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CASE OF MALIGNANT OSSEUS TUMOR ORIGINATING
IN AN EPIPHYSIS (EWING TUMOR)

The case is that of a girl, 17 years old, who has been troubled for about 2 years with a lesion of the right hip, the clinical diagnosis of which was unsettled—chronic osteitis or tumor—till the tumor diagnosis was established by the appearance of a peripheral metastasis.

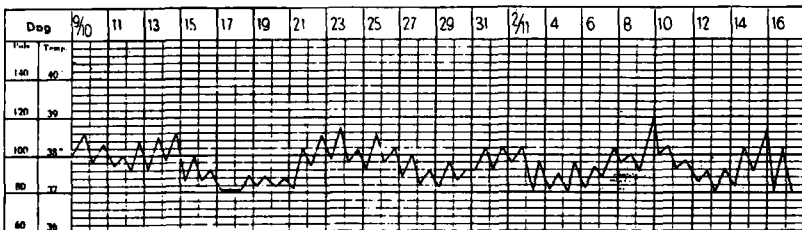
The first symptoms were noticed in July 1936, when the girl was 15 years old. She began to feel some aching in the right hip when she exerted herself. This inconvenience would last only a few days and return at intervals of about 14 days. In the autumn, same year, this inconvenient condition was aggravated. Roentgenography of the hip was performed in a hospital in August and December 1936. In spite of rest and treatment with heat, the trouble kept on; and in the spring 1937 the girl began to limp considerably during the periods of pain. Between these periods she had no trouble worth mention. She was excused from gymnastics in the school, but otherwise she was fully able to keep up with her school work.

On examination in the Stockholm V.F.A. (Orthopedic Hospital) March 23, 1937, there was found to be some atrophy of the right thigh and free mobility in the hip-joint, except for some limitation of the abduction. During the summer 1937 she worked as housemaid in the country and was fully able to do the work, except for a few days now and then when she was troubled with aching in the hip. In the autumn 1937 the aching became worse, and now the interval between the attacks was only one week. Gradually the pain became continual, merely varying

in intensity. On October 7, 1937, examination showed that the limp had increased, and the mobility in the hip was further reduced. She also noticed a slight tenderness over the trochanteric area. Since her first visit to the V.F.A., on March 23, 1937, the patient has been under continual observation, and she has been admitted periodically to another hospital for special examinations. Her illness has been varying to an extreme degree: on some visits to the hospital she has appeared rather ill, complaining over pain and tenderness in the hip, and presenting reduction of the mobility; on other visits her state of health has appeared fairly good, with only quite insignificant symptoms. This periodicity of the clinical symptoms was even manifest in the variations of the outcome of the *sedimentation test*:

10/10-37:	75 mm.	25/1-38:	10 mm.
19/10- "	: 80 "	8/2- "	: 10 "
3/11- "	: 92 "	22/3- "	: 50 "
26/11- "	: 46 "	24/4- "	: 15 "
8/12- "	: 7 "	7/6- "	: 33 "
20/12- "	: 18 "		

During her stay in the hospital from October 9 to November 15, 1937, she presented a peculiar temperature curve: for one week the temperature was about 38° C., in the next week it rose only to 37.5° at the most, and then there came a new fever week with a temperature of about 38°, and so on.



In the first days of June 1938 there appeared a rapidly increasing swelling in the left supraclavicular fossa. On June 8th, an operation was performed, with extirpation of a walnut-sized

tumor. The histological examination, carried out by Dr. Fredrik Wahlgren, gave the following result:

"The specimen received, a lymph gland, is invaded throughout by metastases of a malignant tumor. In some parts this tumor is made up of small, markedly polymorphous, undifferentiated cells that are grouped in a sort of alveolar clusters, between which there are streaks of coarsely trabecular but rather

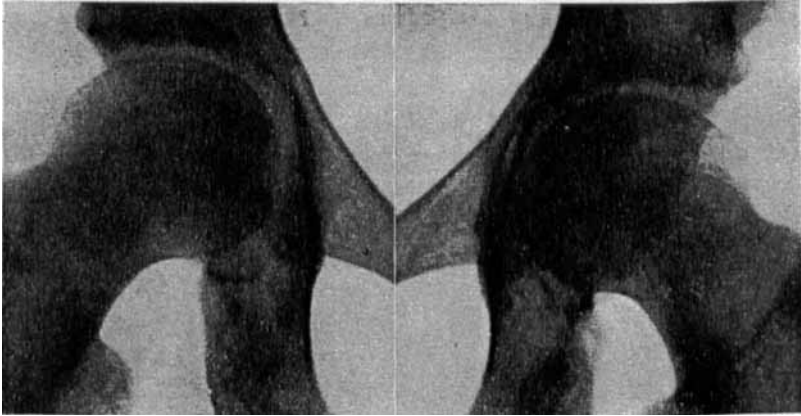


Fig. 1.

cell-rich connective tissue. Even between the cells there appears to be a delicate network of connective tissue. Other areas present an entirely different picture. Here the tissue is composed essentially of connective tissue, rather rich in cells, and this too shows an abundance of intercellular collagen substance with delicate fibres. In this connective tissue there are numerous irregular areas that look most like necrosis, but it still contains a good deal of nuclear fragments and shrunken cells. In the connective tissue there are also numerous spaces resembling blood vessels, lined with endothelial-like cells and containing red blood cells. The nature of the tumor cannot be established with absolute certainty. The differential diagnosis lies between metastasis of an osteogenous sarcoma and a Ewing tumor.

"Certain structures, which are particularly conspicuous in Mallory-stained sections, remind of the picture of a Ewing tu-

mor. Still, the cells in the solid alveolar cell-rich areas appear to be somewhat more polygonal than those usually seen in the Ewing tumor, besides their protoplasm is lighter in colour than that of the lymphocyte-like cells in Ewing tumors. The abundant connective tissue in this tumor is more rich in cells and seems

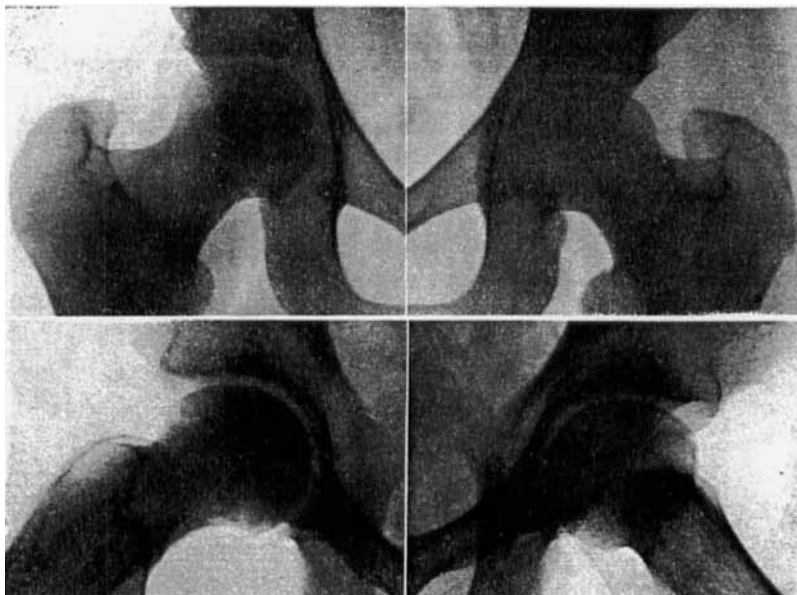


Fig. 2.

more loose than is usually the case in the Ewing tumors. Notwithstanding these deviations from the characteristic picture, the present lesion seems more likely to be a Ewing tumor than an osteogenous sarcoma."

Thus the tumor diagnosis was established, and the patient was transferred to Radiohemmet where, in July 1938, she was given intensive X-ray treatment, applied to the right hip and post-operatively to the left supraclavicular fossa.

After this clinical orientation I shall give an account of the successive roentgenological development of the tumor:

X-ray examination on August 25, 1936, showed, on comparison of the two hip-joints, only a very slight density in the head

of the right femur and a somewhat less distinct spongiosa structure within this area (Fig. 1).

X-ray examination on March 23, 1937, showed an increased sclerosis, involving the central and medial parts of the head of the femur. This process was diffuse, and no particular destruction could be made out. Like before, the joint slit was intact (Fig. 2).

X-ray examination on October 31, 1937, showed that the

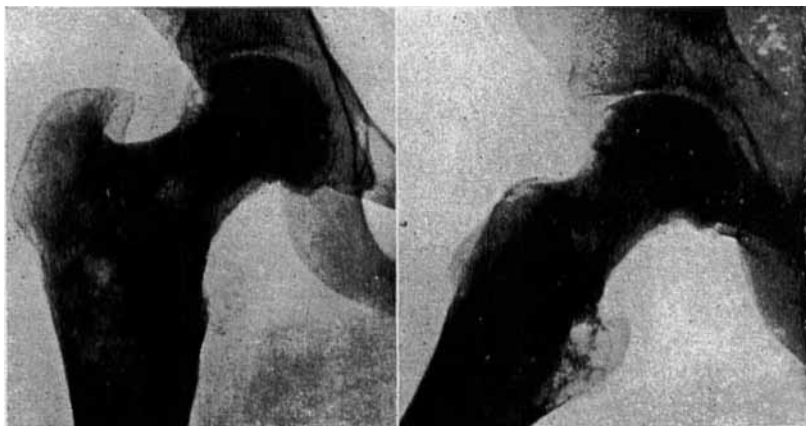


Fig. 3.

sclerosis had extended down in the neck of the femur and increased in intensity. On March 22, 1938, this process was found to involve the head, neck and trochanteric areas. It was characterized by sclerotic areas alternating with areas of focal destruction. No periosteal changes. Joint slit still intact. (Fig. 3).

In this case the process has invaded the lesser trochanter, which is undergoing destruction, so that only a thin fragmentary shell of bone is left here. The circumference of this bony shell is greater than that of the normal trochanter—a feature suggestive of expansive tumor growth, supporting directly the tumor diagnosis (Fig. 4).

Thus the roentgenological development of the lesion is characterized by the fact that the tumor appeared first in the

head of the femur, and then it successively extended down through the neck and the trochanteric areas, following the long axis of the bone—and not expanding perpendicularly on the long axis. This explains why there was no palpatory swelling. The tumor began with a diffuse sclerosis, and focal areas of destruction did not appear till later on. This roentgenological development together with the periodicity of the fever, the



Fig. 4.

variability of the sedimentation rate and the histological features of the metastasis make the diagnosis Ewing tumor most likely.

In contrast to the considerable variations in the clinical picture, the tumor has shown a constant progression in roentgenological respect.

This case is of fundamental interest because the tumor originated in the head of the femur. From the present extensive literature concerning the Ewing tumor and osteogenous sarcoma, contributed above all by American authors, it is emphasized as a regular condition, that the tumor commences in a metaphysis, at any rate never in an epiphysis. That this rule is not absolute is evident from the case here reported, which is interesting not only with a view to the localization of the initial

changes to an epiphysis, but also because of its development from a diffuse sclerosis into alternating sclerotic and destructive changes. At the present stage the differential diagnosis between osteogenous sarcoma and Ewing tumor has not been established absolutely, but there are good reasons to make the diagnosis Ewing tumor. Regardless of this distinction, the present case is of great interest because a malignant osseous tumor of this kind does never commence in an epiphysis—that is, according to the statements in the textbooks.

SUMMARY

A report is given of a malignant osseous tumor, with the diagnosis balancing between osteogenous sarcoma and Ewing tumor, although with strong clinical evidence in favour of the latter diagnosis. This case is of fundamental interest, as the tumor commenced in an epiphysis (head of the femur) wherefrom it extended successively through the metaphysis and diaphysis. This development is an entirely new experience, as hitherto it has been claimed that a malignant tumor of the kinds here mentioned cannot originate in an epiphysis.

DISCUSSION:

Bentzon, Copenhagen:

In this case Dr. Knutsson applies the term *sclerosis* to the decreased permeability to the X-rays observed as the first radiological symptom in the neck and head of the femur. Presumably we have to take this designation as meaning that there is an increase in the *lime* content. But this is something we cannot know with certainty. For the well-known structural picture presented by normal spongiosa tissue on the X-ray plate is due in part to a physical phenomenon which I shall compare to the very pronounced "clearing" (*i.e.*, accentuation of the ray intensity) that may appear on an abdominal roentgenogram when there are even quite small amounts of gas present in the intestines—a far stronger clearing than can be explained by the fact that the gas has displaced a little tissue of higher specific

gravity. The X-rays are intensified on passage from tissue to gas and from gas to tissue.

A similar physical phenomenon (intensification of the rays) appears on the passage of the X-rays from bone to fatty marrow. If the fatty marrow is replaced by a fibrous mass or some other sort of more "heavy" tissue the phenomenon is reduced considerably, and then we find on the picture a "condensation" that needs not be connected with an increase in the lime content of the bone.

In some cases of lunatum malacia this phenomenon is particularly pronounced. If operation is performed in such cases of lunatum malacia where the roentgenogram of the bone shows a very intense shadow, one will most often find a marked decrease in the lime content of the bone—but a fibrous bone-marrow.

To me it seems most likely that the initial changes in this case of Ewing's sarcoma have originated in the bone-marrow, and the roentgenographic findings lend support to this view.

Rob. Hanson

With reference to Bentzon's remark, that the cause of the density of the caput might be due to replacement of the fatty bone-marrow by fibrous tissue, I wish to call attention to the well-known fact that metastasis of malignant tumors to the skeleton shows an increased density which is not due to sclerosis—or to sclerosis alone—but to tumor tissue, possibly with capacity for bone-formation.