

Sometimes we get it wrong

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Pediatric cancer

- Survival ~80%
- One in five children still die
- Second commonest cause of death in developed countries

Delay in diagnosis in children with cancer

- In at least 25% the delay is more than 3 months
- GPs involved in 80% of initial presentation – Important role
- Significantly shorter delay when GPs not involved
- Likely due to more rapidly progressive cases with aggressive tumor growth presenting in Emergency.

Ahrensberg JM, Oleson F et al, 2013; Vedsted & Oleson, 2009,

Presenting symptoms(

Ahrensberg JM al. British J Clinical practice 2012

- In daily clinical practice cancer often presents with symptoms mimicking frequently seen symptoms of benign conditions
- Symptoms were few –average of 2.4 per child
Only one symptom in 38%
- 72% in category “general and unspecified” except for CNS (headache , vomiting)
- Only 20% presented with symptoms severe enough to cause “alarm”

Danish National study

Ahrensberg JM et al Acta Paed 2012

- Girls almost twice as likely to have delay
- Older children > younger
- Bone and CNS longest (med 88 days)

- Socioeconomic: Mothers with low education
25% experienced a delay of minimum 18 days
cf to 3-7 days for more educated mothers

Does delay matter?

- **Does delay affect survival?**

Depends on tumor type

Survival more dependent on tumor biology and chemo-responsiveness than time to diagnosis

-osteogenic sarcoma –long standing history prior to diagnosis –survival better

-leukemia –high risk vs standard risk ---related to biology and not to delay

In some cases delay does matter

- **Does delay affect quality of survival?**

If delay results in increased therapy given then undoubtedly risk of long term consequences increases

- **Affects family's level of confidence in caregiver and the entire health system –increases anxiety and distrust**

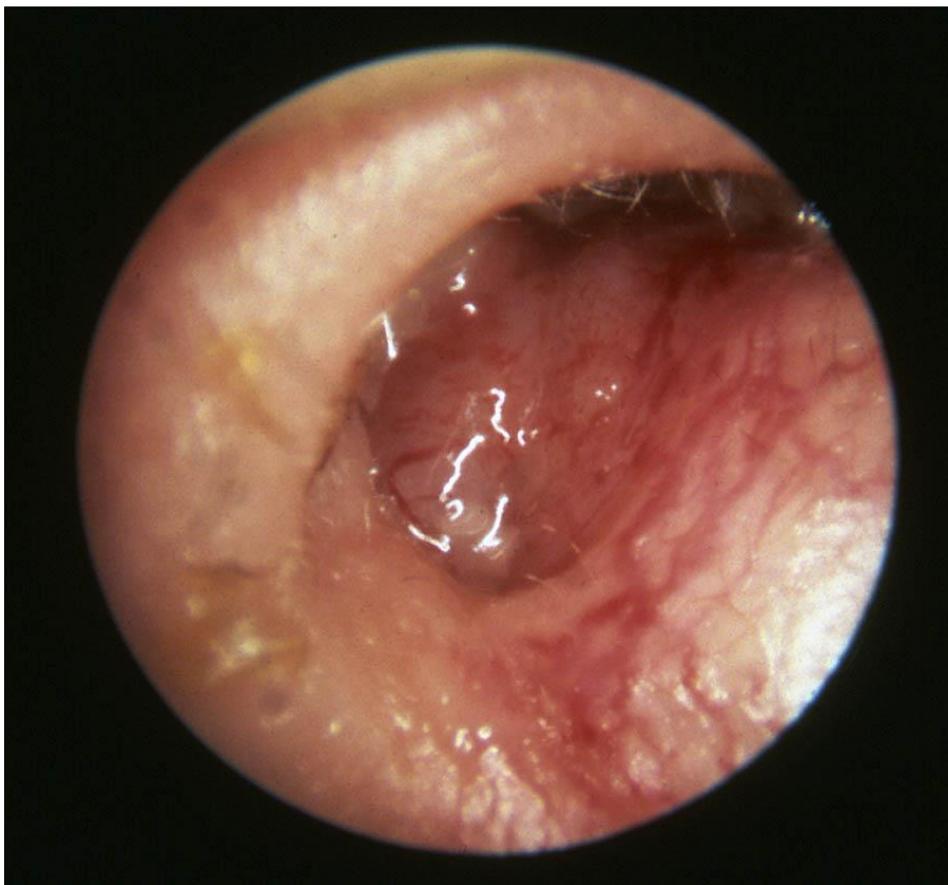
Case 1 –effect on survival

- Patient JS –intranasal mass as a young infant

At first presentation (picture courtesy
of her mother)



At presentation to ENT surgeon –
diagnosis hemangioma without biopsy



Case 1 cont

- Referred by GP to an ENT surgeon who diagnosed a hemangioma and did not follow up.
- She presented 6 weeks later with a large and now inoperable intranasal rhabdomyosarcoma
- Treated with chemotherapy with good response and complete resection attempted but clear margins could not be obtained without mutilating surgery.
- Decision was made with her mother not to offer high dose radiation therapy in view of her very young age . The tumor recurred and proved to be fatal.

Case 2

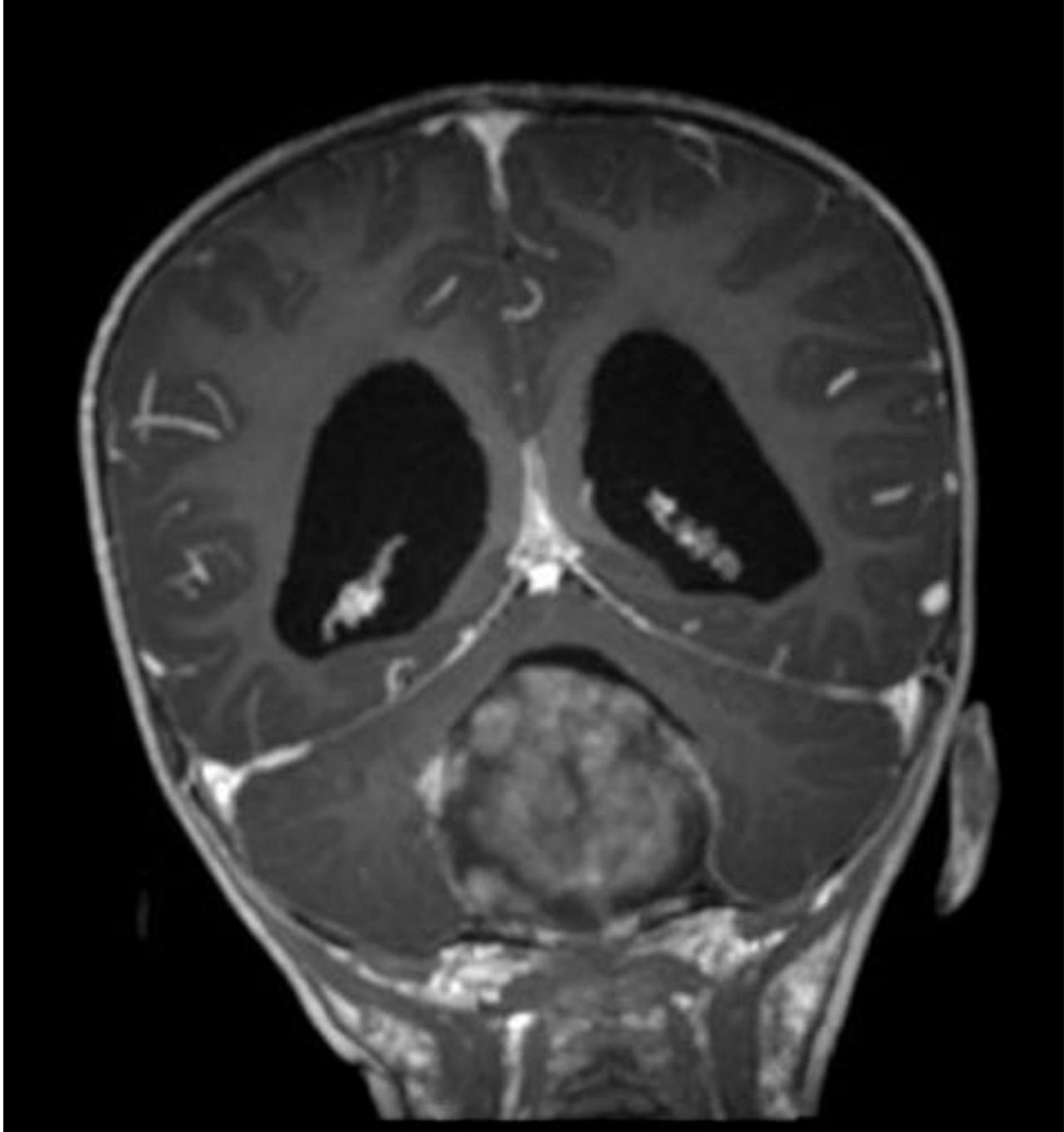
- 22 month old girl

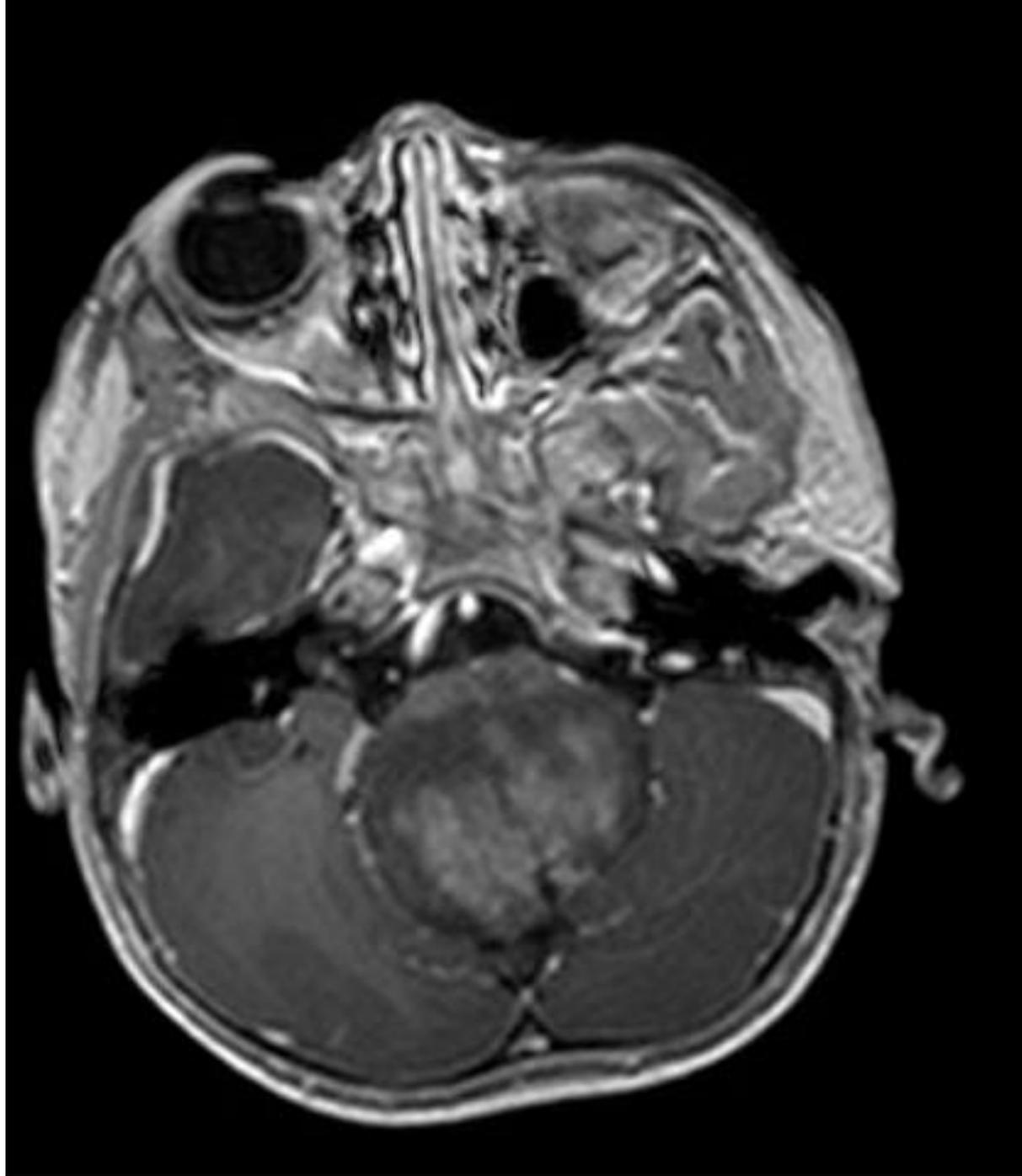
Vomiting for 6 months

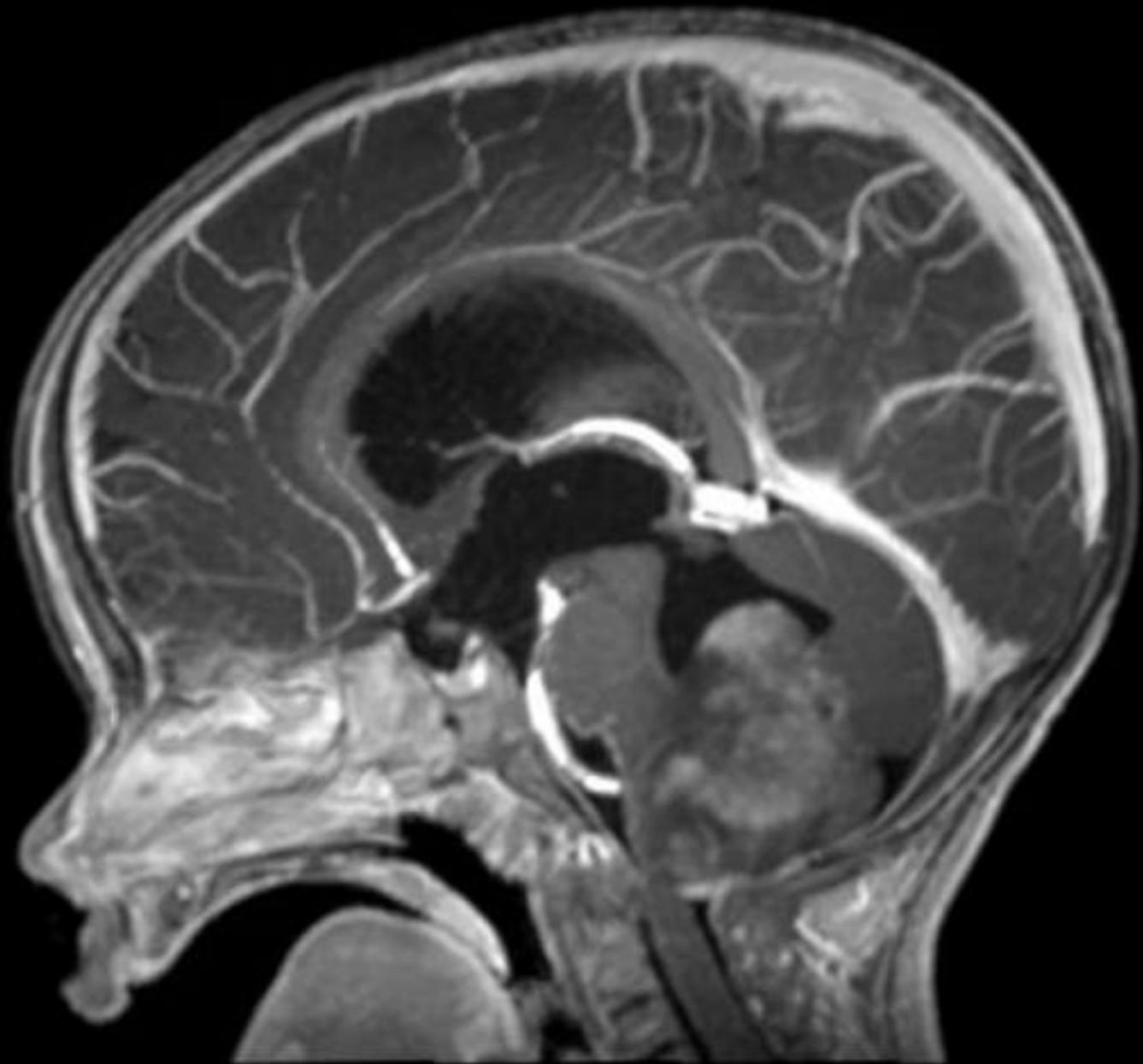
Seen many times by primary caregiver and by GI service at for “reflux” and “failure to thrive”

2 x GI endoscopies done

Eventually mother noted stridor and she was readmitted and MRI brain done







Case 2 cont

- Pathology showed a low grade pilocytic astrocytoma
- Low grade tumors less responsive to chemotherapy
- Newer targeted type therapies being tried
- Best chance for cure in low grade glioma is a complete resection but at this stage only a partial resection was possible

Convinced?

- Even though biology of the tumor and the chemosensitivity are the most important factors affecting survival, undue delay can clearly affect survival

Does delay matter?

- What about quality of survival?

Case 3

- Male patient DB—presented at 8 years of age with a mass arising from the mandible
- Diagnosed as a benign tumor –non-urgent referral to surgery ---
- Surgeon decided to do an excisional biopsy –attempt to resect the entire tumor

Pathology showed Ewings sarcoma but all margins were positive for tumor.

Referred to tertiary center

Chemotherapy was given but because of residual tumor he also needed high dose radiation to the jaw for local control.

He survived but with a significant cosmetic defect from the radiation and a risk of developing a second cancer (radiation induced osteogenic sarcoma)

Case 4 --11yr old male presenting with a jaw mass -very similar presentation to case 3



Case 4 11yr old male:

- Percutaneous needle biopsy -showed Ewings sarcoma
- Preoperative (neoadjuvant) chemotherapy given to shrink the tumor
- Followed by surgical resection –pathology showed that all surgical margins were clear o tumor
- He continued on chemotherapy alone –no Radiation therapy was required and he had an excellent cosmetic result and much less risk of a second tumor

Cough as the first symptom of malignancy

- A common symptom when the diagnosis is delayed

Case 5

- 16 yr old female with a 2 month history of persistent cough –diagnosis “asthma”
- Mother eventually brought her to the ER – puffy, chest pain, swollen neck, headache
- Now typical appearance of a superior vena caval obstruction
- Chest X-ray was done

Anterior superior mediastinal mass



When you see this on X-ray --the primary caregiver needs to remember:

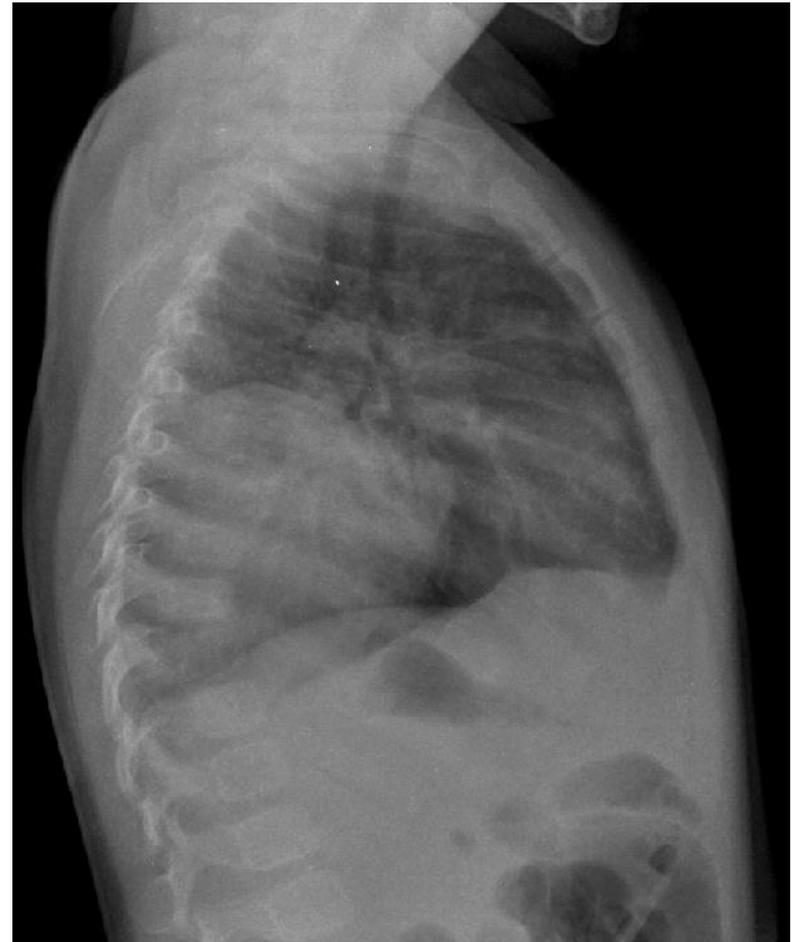
- NEVER lie the patient flat
- Not for CT scan, not for Echocardiogram
- It is the physicians job to tell the radiology /ECHO tech not to lie the patient flat in supine –do CT with the patient on their side or in prone.

Case 6

- 11 month old with persistent cough – diagnosed as “pneumonia”
- 1 week history of not moving her legs

Case 6

11 mo girl with history of “pneumonia”, not using legs for 1 week!—X-ray shows a posterior mediastinal mass – neuroblastoma



Neuroblastoma with spread into spinal canal and compression of the cord



Spinal cord compression

- Back pain 90%

Persistent

Wakes the child from sleep

- Back tenderness 90%

Think spinal cord!!

Complete loss of function for >24 hours may not recover

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- Delay in diagnosis may affect survival and quality of survival
- What the first caregiver to see the patient does is really important

What lessons can we learn

Some symptoms and signs should not be missed

Have been called “alarm” symptoms and signs and include:

pallor, bleeding , bone pain and tenderness, periorbital bruising, limp, weakness, new onset squint, head tilt, ataxia, masses, back pain and tenderness,

What lessons can we learn

However, many more patients present with non-specific and vague symptoms and signs

----Vomiting, cough, headache, “growing pains”, fever

Cancer survivor J Machta --- editorial BMJ 2014 wrote

“Misdiagnosis of pain as “growing pains” is a painfully recurrent theme”

Brain tumors

- ***Persistent*** vomiting ---Early morning vomiting
- ***Persistent*** headaches
- ***Any headache in a child <4 years of age***
- New unsteady gait
- Proptosis
- New onset squint
- Head tilt
- Increasing head size in a young infant
- Weakness

Other warning signs:

- ***Persistent /recurrent*** cough
- ***Persistent/unexplained pain***
- Generalized lymphadenopathy
- Unexplained bruising /bleeding
- ***Persistent/recurrent*** fever

Conclusion

- What the first physician to see the patient with cancer does may be very important for survival and for quality of survival
- Some symptoms and signs should always raise concern

but

Even if vague and non-specific, persistent symptoms and signs need to be investigated

Conclusion cont.

- 1 in 500-600 children will develop cancer by age 15 –you will see them in your practice
- High index of suspicion!!