OSTEITIS FIBROSA OF RECKLINGHAUSEN, HETEROTOPIC PARATHYROID ADENOMA, METASTASES OF A BENIGN ADENOMATOUS STRUMA AND ADENOMA OF THE LEFT ADRENAL IN THE SAME PATIENT

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Two years ago the author had the opportunity of observing a remarkable case of pluriglandular neoplastic growth.

![Image of roentgenogram showing the metastasis from the thyroid adenoma in the left humerus.](image_url)

**Fig. 1. Roentgenogram of the Metastasis from the Thyroid Adenoma, in the Left Humerus**

**Case Report**

A woman sixty-four years of age, who for the last twenty years had on the whole been in good health, suffered a spontaneous fracture of the left arm just above the elbow. X-ray examination showed grave changes in the bone structure of the humerus, spread...
over some 9 cm. of the distal end. This cast a bulky shadow of multicystic appearance, and 5 cm. of the shaft had disappeared (Fig. 1). As the blood calcium was increased to 15.45–17.05 mg. per cent, there was no doubt of the diagnosis—osteitis fibrosa Recklinghausen.

The patient suffered from vomiting which was difficult to control. She gradually became apathetic and anemic, and finally died marantico modo, seven weeks after the injury.

Autopsy: The body is well nourished. The spinal column shows a lordosis with the convexity to the left. In the left lobe of the thyroid gland is a partly calcified, encapsulated solitary adenoma, the size of a plum (Fig. 2). Four parathyroids of normal size were found in the usual places. In both lungs were numerous tumor metastases, the largest as big as a pea and with a reddish-gray, rather soft cut surface. There was a large tumor metastasis in the pleura between the 6th and 7th ribs.

Examination of the fatty thymus reveals in its lowest part, in front of the pericardium, a tumor as big as a pigeon’s egg, and of an appearance somewhat different from the metastases described above. This tumor is brown and soft and has a smooth glossy surface (Fig. 2).

On the lesser curvature of the stomach is a pedunculated polypus. All other organs are free from metastases and show no noteworthy changes.

The lower part of the left humerus is taken out with the surrounding muscles.

Microscopic Examination: The adenoma of the thyroid is strictly circumscribed by a fibrous, partly calcified capsule. There is no infiltrative growth in the parenchyma of
FIG. 3. **Area of Thyroid Adenoma Composed of Solid Bands and Fields of Tumor Cells Separated by Capillaries**

In the left upper corner is a small follicle distended by thin colloid.

FIG. 4. **The Parathyroid Struma**

On the left is the microscopic appearance of the main bulk of the tumor. On the right is an adenomatous area with cells of a type similar to the cells of Welch.
the gland. The structure of the tumor varies. In some places the alveoli are small, and
more or less lack colloid. In some areas this small-folliculated parenchyma is of a still
less differentiated shape, the tumor cells forming bands and fields separated by capillaries
(Fig. 3). Mitoses are not found, nor any polymorphism of the cells or hyperchromasia
of the nuclei. There are no changes in the surrounding thyroid.

The parathyroid struma in front of the heart has an appearance which I have found
in several other cases of von Recklinghausen's osteitis fibrosa (1). The tissue consists
of large epithelial cells rich in protoplasm and arranged in solid follicles surrounded by
capillaries (Fig. 4). They contain practically no fat. Some areas are adenomatous,
and in these the cells are large, with multiple nuclei of varying size (Fig. 4). The
protoplasm in these cells is eosinophil, just like the cells of Welch.

The parathyroids show no pathological changes.

Microscopical examination of the adrenals reveals a small encapsulated adenoma of

![Fig. 5. Tumor Tissue of the Metastasis in the Left Humerus](image)

well-known structure. All the chromaffin cells of the medulla stand out sharply blue in
preparations stained with Ehrlich's hematoxylin and eosin.

In the kidneys are vast deposits of calcium, both in the parenchyma cells and in the
interstitial spaces.

The metastases in the lungs show the picture of a parenchymatous thyroid adenoma
with the tumor cells arranged in bands or small follicles, mostly empty.

The tumor of the left humerus is uniform and the structure resembles a struma
colloides, with large follicles filled with colloid (Fig. 5).

Slides from different parts of the skull, the spinal column and the tubular bones show
everywhere the osteolytic process characteristic of osteitis fibrosa Recklinghausen. The
Haversian canals are much widened and filled with connective tissue and giant cells of
the osteoclast type, arranged along the walls in small cavities in the bone (Fig. 6).
There is no evidence of new bone formation, cysts and giant-cell tumors being absent,
and the contents of the major marrow cavities of the bones unchanged.
DISCUSSION

The case just described is interesting from many points of view. The presence of adenomata in three internal secretory glands points to an embryonal disturbance as the basis of them all. This view is supported by the fact that the para-struma has originated in a heterotopic fifth parathyroid gland. The position of this, right down on the pericardium, is remarkable but easily explained, as aberrant parathyroid tissue may be found anywhere in the thymus.

Metastasizing adenoma in the thyroid is a rare affection, and deserves publication as further proof of the occurrence of such growths.

The para-struma has given rise to an osteitis fibrosa Recklinghausen of a type which the author has previously described, but which Jaffe (2) only with hesitation accepts as that disease. He believes that new bone formation, cysts and giant-cell tumors, ought to be present before this diagnosis can be established. This opinion is difficult to share, as such changes in the bone are but secondary. The essence of von Recklinghausen's disease of bone appears to be a general osteolysis around the bone corpuscles. This gives rise to new bone canals, and a widening of the old. By amitotic division the liberated bone corpuscles are changed into giant cells, and the canals are filled with connective tissue instead of with bone marrow. That the giant cells have their origin in dissolved trabeculae is proved by the fact, among other things, that they are frequently arranged in groups possessing the configuration of the trabeculae; although all the bony substance is

Fig. 6. Enlarged Haversian Canal in the Shaft of One of the Tubular Bones Filled with Loose Connective Tissue, and Giant Cells Along the Wall
dissolved (Fig. 7). This is what happens first, and the process may be met with anywhere in the osseous system. Later, the tendency towards proliferation of the de-differentiated bone corpuscles will also find expression in mitotic division, and voluminous growths will occur here and there, distending the bone substance and forming the well known multiple giant-cell tumors.

Symptoms of hyperparathyroidism from other than osseous organs will, of course, also appear in the course of the disease.

The para-struma deserves a brief mention. Adenomata are occasionally found in the parathyroids post mortem. These differ from the usual parathyroid tissue in two ways: there is practically no fat in the actual tumor cells, and there are no fatty cells in the interstitial tissue. Staining will therefore make the adenomata show up very distinctly against their surroundings. In other words, the tumor tissue is structurally the same as that of the normal parathyroid gland of a new-born infant. Some thyroid adenomata are, as we know, quite analogous to this.

The parathyroid tumors in osteitis fibrosa Recklinghausen, at least in the 6 cases I have had the opportunity of examining, appear to be of a like nature. In all these cases, however, the tumor or tumors were so large that the old parathyroid tissue was so completely displaced that it could not be discovered. In the case under consideration the parathyroids which were in their right places proved to have a

**Fig. 7. Totally Dissolved Bone Trabecula in a Giant-Cell Tumor from a Case of Osteitis Fibrosa Generalisata Recklinghausen**

Only débris of collagenous substance and giant cells is left, indicating the outline of the original trabecula.
normal amount of fat in the parenchyma cells, as well as fatty cells in the interstices.

The fact that the cells in the regressively changed portions of the para-struma had become like the cells of Welch seems to me to support the view which I have previously advanced (3), viz. that the cells in the parathyroids are of only one kind, and that the cells of Welch are degenerative formations in the aging gland.

References

2. JAFFE, H. L.: Hyperparathyroidism (Recklinghausen's Disease of Bone), Arch. Path. 16: 63, 1933.