Approach to a locally invasive pleural/ chest wall mass in a child

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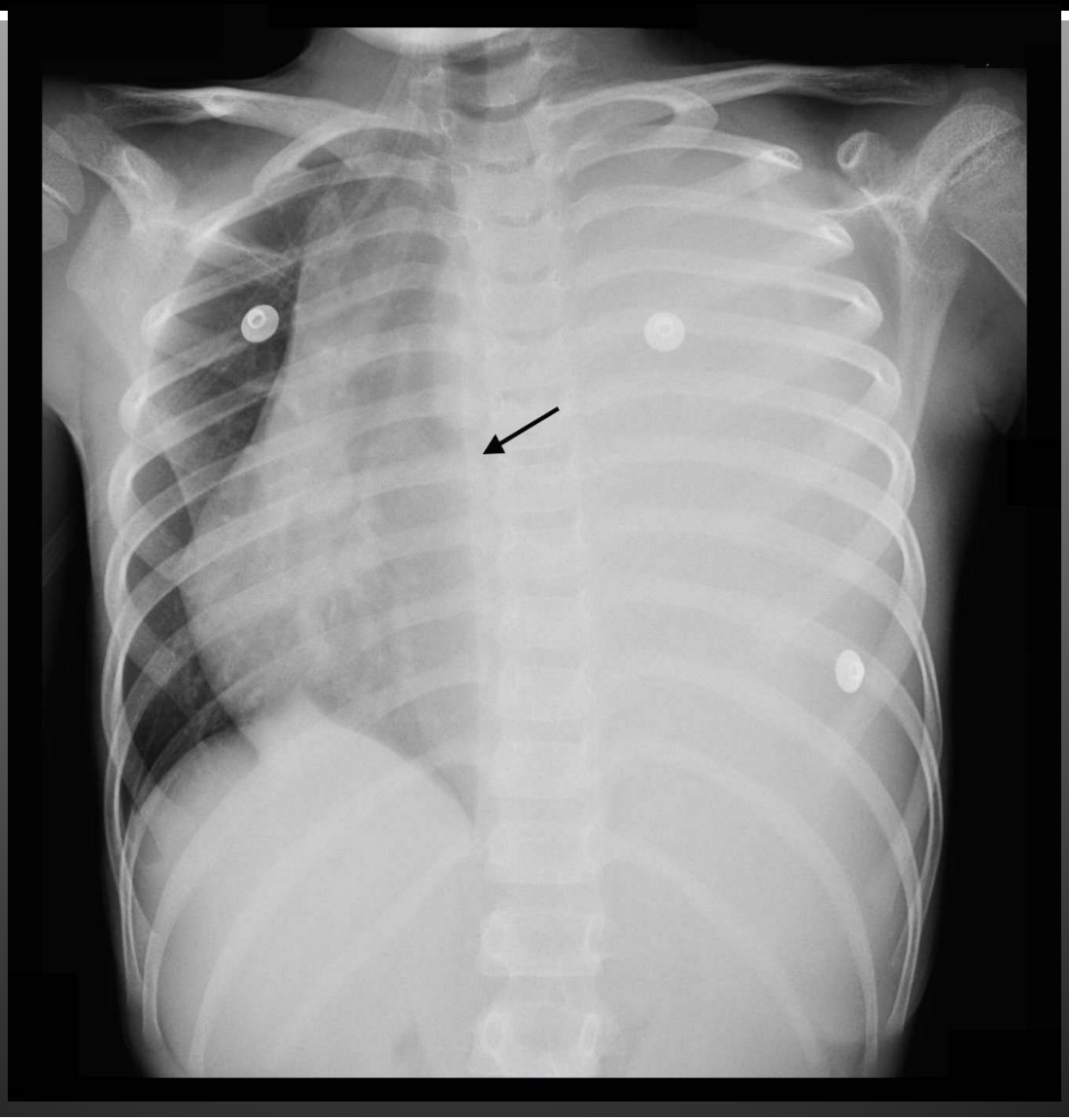
Objectives

- Formulate a differential diagnoses for a locally invasive pleural/chest wall lesion in a child
  - Clinical history (**N.B. age)
  - Imaging findings
- Highlight an entity that is infrequently reported in the literature

Case presentation

- 7-year-old boy who presented with a progressive history of dyspnoea
- No known co-morbidities
- Not immune-compromised

Work-up: CXR



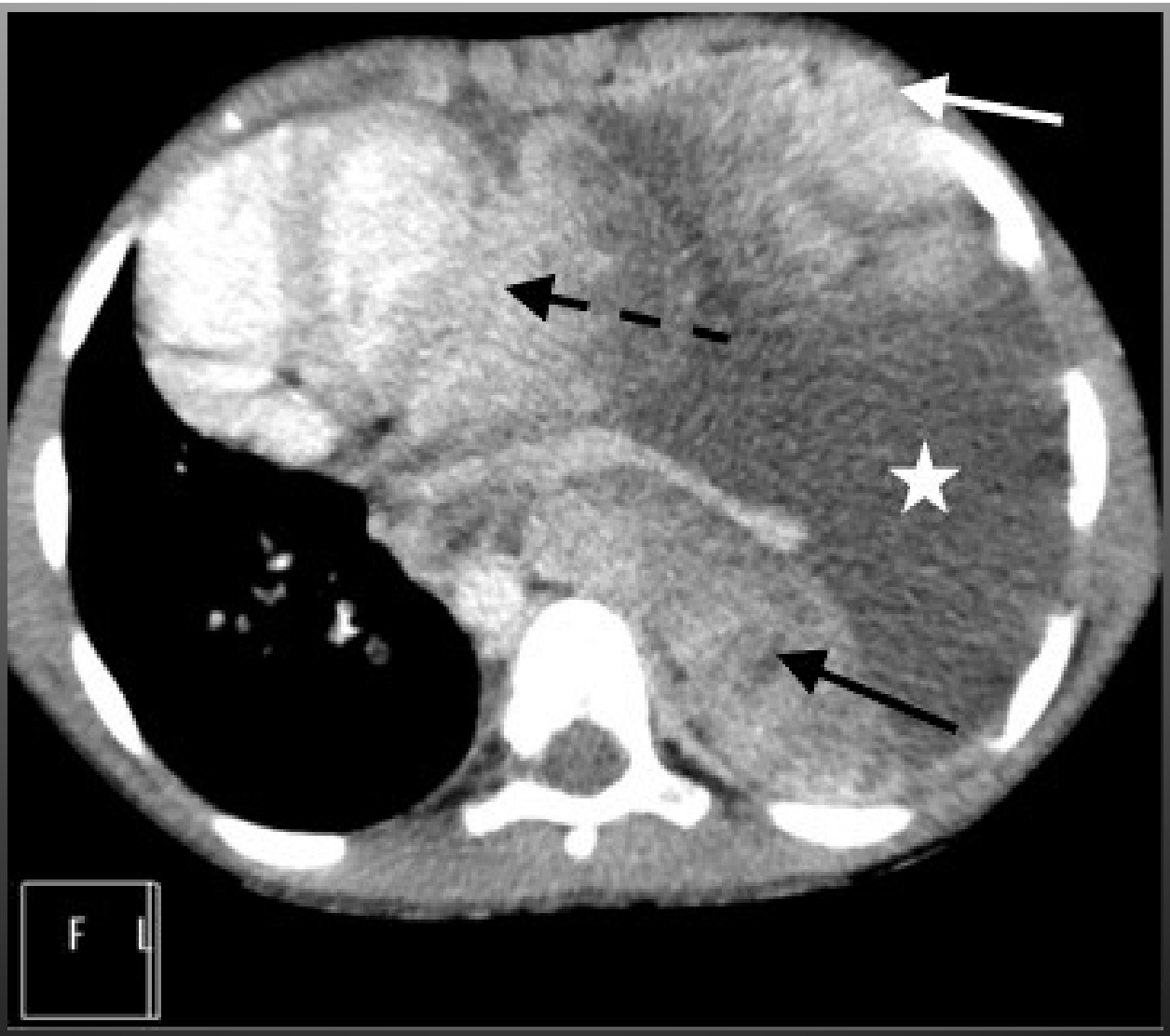
Work-up: Ultrasound

- Advantages:
  - absence of ionising radiation
  - easy accessibility
  - dynamic study that can be performed at the patient's bedside
  - guide needle-biopsy for the histological diagnosis

Work-up: Ultrasound

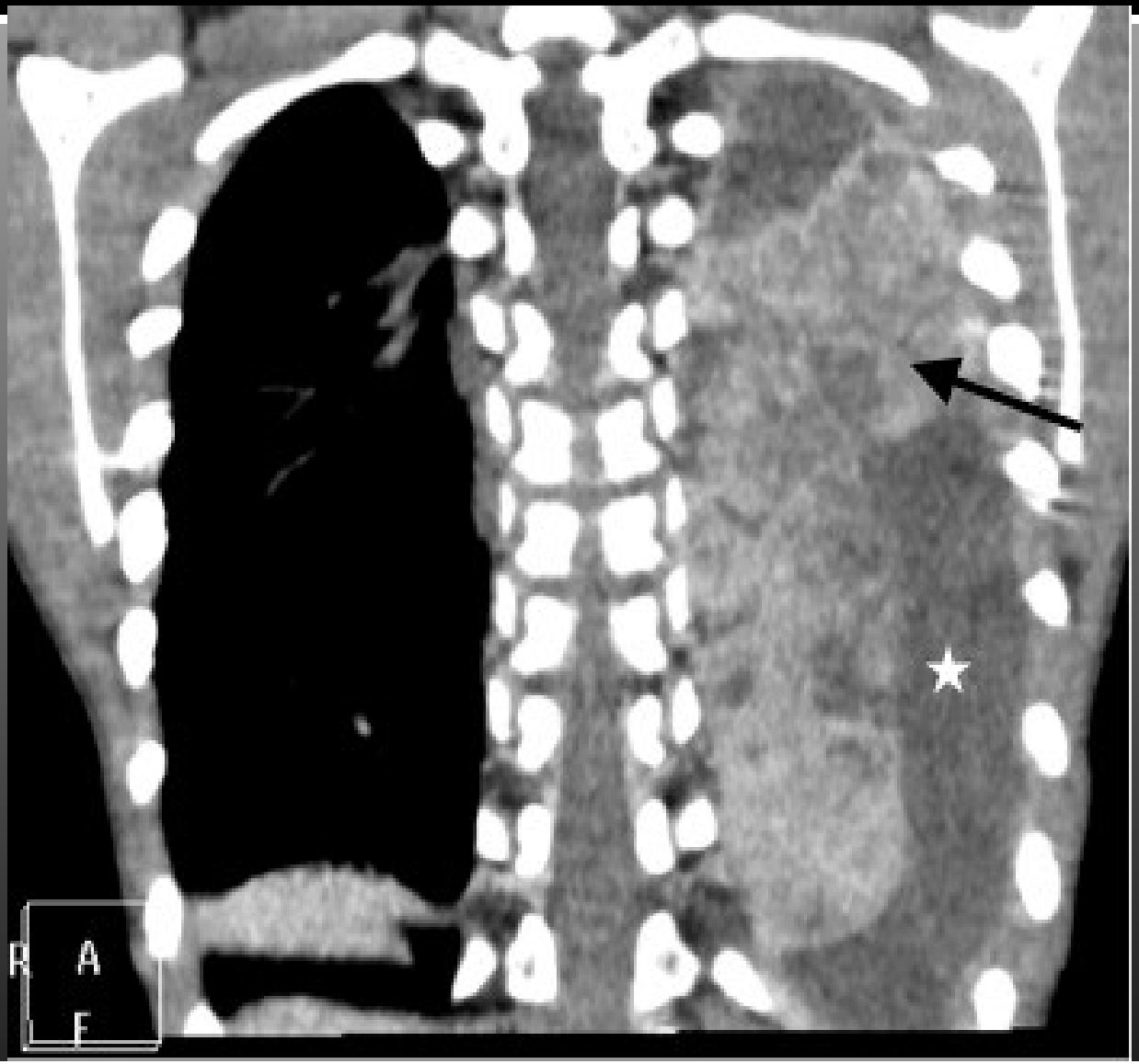
- Multiple pleural-based masses with a heterogeneous echotexture
- Doppler - vascular flow within the lesion
- Associated complex left pleural effusion

Work-up: CT



CT

CT

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Differential diagnosis

- Locally invasive pleural/chest wall tumours in a child
  - heterogeneous group of lesions
- Useful to categorize them on the basis of[1]:
  - primary versus secondary
  - site of origin (osseous and cartilage, or soft tissue)

Differential diagnosis

1. Metastases	2. Malignant soft tissue tumour	3. ESFT	4. Benign lesion with possible locally invasive features
• Rhabdomyosarcoma • Neuroblastoma • Lymphoma • Leukaemia	• Rhabdomysarcoma • Malignant peripheral nerve sheath tumour (MPNST) • Pleuropulmonary blastoma - type II/III	• ESFT - Extra-skeletal Ewing's Sarcoma (ES-EWS) **if primary osseous origin: • *EWS of the bone*	• Inflammatory myofibroblastic tumour (IMT) of the lung/pleura

1. Metastases

- **Neuroblastoma**
  - posterior mediastinal mass
  - may have direct invasion of the chest wall and extend into the adjacent neural formamina[2]
- **Lymphoma**
  - associated with immune compromised patients
- **Leukaemia**
  - constitutional symptoms in conjunction with the associated biochemical and bone marrow findings

2. Malignant soft tissue tumour

- **Rhabdomyosarcoma**
  - aggressive chest wall lesion [4]
  - Bony involvement typically occurs in the late stage of the disease - reported in ~20% of patients[2]
  - Embryonal rhabdomyosarcoma is a subtype which is unique to the paediatric population [5]

2. Malignant soft tissue tumour

- **MPNST's**
  - Arise within the nerve sheaths
    - sporadic or associated with neurofibromatosis type 1 (NF1)[2]
  - **Clue** - mass lesion that courses along the expected location of a peripheral nerve (may be associated with underlying bony erosions)[2]

2. Malignant soft tissue tumour

- **Pleuropulmonary blastomas**
  - Typically affects infants and children (overall median age of 4 years).[11]

3. ESFT

- Group of malignant small round blue cell tumours with varying degrees of neuro-ectodermal differentiation[6]
- Include:
  - Ewing sarcoma of the bone (EWS)
  - Extra-skeletal Ewing sarcoma (ES-EWS)
  - Peripheral primitive neuroectodermal tumour (pPNET)
  - Askin's tumour[2]
- Believed to represent the same entity that are histogenetically related, with minor differences in their differentiation[4,6]

3. ESFT: ES-EWS

- Typically manifests in children and young adults
  - 85% of reported cases 20 months - 30 years of age[4]
- Although ES-EWS is primarily a soft tissue tumour, it may invade adjacent osseous structures [7]

3. ESFT: ES-EWS

- **Imaging:**
  - solitary or multiple extra-pulmonary mass/es
  - eccentric growth along the chest wall [3,8]
  - expansion may result in the secondary collapse of the lung/directly invade the lung parenchyma [3]
  - paravertebral origin may result in extension through the vertebral foramina with secondary bony changes [3,4]
  - other reported findings:
    - pleural effusions, lymphadenopathy, lung and bone metastases [8]

4. Benign lesion with possible locally invasive features

- Lung IMT
  - usually appears as a solitary, well marginated, peripheral mass on CXR
  - heterogeneous enhancement on CT
  - Calcifications: children > adults
- Pleural IMT is exceedingly rare - may appear locally invasive with infiltration of the adjacent chest wall

Histology

- Biopsy of the anterior pleural-based mass
  - small round blue cell tumour
- Examples of small round blue cell tumours which may occur in the chest wall/pleura[7]:
  - Neuroblastoma
  - Rhadomysosarcoma
  - Lymphoma
  - **Ewing Sarcoma Family Tumours**

Diagnosis

- Immunohistochemistry of the biopsy specimens was then performed to further subtype this neoplasm
  - **ESFT** – Extra skeletal Ewing Sarcoma

Management

- Multidisciplinary
- Neoadjuvent chemotherapy is initially used:
  - decreasing the size of the primary lesion
  - eliminating micrometastases [4]
- Surgery - primary method for local control (dependant on tumour stage) [4]

Conclusion

- Locally invasive pleural/chest wall tumours
  - heterogeneous group of lesions
  - relatively infrequent in children and infants
- Categorizing them assists in narrowing the differential diagnosis:
  1. Metastases
  2. Malignant soft tissue tumours
  3. ESFT
  4. Benign lesions with possible locally invasive features

Conclusion

- ES-EWS should be considered:
  - child or adolescent presents with an ill defined, eccentric, chest wall mass, in the absence of a lesion with a primary osseous origin
- Imaging plays a key role:
  - tumour staging
  - therapeutic planning
  - follow up
- Biopsy required for a definitive diagnosis

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