In their volume *Tumours of Bone*, Geschickter and Copeland describe 65 cases of Ewing sarcoma, of which only one occurred in the ribs. The inference might be that such a location is rare, but we are inclined to believe that the rarity is only apparent, since no less than 4 instances have been observed in our own small material. These cases seem to merit description, more especially as they throw a certain amount of light on the clinical features and pathology of the growth.

**Case 1:** A boy of fifteen became suddenly ill, with stinging pains in the right half of the thorax. There was swelling in the region of the pain, and x-ray examination indicated changes extending over 13 cm. of the 6th right rib. The bone had become thinned in patches, alternating with denser areas. A tumor the size of a fist was observed in the adjacent pleura, extending into the space between the lower and middle lobes.

Two weeks after the onset of symptoms the patient was operated upon (Sept. 3, 1930). It was found that the tumor bulged into the pleural cavity, but did not adhere to the lung. It also protruded between the 5th and 6th ribs, which were resected with it. The tumor weighed 500 gm., was lobulated, and extended along the ribs in the manner characteristic of Ewing sarcoma in the long bones.

A roentgenogram of a section of the specimen showed the characteristic increase in volume and density of the rib, and a roughness of the surface toward the pleura. The outer surface, on the other hand, was smooth and well defined. Microscopic examination showed that the rib was entirely surrounded by masses of tumor, but that the periosteum
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FIG. 2. CASE I: EXTRAPATED TUMOR, WITH THE 6TH RIB

The rib section shows increased volume and density. The surface toward the pleura is rough, with spicules. The outward surface, on the other hand, is smooth. Bone formation has occurred within the tumor.

was lifted only where the surface was rough, the roughness being due to newly formed bone spicules. New bone formation radiating towards the rib could be observed in the tumor tissue.

Microscopically the growth consists of small, uniform cells containing but little protoplasm and a round, deeply stained nucleus showing a definitely limited membrane. No distinct cell boundaries can be seen. In the tissue are occasional cavities lined by tumor cells. The relation between the tumor cells and the vascular interstitial connective tissue
differs in different parts of the tumor. In some areas irregular masses of tumor cells are surrounded by strands of connective tissue with no collagen or reticular fibers between the cells. Elsewhere the connective tissue is edematous and resembles embryonic connective tissue, while the tumor cells appear to be undergoing a change in this direction. When stained by special methods these areas show fine reticular fibers, not only in the interstitial connective tissue but also among the tumor cells.
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The tumor cells give an impression of being changed into embryonal connective-tissue cells.

In some areas the vessels are of peculiar appearance, their walls being distended and rich in cells, while the lumen is narrowed. These vessels lack elastic tissue and smooth muscle. The tumor cells are frequently arranged around these vessels, and shrinkage may then produce pseudo-papillary formations in the embedded preparations.

No bone formation on the part of the cells has been observed. The bone formation within the tumor is apparently of periosteal origin.
The earlier film (left) shows metastases the size of walnuts in the right lung. At the time the second was made these had all disappeared.

This case was reported in 1932 as a mesothelioma. The patient was then free of recurrences or metastases. An x-ray examination in December of that year, however, revealed a number of metastases in the right lung, some of which were 4 or 5 cm. in diameter. These gradually regressed under x-ray treatment and in six months had entirely disappeared. In view of the good results the tumor was again examined, and the diagnosis changed to Ewing sarcoma.

The patient remained well until September 1935, when he returned with symptoms referable to the right pleura. X-rays disclosed a shadow over the base of the right lung, the nature of which could not be determined. Following roentgen therapy the symptoms disappeared, and in March 1936 had not recurred.

Case II: F. S., a boy of ten, was admitted Nov. 19, 1934. He had been listless and pale during the preceding months but had attended school. Eight days before admission he had become ill, with a temperature of 39°-40° C. but without local symptoms. The temperature gradually fell to about 38°. The patient complained of slight pain in his left side. The chest over the entire left lung was dull to percussion. Puncture yielded only a few drops of pus. A tumor in the left axilla was discovered on Nov. 22.

The tumor was at the level of the nipple in the axillary line, was the size of a hen’s egg, soft, but not fluctuating. Its limits were difficult to determine and it could not be moved over the underlying tissue. The skin was not adherent.

On x-ray examination changes were observed on the inner side of the 6th rib for a distance of 2 cm., immediately beneath the soft axillary swelling. The contour was unevenly thickened by layers of spicule-like deposits. A periosteal sarcoma was first suspected, but tuberculosis could not be excluded.

On Dec. 4, 1934, about 12 cm. of the 6th and 7th ribs were resected. As soon as the skin and the outer layer of muscles were cut through, a soft tumor could be seen bulging out between the ribs. The pleural cavity was found to contain a tumor as large as a fist, apparently arising from the costal pleura. It was so friable that it fell to pieces as soon as it was touched. Two isolated tumors, the size of a cherry, were found on the surface of the lung. As the tumor was apparently connected with the 7th as well as the 6th rib, both were resected at the same time. Three months later the patient had gained weight, was feeling well, and was quite active. Ten months after operation he was free of symptoms and x-ray examination was negative.

Fig. 10 is a roentgenogram of 4 sections of the tumor, showing the same increase of
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FIG. 10. CASE II: ROENTGENOGRAM OF SECTIONS OF THE 6TH RIB AND THE TUMOR

Attachment of intercostal muscle at A. At this point the periosteum is lifted. Spicules (S) have formed only where the periosteum is elevated. The outward surface of the rib (B) is smooth, although the tumor tissue is infiltrating the outer portion of the rib. P = periosteum. T = tumor tissue.

density of the bone as was seen in Case I. Here, too, the outer side is smooth and the pleural surface rough, with spicules.

The microscopic findings are identical with those in Case I. The periosteal deposits on the bone can be distinctly seen and here the periosteum was loosened from the bone. This was obviously the point of attachment of the intercostal muscles on the rib.
In March 1936 the patient was still alive, but with multiple metastases in the lungs and skeleton.

CASE III: S. O., a boy ten years of age, had a tumor on his back. It was first noticed three weeks before admission, by his mother, who thought it had not grown since. The child's only complaint was local pain when "flinging his arms about."

Medially to the left scapula, at the level of the 3rd and 4th ribs, was a rounded area of resistance the size of an egg, over which the skin was freely movable. It was firm in consistency, and the surface was slightly uneven. The leukocyte count was 7,300.

X-ray examination revealed a lobulated shadow in the center of the lung field on the left side, extending from the second to the fifth intercostal in front across the field. A lateral view showed the tumor to be situated at the back, and its lobulated contour was here very distinct. The 6th and 7th ribs were forced apart, most widely at a distance of about 5 cm. from the vertebral column. The ribs showed a distinct pressure effect, the margins having become concave.

X-ray treatment brought about progressive reduction in the size of the tumor, and about five weeks after irradiation was instituted operation was done. An incision was made above the tumor in the 6th interspace. The tumor was found to be situated along the ribs, and to be movable and well encapsulated. It was isolated up to the point of entry into the 6th interspace. The 6th and 7th ribs were then cut some distance beyond what was assumed to be the lateral limit of the intrathoracic tumor. An incision was made in the 8th interspace, and 7 cm. of the 8th rib, from the transverse process outwards, were resected. As the tumor seemed to extend up towards the 5th interspace, it was decided to include this, also. The 6th and 7th ribs were divided medially, close to the transverse processes. The tumor proved to be adherent both to the lower and upper lobes of the lung, and in separating it a superficial layer of the lung was also taken. The operation apparently went some distance beyond the tumor tissue on all sides.

Grossly the operative specimen was an oval-shaped mass about 8 × 5 cm. Its pleural surface was in general smooth, but coarsely tubular. On cross-section the tumor was seen to have grown out like a pocket from the space between the 6th and 7th ribs, both towards the pleural cavity and the inner surface of the 6th rib. This pocket had thus been formed between the tumor and the parietal pleura.

A roentgenogram of a cross section of the operative specimen shows the 6th rib twisted upward and the 7th downward. The outer surfaces of both were smooth and the pleural...
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surfaces rough. A spontaneous fracture of the 6th rib was observed, a rare occurrence in Ewing sarcoma. This may possibly have been due to manipulations during operation.

Microscopic examination showed that the tumor tissue was largely replaced by loose connective tissue. In certain parts it remained unchanged, however, and had the same appearance as in Cases I and II. There was no tumor tissue in the ribs. The cortex on the inside of the 7th rib was pressed in.

It is impossible to say definitely whether the tumor had originated from the 6th or the 7th rib; probably the 6th.

CASE IV: J. G., a fourteen-year-old boy, repeatedly complained of pains in his right shoulder during October and November 1934. The pains occurred only during effort and disappeared on resting. In the spring and summer of 1935 similar pains occurred on exertion, and the boy was short of breath, tired, and faint. He lost weight and complained

of feeling hot and of perspiring. He had pain in the right half of the thorax and a dry cough. At the beginning of September 1935 the patient noticed that the right half of the thorax was increasing in size and dyspnea became increasingly severe. The pain disappeared as the other symptoms grew worse.

Examination on Oct. 3, 1935, showed displacement of the heart towards the left. The thorax was much deformed, the right half bulging forward. Percussion gave a wooden dullness over the whole area of the right lung. No respiratory sounds were audible here. The liver was palpable and slightly tender. There were changes in the urine. The temperature was elevated. The leukocyte count was 25,700.

X-ray examination, Oct. 5, showed a diffuse, intense shadow over the whole right lung field, without any discernible details. The heart and mediastinum were displaced considerably towards the left, even the right main bronchus being wholly to the left of the median line. The roentgenogram showed what looked like a copious exudate, but this may conceivably have been due to a massive tumor in the pleura.

On biopsy tumor tissue was obtained made up of cells with closely lying, uniform nuclei, fairly rich in chromatin, and displaying mitoses. The protoplasm in the cells was hardly discernible. The tumor cells were arranged in heaps in the meshes of an abundant and regular uniform network of vessels. No connective tissue or reticulin fibers could be seen between the cells with Perdrau and Mallory stains. There was no bone formation. In some areas the vessels were abundant. They were of a rather primitive character, having no boundary between intima, media, and adventitia. The walls of even fairly large vessels
consisted of a layer of voluminous cells with large nuclei, and an outer abundant layer of connective tissue without elastic fibrils or muscle. Arteries and veins could not be differentiated. The difference between capillaries and larger vessels was also very indistinct. The thoracic wall was infiltrated by the tumor tissue. Diagnosis: Ewing sarcoma.

On Oct. 12 the patient was extremely dyspneic and the temperature reached 40.5°. A blood count showed hemoglobin 68; red cells 3,420,000; white cells 28,600 (81 per cent neutrophils, 1 per cent eosinophils, 1 per cent basophils, 3 per cent monocytes, and 14 per cent lymphocytes).

On the same day a roentgenogram showed fairly extensive destruction of the 8th rib, which for a distance of about 15 cm. showed peripheral erosion and a patchy disintegration of the central portion. Traces of a thin shell of bone surrounded the involved area. Along the 7th rib was thick periosteal new bone formation extending for a distance of 10 to 15 cm. along the lower edge, but in one place involving the entire circumference of the rib. No destruction of the spongiosa was demonstrable. Diagnosis: Ewing sarcoma.

Following x-ray therapy the patient felt better and breathed more easily. A blood count on Oct. 20 showed 3,300,000 red cells and 21,300 white cells. Two days later the red cell count was 3,380,000 and the white count 16,000 (neutrophils 71 per cent, eosinophils 7, basophils 0, monocytes 7, lymphocytes 15). The urine was negative for Bence-Jones albumen. The diazo reaction was negative.

X-ray examination, Oct. 28, after a period of x-ray treatment showed considerable regression of the process in the right half of the thorax. The heart and mediastinum had resumed almost normal positions, and the right lung field in the upper median part showed well aerated portions sharply differentiated from the process in the pleura. The 8th rib showed little change in appearance, but no progress of the lesion since the previous examination was demonstrable. The periosteal bone formation along the seventh rib was unchanged, and this rib still did not show any distinct signs of destruction.

On Oct. 30 a blood-count showed red cells 3,900,000; white cells 10,800 (neutrophils 73 per cent, eosinophils 2, monocytes 5, lymphocytes 20). On Nov. 4 continued improvement was reported. The boy was no longer short of breath and the bulging of the right half of the thorax had diminished. The temperature was sub-febrile.

**Discussion**

These 4 cases are much alike. The patients had just reached or were about to reach the age of puberty. The tumors were localized in the 6th to the 8th ribs, and always at the back. They grew into the pleural cavity like

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**Fig. 14. Case IV: Roentgenogram of Right Half of Thorax; Changes in the 8th Rib**
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A sponge, pushing the pleura away. The tumor tissue had lifted the periosteum on the pleural surface of the ribs, and this process ceased at the attachment of the intercostal muscles. As a consequence, the inside of the rib was rough, with periosteal spicules, while the outside was smooth, although tumor tissue had infiltrated all around the bone. The tumor tissue had also filled the haversian canals throughout the thickness of the rib.

The fact that the periosteum was elevated only on the inside does not, of course, necessarily mean that the tumor had originated on that side, but only that the periosteum is more easily loosened there. Neither can any conclusions be drawn from the fact that the tumors proliferated chiefly into the pleura.

The localization of the tumors to the posterior parts of the 6th-8th ribs is noteworthy, this being the site of earliest ossification. The time when ossification begins in the bones in which Ewing sarcoma occurs was therefore examined.

A study of the cases published by Geschickter and Copeland and by Connor show that in practically all instances the tumors occur in those parts of the skeleton where ossification begins towards the end of the second month of fetal life (Keibel and Mall). This conforms with observations previously made that this form of malignant growth occurs primarily in the shafts of the long bones and never in the epiphyses. Geschickter has also published 19 cases in which the tumor occurred in the maxillae, which are formed at the same early stage but are not preformed in cartilage.

Only a few cases are not in accord with this rule, and of these, several are doubtful. One of Connor’s cases occurred in the ischium, but of this he says: “The section was too poor for accurate diagnosis.” Another case in the os calcis was variously diagnosed. Connor’s own conclusion was that this was a “good example of the angio-endothelial type of bone tumor.” In another case in the os calcis plasma cells occurred and the possibility of myeloma is therefore not excluded, especially as there were tumors throughout the skeletal system. On the other hand the occurrence in the mastoid process, of which Geschickter and Copeland cite one case, is contrary to the general rule. It must be pointed out, however, that the squama temporalis, which is formed early, is in such close proximity that we are not justified in attaching too much significance to these cases.

Conclusions regarding the histogenesis of Ewing sarcoma might be permissible on the basis of the peculiar localization. Ewing himself considered the tumor to be an endothelioma arising in the endothelium of the lymphatics in the haversian canals. Connor suggested (1926) that the tumor arises from the reticulo-endothelial system, an opinion shared by Oberling, who therefore includes the Ewing sarcoma in his system of reticulo-endothelial tumors. Geschickter and Copeland believe that the growth originates in the intracortical or subperiosteal lymphoid tissue. Melnik rejects all these theories and maintains that the tumor is a round-cell sarcoma arising in the undifferentiated embryonal connective-tissue cells in the haversian canals.

In view of what has been said above, however, it would seem that the Ewing sarcoma might possibly be traced back to a disturbance in the formation of the skeleton at a very early stage of fetal life. This, we know, is
characterized by a condensation of the mesenchymal blastema, the cells of which ultimately form the precartilage. These early cells are very similar to the tumor cells of the Ewing sarcoma.

According to Broman the formation of bone as a rule begins in the places where the first "blastem-anlagen" are formed. A disturbance at an early state of embryonic life would then affect only those parts of the skeleton where the Ewing sarcoma is found. The marked sensitivity of these tumors to radiation would be explained if the cells are comparable to such primitive embryonal cells.

**Summary**

Four cases of Ewing sarcoma in the ribs are described, all exactly similar. The tumors originated from the posterior portion of one of the ribs, and grew like sponges into the pleural cavity, pushing the lifted periosteum and the pleura before them. The greatest length of the tumors was along the rib. The surface was coarsely lobate. The ribs presented a characteristic increase in density and volume, and spicules were formed. The latter occurred only on the inside of the rib, however, where the periosteum was elevated, but not on the outside, even though the tumor tissue grew around the rib. The loosening of the periosteum began at the point of attachment of the intercostal muscles.

In one case, treated by extirpation and x-rays, the patient is alive and well five years and a half after the operation, although metastases had occurred in the lungs two years after the operation. These disappeared under x-ray treatment.

The author believes that practically all Ewing sarcomas are localized to those parts of the skeleton in which ossification begins in the second month of fetal life. The first formation of the later ossifying blastema takes place at the same points. As the cells of Ewing sarcoma are very similar to these blastema cells, the conclusion might tentatively be drawn that Ewing sarcomas are due to a disturbance in the formation of the skeleton at a very early stage of fetal life.

**References**


1 Personal communication.