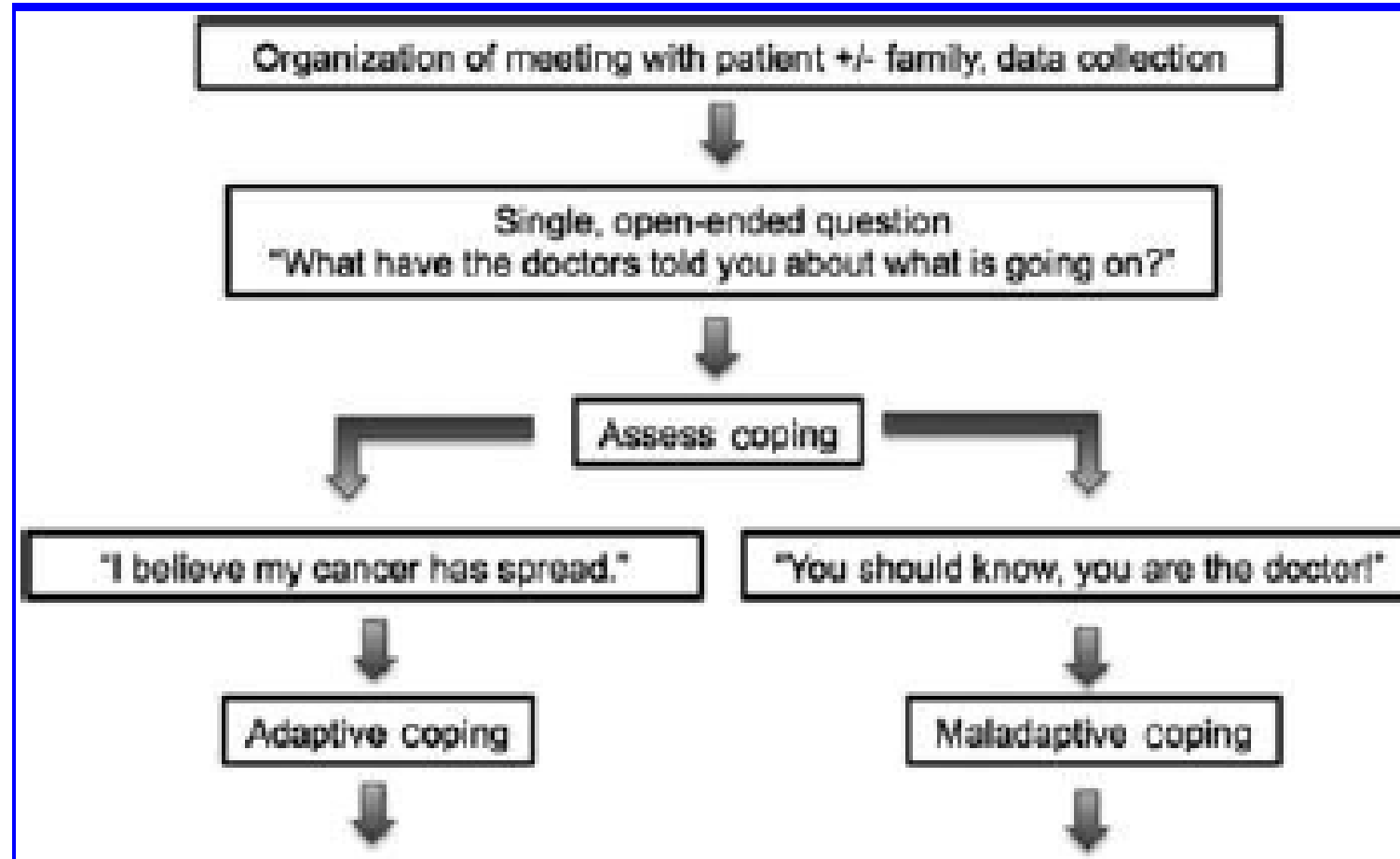
Emotional/psychological

- Emotionally arrested
- Shame is a prominent emotion; patient may inaccurately believe it is his or her fault he or she is sick
- Posttraumatic stress disorder
- Emotionally reactive
- Serious mental illness
- Magical thinking
- Need to assert own authority in spite of possible harm to self
- Substance abuse

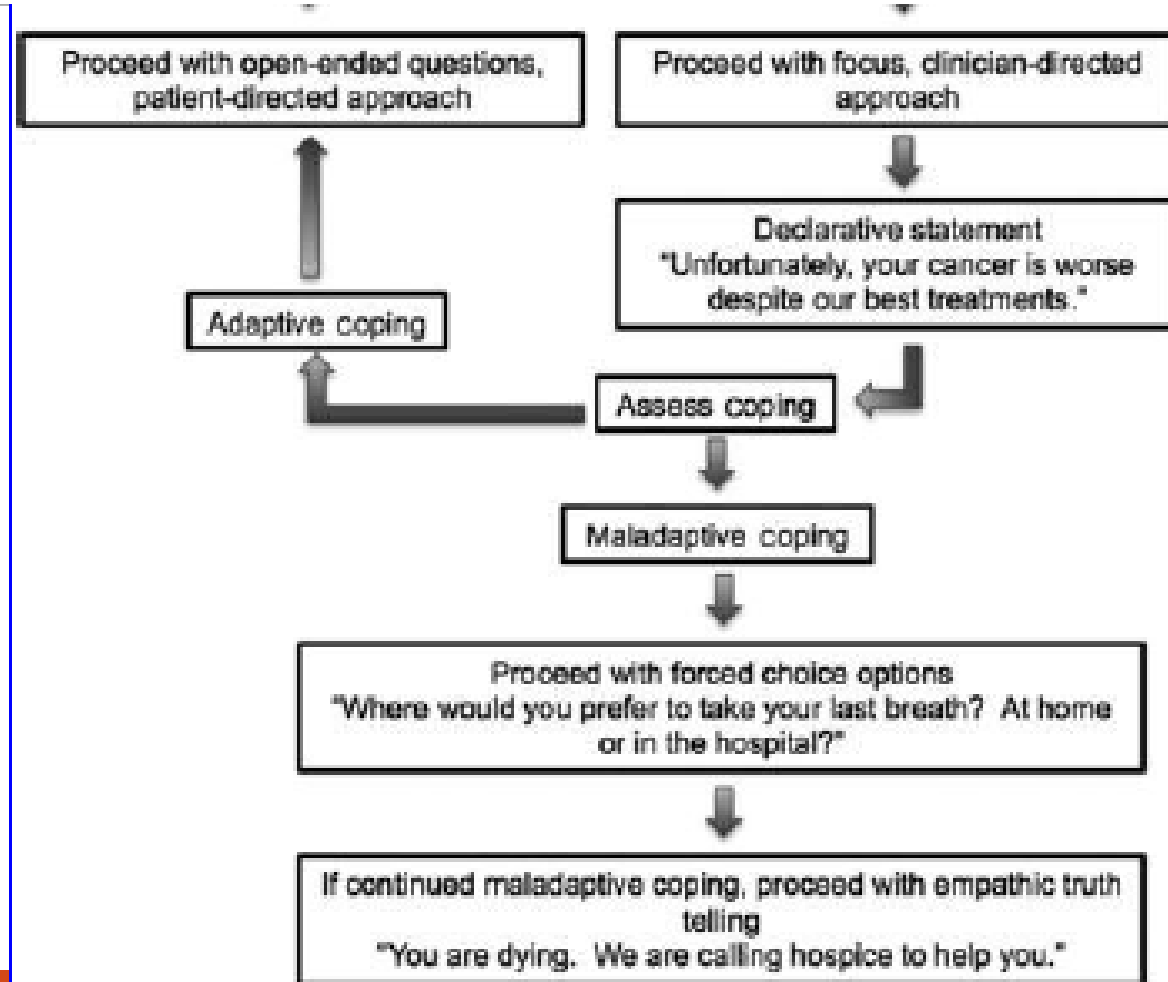
Social/cultural

- Cultures that are traditionally mistrustful of the medical community
 - Cultures that focus on the collective rather than the individual
 - Cultures that value deference to authority
 - Individuals who believe only option is a miracle
-

Palliative Paternalism



Palliative Paternalism



Policies on Futility

UCSD: MCP 531.1 Care which has “no realistic chance of returning the patient to a level of health that permits survival outside the acute care setting”

- “If disagreement arises between family members or surrogate, a family conference should be held with the members of the treating team to discuss the patient’s prognosis, goals of care, and proposed treatments in order to try and achieve consensus. If disagreement persists, ethics consultation should be requested.”
- “If the Medical Ethics Committee agrees with the Responsible Physician’s determination of non-beneficial treatment and the patient’s or surrogate’s position remains unchanged, the Responsible Physician may decline to provide the treatment determined to be non-beneficial. This treatment will not be offered at UCSDHS. The patient and family will be given a reasonable period of time to prepare after notification that life sustaining treatment will be withdrawn (generally 24-72 hours).”

California Probate Law (2003): "When further intervention to prolong the life of a patient becomes futile, physicians have an obligation to shift the intent of care toward comfort and closure."

AMA: Policy 2.035 “Physicians are not ethically obligated to deliver medical treatments that, in their best professional judgment, will not have a reasonable chance of benefiting their patients”

References

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