

# Radical Resection of a Giant Ewing's Sarcoma of the Anterior Chest Wall

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# Background

- The most frequent malignant tumor of the chest wall in children and young adults.
- An extremely aggressive tumor.
- Management requires intensive local therapy with surgical resection, radiotherapy, or both, as well as adjuvant chemotherapy to prevent or treat metastases.
- Local recurrence in this tumor has been frequent.



# Background

- One major clinical decision at presentation of ES of the chest wall is whether to resect the primary tumor initially or after neoadjuvant chemotherapy.
- Complete resection is the goal, and if this is accomplished with adequate margins, it allows the pt to avoid radiation, with its potential long-term complications:
  - pulmonary fibrosis.
  - increased incidence of CAD.
  - significant incidence of secondary tumors.



# Background

- Although thoracic tumors are frequently large and similar to the size of tumors of the pelvis, a primary resection is often attempted for the chest wall lesions while secondary resection is generally the approach for tumors of this size at other sites.



# Patient Description

## History:

32 year old man.

Huge mass causing a bulge in the right chest wall that had increased dramatically in size over the previous 6 mo.

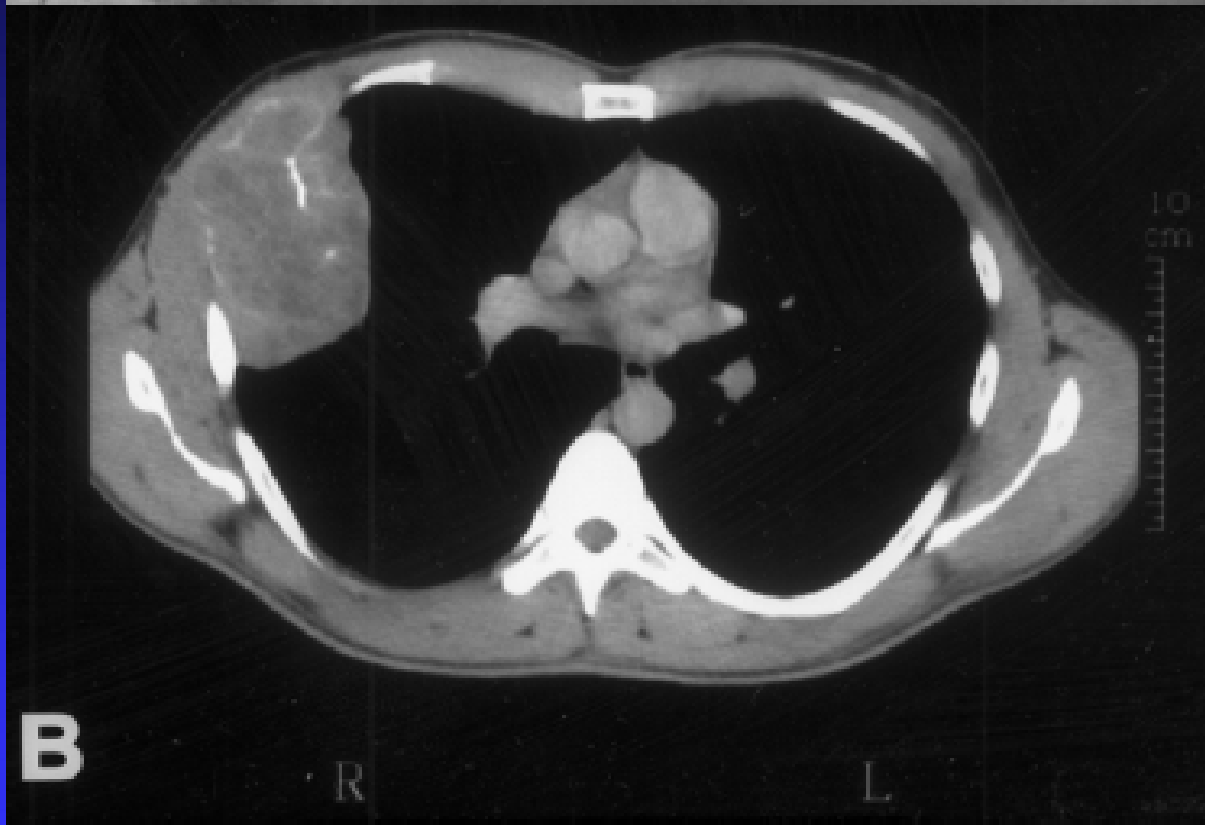
Accompanying chest pains.

## Physical examination:

Mass was hard, fixed and non-tender



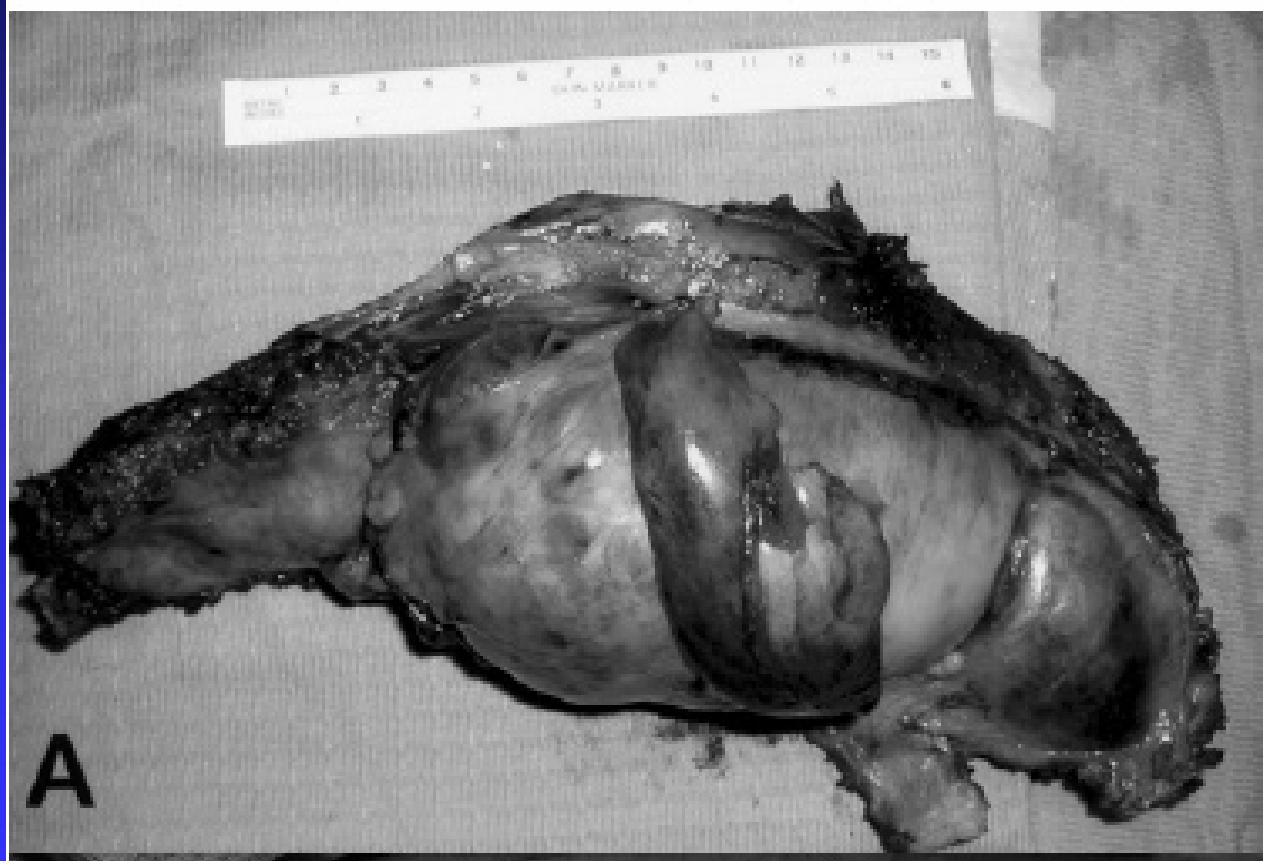




# Surgical Methods ( I )

- Radical en bloc resection of a 7 x 8.5 x 21 cm segment of the anterior chest wall was performed, including the entire gross tumor and a portion of lung and 4 cm macroscopically normal surrounding tissue.
- The microscopic evaluation of the margins by frozen section was negative.





## Surgical Methods ( II )

The large chest wall defect was reconstructed with a double-layer composite of Marlex mesh to confer adequate stability and avoid 'flail' physiology.





# Results ( I )

- Histopathologic study confirmed a completely resected ES/PNET.
- A work-up for metastases was performed that included CT of the abdomen and a radionuclide bone scan.



## Results ( II )

- None of these procedures showed any evidence of distant metastatic lesions.
- Pt underwent 30 wks of chemotherapy that included courses of vincristine, cyclophosphamide, and doxorubicin alternating with courses of ifosfamide and etoposide.
- Pt remained disease free 1 y after cessation of therapy.



# Comment

- ES is the 4<sup>th</sup> most common malignant bone tumor overall & the 2<sup>nd</sup> most common malignant bone tumor in pediatric pts.
- Most ES occur in pts between 3 & 25 yrs (mean 13 yrs).
- ES have a female predilection of approximately 2:1.
- About 10% of ES occur within the ribs.



# Comment

- ES of the ribs has a propensity to spread inward towards the thoracic cavity.
- This extension may manifest as a strictly extrapleural mass pushing into the thoracic cavity, or it may invade the thoracic cavity with direct invasion of deep structures (mediastinum, pericardium, diaphragm)



# Literature Review

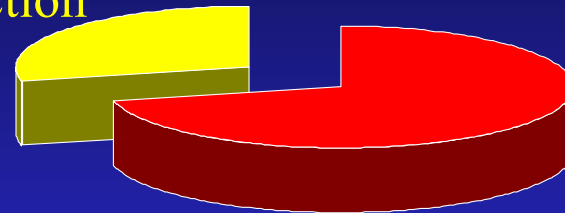
Shamberger RC *et al*, *Ann Surg* 2003;238:563-568

*ES/PNET of the chest wall*

*Impact of initial VS delayed resection on tumor margins, survival, and use of radiation therapy*

➤ 87 Pts

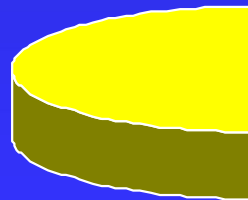
Primary resection



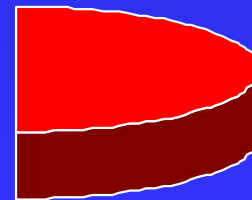
Delayed resections

➤ Negative margins assessed by pathologists for pts with **primary** resection (50%)

Negative margins



Positive margins



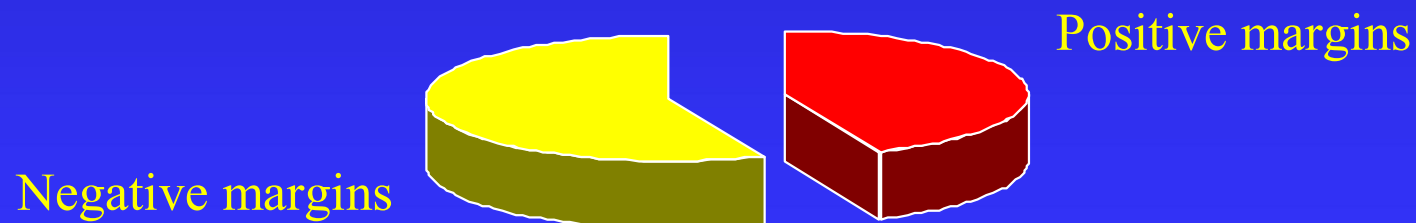
# Literature Review

Shamberger RC *et al*, *Ann Surg* 2003;238:563-568

*ES/PNET of the chest wall*

*Impact of initial VS delayed resection on tumor margins, survival, and use of radiation therapy*

- Negative margins assessed by pathologists for pts with **delayed** resection (77%)



Shamberger RC *et al*, *Ann Surg* 2003;238:563-568  
*ES/PNET of the chest wall*

- No survival benefit caused by primary resection of the tumor
- Pts undergoing definitive surgery after initial chemotherapy had a higher frequency of successful complete resection of the tumor, eventuating in the avoidance of radiation therapy to the chest.

# ESMO treatment plan for ES

Ann of oncol 20 (supplement 4): iv140-iv142, 2009

- Multimodal approaches within clinical trials, employing combination chemotherapy & surgery/radiotherapy, have raised 5-year survival rates from <10% to >60%.
- All current trials employ 3-6 cycles of initial chemotherapy after biopsy, followed by local therapy and another 6-10 cycles of chemotherapy usually applied at 3-wks intervals. Treatment duration is thus 8-12 m. Agents include doxorubicin, cyclophosphamide, ifosfamide, vincristine, dactinomycin & etoposide [Ib, A].



# ESMO treatment plan for ES

Ann of oncol 20 (supplement 4): iv140-iv142, 2009

- Despite lively debate, complete surgery, where feasible, is regarded as the best modality of local control. Radiotherapy should be applied if complete surgery is impossible, and should be discussed where histological response in the surgical specimen was poor [IV, C].
- Radiotherapy is applied at doses of 40-45 Gy for microscopic residues and 50-60 Gy for macroscopic disease [III, B].





Shamberger RC *et al*, *Ann Surg* 2003;238:563-568  
*ES/PNET of the chest wall*

- Majority of pts, adjuvant chemotherapy will significantly shrink the tumor as well as decrease its friability and vascularity, which will enhance the surgeon's ability to define the margins of the lesion and achieve its complete resection.
- This will not avoid the need for radiation in all pts, particularly those with paravertebral or apical involvement or those with extremely large tumors.

