

EWING'S SARCOMA IN RIB

A report on 7 cases with special emphasis to the early roentgen findings

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Abstract

During the last 2½ years 7 patients with malignant rib disease were referred to the Pediatric Oncological Centre at Amsterdam, Emma Children's Hospital. All proved to have Ewing's sarcoma. The clinical histories are outlined and the roentgenographic findings discussed. Special emphasis is given to the initial clinical and radiological data. These data are compared to those from previously published case reports.

Because of the intermittent character of symptoms, the excentric location of the soft tissue mass, and the often subtle rib lesion the diagnosis is frequently delayed for several months. Therefore, a good quality X-ray examination of the chest is necessary in every day practice. The recognition of a soft tissue mass in relation to the thoracic wall and/or the presence of "pleuritic" complaints make penetrated detail films of the ribs concerned obligatory. The combination of a large intrathoracic extrapleural mass and local rib abnormalities is highly suggestive for Ewing's sarcoma.

Information about the radiological suspicion of a Ewing's sarcoma of a rib is of great help to the surgeon who should take an extrapleural biopsy and also to the pathologist who may have serious difficulties in establishing the histological diagnosis.

Key-words: Ewing's sarcoma — Rib tumours — Thoracic wall — Child — Soft tissue swelling.

Introduction

Ewing's sarcoma is a highly malignant tumour with a poor prognosis. Localization in a rib accounts for 10% of the cases (2) and 10% of all malignant rib lesions are Ewing's sarcomas (2). The fast majority of the malignant costal tumours in children consists of Ewing's sarcoma (11).

Recently special treatment regimens consisting of cyclical combined cytotoxic chemotherapy with radiotherapy caused improvement in

disease-free-survival, and, according to the primary site ribs were found to have the more favourable immediate prognosis (9, 11).

The clinical picture is unspecific. Intermittent complaints, sometimes with a "pleuritic" nature, periods of fever and other infectionlike symptoms are mentioned in the literature, including elevation of the erythrocyte sedimentation rate (ESR) (10). Intermittent pains gradually become more constant and a tender swelling develops over the affected rib. Roentgenographic examination at this time may reveal a mottled appearance of the rib caused by lytic destruction, and periosteal elevation. Also reactive bone formation may be seen, whether laminated (onion-skin) or as radiating spicules. The bony changes, however, may be easily over-

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looked, the soft tissue component being far more impressive. It consists of an excentric mainly intrathoracic but extrapulmonary mass with a spherical contour which is, in combination with the costal abnormalities, highly suggestive for Ewing's sarcoma. In rare cases pleural fluid, a haemothorax or a Horner's syndrome can be found (16). In adults systemic symptoms are almost absent and the soft tissue swelling is less extensive, but otherwise the findings are the same as in children (7).

Because of the intermittent character of symptoms, the excentric location of the soft tissue mass, and the often subtle rib lesion the diagnosis is frequently delayed for several months. Therefore, already in 1956 Sherman (12) stated that the radiologist should be alert to the occurrence of malignant disease even in circumstances in which such a possibility does not seem likely.

Material and methods

During the last 2½ years we encountered 7 children with malignant rib disease. They all had their initial investigations elsewhere in the Netherlands. They were referred to the Pediatric Oncological Centre Amsterdam (Emma Children's Hospital) after the malignant character of their disease had been established. All proved to have a Ewing's sarcoma. In some cases the diagnosis was clear from the beginning, while in other cases problems raised in establishing the definite histological diagnosis. In these cases the material was reviewed by the Dutch Committee on Bone Tumours and by the pathologist of the Pediatric Oncological Centre.

Case I (fig. 1) — November, 1978

An 11-year-old boy was admitted with pain in the left upper thorax and arm, gradually increasing during the last 6 months. Three months ago there was a short period of high fever. A chest film showed a large extrapulmonary mass in the left upper thorax and a lesion of the 5th rib. There was broadening of the rib, with an irregular trabecular structure, the cortex being intact. A biopsy confirmed the radiological suspicion of Ewing's sarcoma.

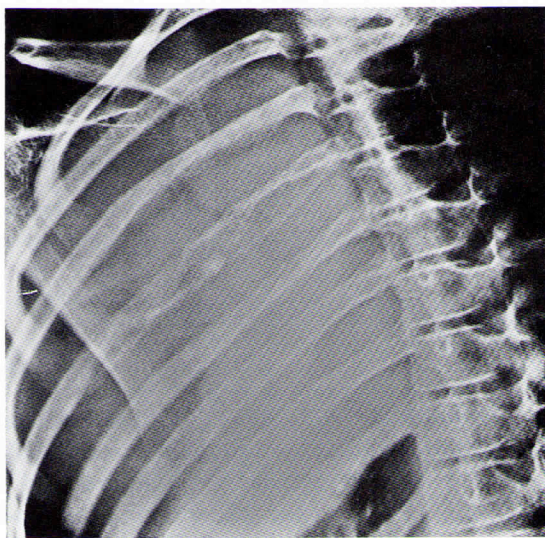


Fig. 1. — Case I: 11-year-old boy. The 5th left rib is broadened and the bony structure is irregular. No periosteal elevation or reactive bone formation is visible. There seems to be some erosion of the 4th and 6th ribs. Note the large intrathoracic soft tissue mass.

The patient was treated with combined cytostatic therapy, consisting of vincristin, actinomycin D and cyclophosphamide. During the following 4 months the tumour gradually decreased in size and was subsequently surgically removed with partial resection of the 3rd, 4th, 5th and 6th rib. A scoliosis developed afterwards, but apart from that the course was uneventful.

Now, more than two years after the operation, the patient is well without signs of local recurrence or metastasis.

Case II — April, 1979

A 9-year-old boy presented with a 5 months' history of pain in the right axilla and ribs. He had noted a swelling on the right thoracic wall. Roentgenographic examination revealed a lesion of the right 4th rib with marked soft tissue swelling. The tumour was surgically removed, followed by radiation therapy. The histological diagnosis was lymphocytary thymoma. 4 Months after the operation a lesion in the right distal femur was discovered which proved to be of the same histological type. A metastasis in the left lung was also found.

The patient was then referred to the Emma Children's Hospital and the pathological specimen was reviewed: Ewing's sarcoma.

The boy was treated with radiotherapy on the right femur (4320 rads) and cyclical combined chemotherapy. 16 Months later radiation was also necessary for a paralumbar metastasis.

sis, which caused severe irradiating pain. The cytostatics were continued.

Now, 4 years after the operation, the patient is in a reasonable condition without signs of local recurrence or metastatic activity.

Case III (fig. 2) — October, 1979

A 15-year-old girl had noted a swelling at the inner side of her left upper leg, just beneath the groin, since one week. There were muscular pains and there was walking pain. No cough, no fever, ESR was 37 mm per hour.

Roentgenographic examination revealed a rapidly increasing pleural effusion in the right hemithorax and lytic areas in

several vertebral bodies. In between a needle biopsy of the sub-inguinal lymph node revealed embryonal rhabdomyosarcoma or Ewing's sarcoma.

After a spindle shaped soft tissue swelling had been noted on the right thoracic wall, an irregular lytic lesion of the right 6th rib was discovered retrospectively on the initial chest film.

A diagnosis of Ewing's tumour with skeletal, bone marrow and lymph node metastasis was made and the patient was subsequently treated with vincristin, actinomycin D and cyclophosphamide, during one year. Radiotherapy was applied to the vertebral column up to 3500 rad.

The patient died 3 months after the end of the cytostatic therapy, with widespread pulmonary metastases.

Case IV (fig. 3 and 4) — November, 1979

A 12-year-old girl presented with abdominal complaints possibly due to a pleuropneumonia at the left side. On pleural suction bloody fluid was obtained. Intercoastal arteriography was suspicious for a-v-fistula and embolisation of the left 9th intercostal artery was performed. No rib lesions were seen at the time.

On histological examination of several pleural punctures some malignant looking cells were found once. The lesions

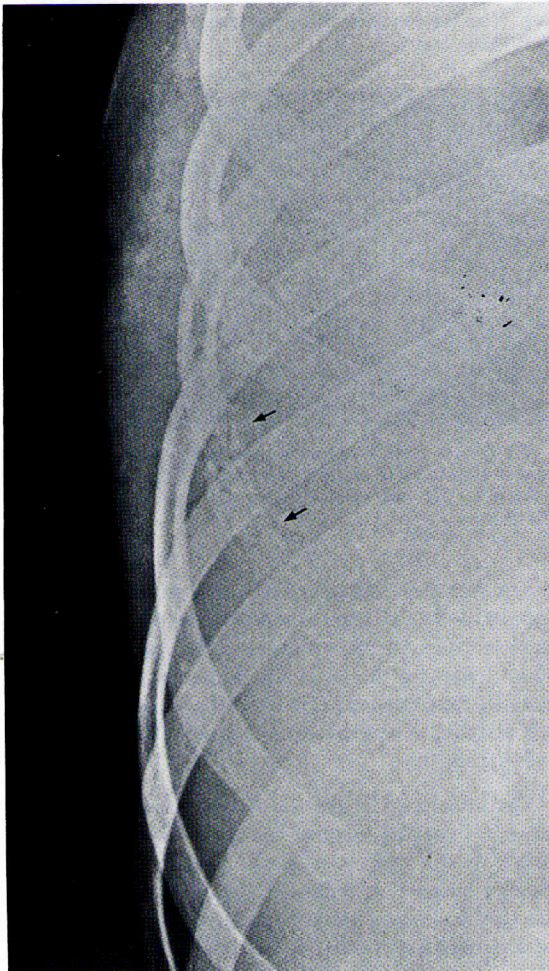


Fig. 2. — Case III: 15-year-old girl. Detail of the initial chest film. The right anterior 6th rib shows a mottled appearance with cortical destruction. Soft tissue swelling outside the thoracic wall.

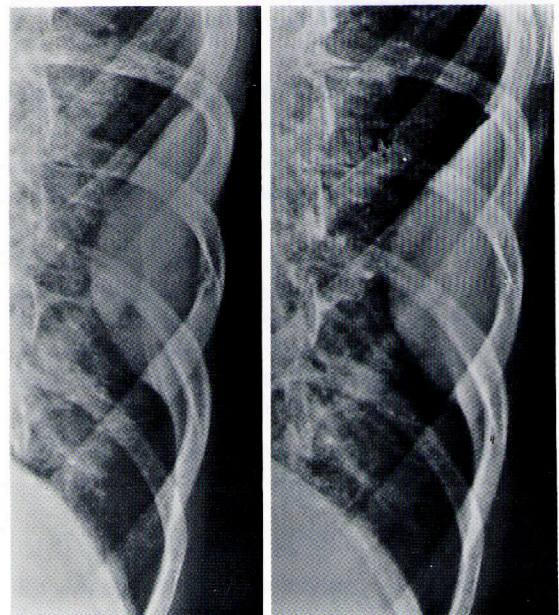


Fig. 3. — Case IV: 12-year-old girl. Oblique detail films showing the intrathoracic soft tissue mass. There is a small lytic area with cortical destruction at the inner side of the rib. This lesion is easily overlooked on a posteroanterior chest film.

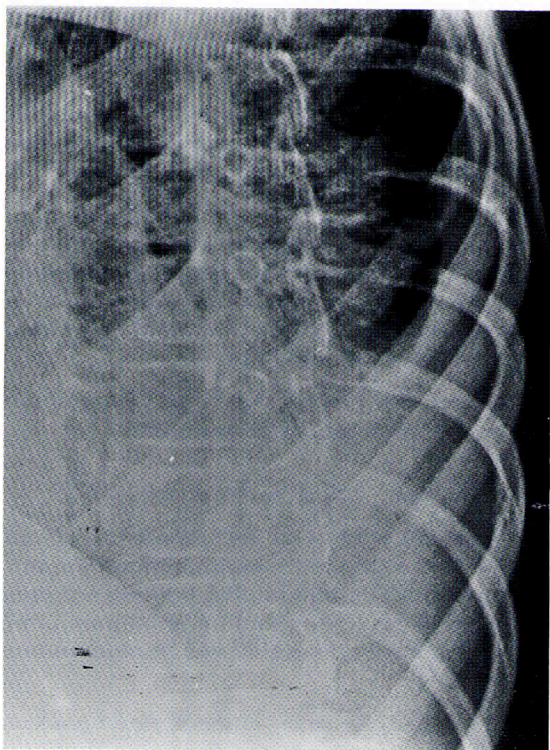


Fig. 4. — Case IV: Detail of one of the initial oblique views obtained to study a possible atelectasis after pleural suction. The tiny rib lesion can be seen in retrospect.

on the chest X-ray decreased, but, 2 months after onset of symptoms they increased again.

Rib detail films showed a small lesion of the left 9th rib, which could be noted in retrospect on one of the original oblique views.

A biopsy was performed at the side of the lesion: Ewing's sarcoma. The patient was treated with a combination of vincristin, adriamycin, endoxan and actinomycin D, and later on with vincristin, actinomycin D and cyclophosphamide. She had also radiotherapy, 2000 rad over the left hemithorax with a surdosage up to 6000 rad on the 8th and 9th rib.

The girl came in a complete remission and is in good health now, without any evidence of metastatic spread.

Case V (fig. 5A and 5B) — December, 1979

A 15-year-old boy presented with a lesion of the right brachial plexus and a Horner's syndrome. 7 Months previously he had been treated with physiotherapy because of complaints of the right shoulder. On admission a tumour was palpable in the right supraclavicular region. Roentgenographic examination revealed a soft tissue mass in the right upper

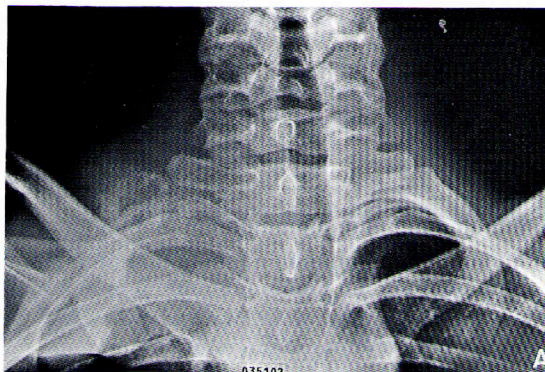


Fig. 5A. — Case V: 15-year-old boy. The posterior part of the right 1st rib shows an irregular trabecular structure with cortical destruction and areas of new bone formation in the extrathoracic soft tissues. The 2nd rib shows periosteal layering. Note the so-called extrapleural sign of the intrathoracic soft tissue mass.

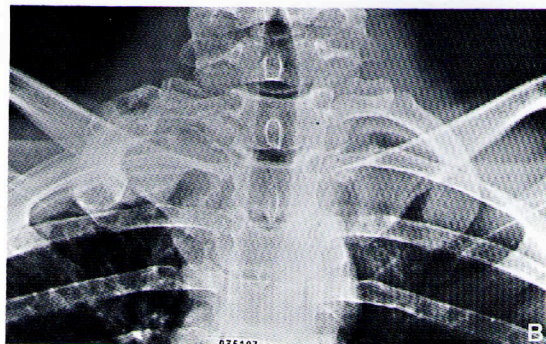


Fig. 5B. — Case V: Months later, after radiation and chemotherapy, there is marked regression of the tumor on both sides of the rib with recalcification and partial restoration of the cortex.

thorax with slight displacement of the trachea. The 1st rib showed patchy osteolysis with destruction of the cortex and bone formation in the soft tissues at the cranial side of the rib. A biopsy of the lesion was performed and the tissue obtained contained malignant mesenchymal cells. A histological diagnosis of neuroblastoma or small cell carcinoma was made at the time.

The pathologists of the Dutch Committee on Bone Tumours reviewed the specimen and considered rhabdomyosarcoma, alveolar type, or, in case of a primary bone tumour, Ewing's sarcoma. A definite diagnosis of atypical Ewing's sarcoma was made and subsequently the boy was treated with cytostatics and radiation therapy (4000 rads with surdosage up to 6400 rads). Fibrosis developed in the cervical region with impaired movement of the head, but the roentgenographic image showed recalcification of the rib and diminishing of the soft tissue mass.

Skeletal metastases developed and the patient died one year after the beginning of the therapy, of intracranial extension of a metastasis in the base of skull.

Case VI (fig. 6, 7 and 8) — April, 1980

An 11-year-old girl presented with a three weeks' history of malaise, loss of appetite and weight, cough and pain in the right hemithorax. During evening and night she had fever and severe perspiration. ESR was 40 mm per hour.

Half a year previously there had been a short period of fever and sudden pain in the right shoulder. A diagnosis of bronchitis had been made at the time and the child was treated with antibiotics.

On admission physical examination revealed signs of pleural effusion on the right side, severe emaciation, anaemia and high temperature.

A posteroanterior chest film showed "elevation of the right hemidiaphragm", a large pleural effusion and lytic destruction of the lateroposterior part of the right 9th rib. Retrospectively the lesion could also be seen on the initial film.

After the diagnosis of Ewing's sarcoma had been established, treatment with cytotoxic chemotherapy started. 5 Months later the tumour had diminished in size, with recalcification of the rib lesion.

The tumour was surgically removed then, the chemotherapy was continued. Now, almost one year postoperatively, the patient is well without obvious recurrence or metastasis.

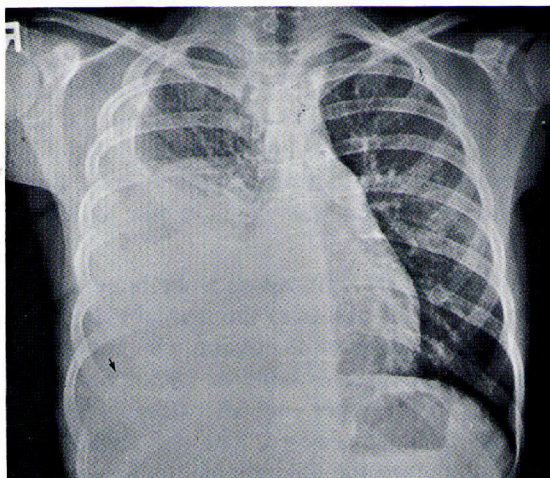


Fig. 6. — Case VI: 11-year-old girl. Posteroanterior chest film on admission. There is a pleural effusion with a big mass in the right lower hemithorax with lytic destruction of the posterolateral part of the right 9th rib.

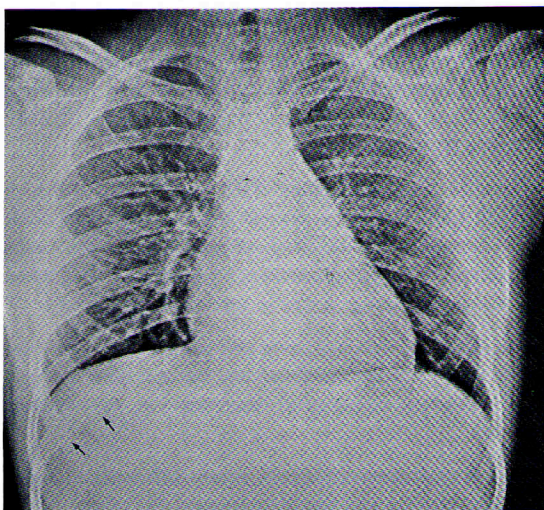


Fig. 7. — Case VI: First chest film made 6 months before that of figure 6. Broadening of the right posterior 9th rib with central osteolysis was initially overlooked. Even retrospectively the rib lesion can hardly be seen.

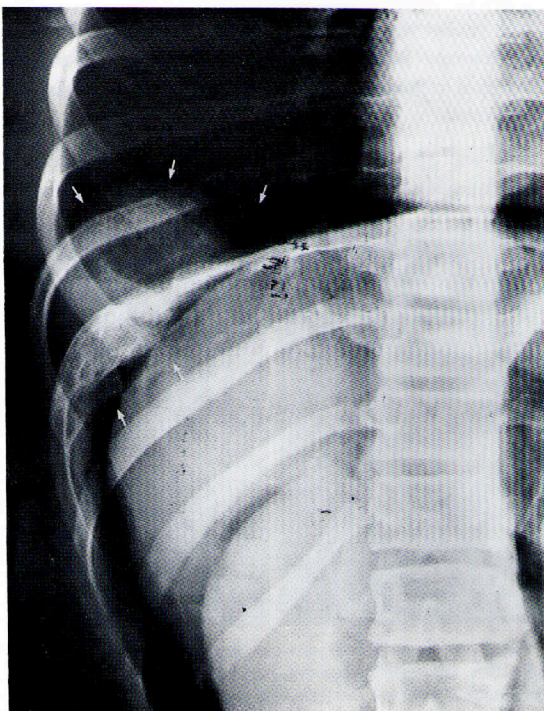


Fig. 8. — Case VI: Preoperative examination. After 5 months of chemotherapy, the pleural fluid has disappeared, the tumour mass is markedly decreased in size, and the rib shows recalcification. The caudal border of the tumour is nicely outlined by a iatrogenic pneumoperitoneum.

Case VII (fig. 9) — May, 1980

A 9-year-old boy was presented by his mother who had noted a paravertebral swelling on the left side two weeks before. There was no history of trauma, nor pain, or fever. ESR was 20 mm per hour.

Detail films showed a lesion of the posterior part of the left 10th rib with extensive periosteal reaction and a soft tissue mass. A presumptive diagnosis of osteomyelitis was made, but cultures of the pleural suction were negative. The lesion was rapidly increasing in size in the following 3 weeks.

The child was operated upon and partial costectomy was performed. A tumour was found and the operation was radical because of extension of the lesion into the vertebral column.

On histological examination a poorly differentiated highly vascular sarcoma was found, suggesting botryoid rhabdomyosarcoma, or liposarcoma. The material was reviewed and a final diagnosis of atypical Ewing's sarcoma was made.

In the same period the chest film showed a round lesion in the right middle lobe and two other lung metastases were found on CT.

The patient was treated with cyclical combined cytotoxic chemotherapy. Radiotherapy was given on the left hemithorax with a tumourdose up to 3800 rad. The lung metastases disappeared.

One year after admission the child is in a good general condition without signs of local recurrence or metastasis.

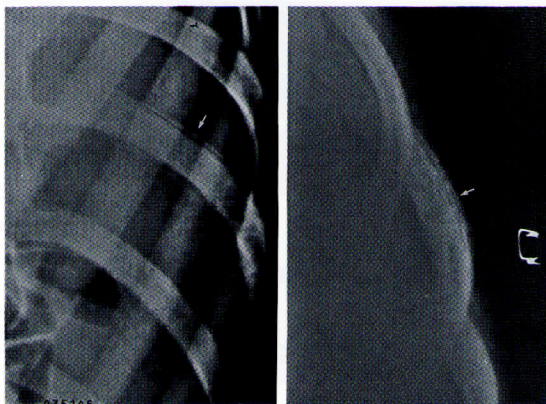


Fig. 9. — Case VII: 9-year-old boy. Irregular lesion of the left 10th rib with extensive periosteal reaction. Vertebral involvement can not be seen, but was found at operation.

The following table gives some data from the patients presented here as well as from previously published case reports.

Discussion

When this material was presented at the ESPR Oslo meeting, 1981, Dr Crispin¹ said he had the impression of an increasing frequency of Ewing's sarcomas of the rib. This remark emphasizes the importance of good quality X-ray examination of the chest in every day practice. Only after having detected a rib lesion the radiologist can come to the differential diagnosis of this rib lesion! Every young resident should learn how to look at ribs on posteroanterior chest films. This is not only worth while for the older age group with frequently occurring metastasis and myeloma; in the pediatric age group neurogenic tumours, Ewing's sarcoma and osteomyelitis are not infrequent.

Because of the relative smaller differences in tissue density in children, the different parts of the skeleton including the lower ribs under the level of the diaphragm can be seen better than in adults. Many times, however, rib lesions are easily overlooked, or are not visible at all on a pa-view. Many authors recommend overpenetrated detail films, but when shall we make them? The recognition of a soft tissue mass in relation to the thoracic wall and/or the presence of "pleuritic" complaints make penetrated detail films of the ribs concerned, obligatory.

In most cases Ewing's sarcoma can be differentiated from osteomyelitis by the coexistence of destruction and repair in the osteomyelitic bone. Rib lesions in relation to neurogenic tumours mainly show signs of arrosion rather than destruction. In rare circumstances a very extensive aneurysmal bone cyst may cause differential diagnostic problems (5). A good differential diagnostic survey is given by Weigel (14).

In our case V there seems to be bone formation in the soft tissues at the cranial side of the rib. It is a matter of discussion whether we are dealing with reactive bone or tumor bone formation. In the latter case an osteogenic sarcoma has to be considered. The soft tissue mass protruding into the thoracic cavity, however, is in favour of Ewing's sarcoma as is the relative infrequency of osteogenic sarcoma of the rib.

1. From the Hospital for sick children, London.

Table. — *The references are numbered according to the list of references. Our own cases are numbered in Romans. The table shows the relative frequency of intermittent complaints and infection-like symptoms and gives also an impression of the difficulties in establishing the histological diagnosis*

Reference	(1)	(1)	(1)	(2)	(2)	(2)	(2)	(2)	(3)	(4)	(4)	(6)	(8)	(10)	(14)	(14)	(14)	(14)	(15)	I	II	III	IV	V	VI	VII
Age	15	10	10	14	14	16	13	16	12	10	11	4	8	12	10	8	12	8	15	11	9	15	12	15	11	9
Sex	m	m	m	m	m	m	m	f	m	f	m	f	m	m	m	m	f	f	m	m	m	f	f	m	f	m
Rib(s) involved	6	6	6	8	4	9	7	11	7-9	2	3	6	5	8	10	5	6	1	7	5	4	6	9	1	9	10
Posterior, lateral, anterior	1	1	p	p	a	p	1	p	1	a	a	a	a	a	p	a	a	a	1	p	a	a	1	p	p	p
Period of latency (months)	—	3	—	12	4	3	2	2	?	2	1	11	4	2	—	—	9	5	4	6	5	2	2	7	6	—
Intermittent complaints	—	—	—	+	+	—	+	+	—	+	—	+	+	—	—	—	—	+	+	+	—	—	—	+	+	—
"Infection", fever, and/or elevated ESR	+	+	—	+	—	—	+	+	+	+	+	+	+	+	—	—	—	+	—	+	—	+	+	+	+	+
Difficult pathological diagn.	+	+	+	—	+	—	—	+	+	—	—	+	—	—	—	—	—	+	—	—	+	+	—	+	—	+

Swenson (13) stated in 1943 that Ewing's sarcoma shows great variations in roentgenographic (skeletal) manifestations as well as in its histopathological picture. Apart from the question if this is true for all localizations, this statement seems applicable to our present material as can be seen from the comparison of the figures 1, 2, 3, 5A, 7 and 9. But the combination of all roentgen findings including the large excentric soft tissue mass, is of great help in establishing the diagnosis of a Ewing's sarcoma of the rib. Sometimes the roentgenographic image may even lead the pathologist to the final diagnosis (16), as in Weigel's case 4 and our case III.

The recognition of the extrapleural character of an intrathoracic mass is of special interest to the surgeon. An extrapleural biopsy must be taken without contaminating the pleural cavity.

Conclusions

The study of 7 recently encountered cases of Ewing's sarcoma starting in a rib and the comparison of the data obtained with those from the literature lead us to the following conclusions:

1. Intermittent and/or infectionlike complaints do not exclude underlying malignant disease.
2. Every mass of unknown origin in relation to the thoracic wall makes detail films of the ribs in at least two directions necessary.
3. Information about the radiological suspicion of a Ewing's sarcoma of a rib is of great help to the surgeon who should take an extrapleural biopsy and also to the pathologist who may have serious difficulties in establishing the histological diagnosis.

Résumé

Sept patients présentant une tumeur maligne au niveau costal ont été envoyés ces deux dernières années et demie au Centre Oncologique pour enfants d'Amsterdam, dépendant de l'Hôpital Pédiatrique Emma. Un sarcome d'Ewing a été mis en évidence chez tous ces patients. Leurs antécédents cliniques sont relatés et les observations radiologiques discutées en accordant un intérêt particulier aux renseignements

cliniques et radiologiques initiaux. Ceux-ci sont comparés aux données contenues dans les rapports individuels déjà publiés.

Il n'est pas rare que le diagnostic soit retardé de plusieurs mois du fait du caractère intermittent des symptômes, de la localisation irrégulière de la masse de tissu mou et de l'identification souvent difficile des lésions costales. C'est pourquoi il est nécessaire dans la pratique courante de soumettre le thorax à des examens radiologiques de bonne qualité. L'identification d'une masse tissulaire molle au niveau de la paroi thoracique, s'accompagnant ou non de plaintes «pleurétiques», rendent indispensables des prises de vues détaillées et en profondeur de la région costale en question. La combinaison d'une masse intrathoracique et extrapleurale importante avec des anomalies costales localisées plaide fortement en faveur d'un sarcome d'Ewing.

Les renseignements radiologiques faisant soupçonner la présence d'un sarcome d'Ewing au niveau d'une côte sont d'un grand secours pour le chirurgien, qui devrait alors effectuer une biopsie supplémentaire, ainsi que pour le pathologiste, qui peut éprouver de sérieuses difficultés à établir le diagnostic histologique.

Samenvatting

In de loop van de voorbije 2½ jaar werden 7 patiënten met kwaadaardige ribaandoeningen verwezen naar het Pediatrisch Oncologisch Centrum van het Emma Kinderziekenhuis te Amsterdam. Bij al deze patiënten werd een Ewingsarcoom gevonden. De klinische geschiedenis van deze gevallen wordt nagegaan en de radiologische bevindingen worden besproken. Bijzondere aandacht wordt geschonken aan de initiële klinische en radiologische gegevens. Deze gegevens worden vergeleken met gegevens die gepubliceerd werden in vroegere gevallenstudies.

Vanwege de intermitterende aard van de aandoening, de excentrische lokalisatie van de zachte weefselmassa en de vaak subtiele riblaesies duurt het niet zelden meerdere maanden voordat de juiste diagnose gesteld wordt. In de dagelijkse praktijk is een kwalitatief goed radiologisch onderzoek van de thorax dan ook een noodzakelijkheid. Het herkennen van een zachte weefselmassa tegenover de thoraxwand en/of de aanwezigheid van „pleuritische” klachten maken een detailopname van de betrokken rib noodzakelijk. Het samengaan van een grote intrathoracale extrapleurale massa en plaatselijke ribafwijkingen is zeer suggestief voor een Ewingsarcoom.

Informatie in verband met het radiologisch vermoeden van een Ewingsarcoom is van groot belang voor de chirurg die een extrapleurale biopsie moet nemen, evenals voor de anatomopatholoog die voor ernstige moeilijkheden kan staan bij het stellen van een histologische diagnose.

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