A CASE OF MEDIASTINAL DERMOID CYST CONTAINING PANCREATIC TISSUE, SIMULATING INTRATHORACIC GOITER

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The mere addition of another case of mediastinal dermoid cyst to those already recorded in the literature would hardly seem to be justified. Our purpose in reporting the following case is to emphasize the possibility of the confusion of mediastinal dermoid cyst with intrathroacic goiter, and more particularly to record the occurrence, unique in this case so far as we have been able to determine, of pancreatic glandular and islet tissue in the cyst wall.

CASE REPORT

A woman, 35 years of age, entered the Cleveland Clinic Hospital on October 22, 1929, complaining of marked dyspnea on exertion and a tumor in the suprasternal notch.

The tumor, which was first noticed two years previously as a slight, tender swelling, had persisted for two days and then disappeared. At that time the radiographic examination of the teeth gave negative findings; the tonsils were removed on account of chronic infection. Since its first appearance the swelling had recurred intermittently in the suprasternal notch and above the sternal end of the right clavicle, remaining for a few days and then subsiding.

At the time of admission the patient complained of shortness of breath, difficulty in swallowing, a slight non-productive cough and hoarseness of the voice. There had been no increased nervousness, no loss of appetite or weight and no gastrointestinal or genitourinary symptoms. The patient was well developed and was not acutely ill. The past and family history revealed nothing of importance.

Physical examination showed a tumor mass about the size of a hen's egg protruding into the suprasternal notch. During the act of swallowing this mass followed the movement of the trachea. There was no thrill, bruit or pulsation. An area of upper mediastinal dullness extended 4 cm. to the right and 5 cm. to the left of the midsternal line. Examination of the heart, lungs, abdomen and extremities revealed no significant abnormality.

Laboratory Findings. A roentgenogram of the chest (Fig. 1) revealed a large tumor mass in the upper mediastinum which was interpreted as a substernal goiter extending down to the third interspace anteriorly. The red blood cells numbered 4,000,000; white blood cells, 9,200; hemoglobin, 75 per cent. The basal metabolic rate was minus 7 per cent. The blood sugar before operation was 108 mg. per hundred cubic centimeters, one and one-half hours postprandial. The blood sugar after operation was 75 mg. per hundred cubic centimeters fasting. The Wassermann and Kahn reactions were negative. The urine was essentially negative on four examinations.

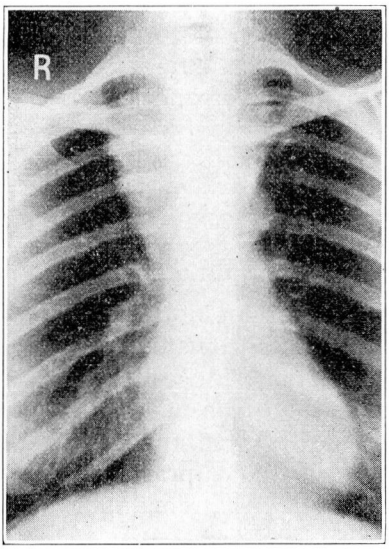

Fig. 1. Roentgenogram of the chest showing the mediastinal tumor.

A clinical diagnosis of substernal goiter was made and the patient was operated upon by Dr. G. W. Crile. Under nitrous oxide-oxygen anesthesia and novocain infiltration, a low collar incision was made as for thyroidectomy. The pretracheal fascia and muscles were separated in the median line exposing an apparently normal thyroid gland. Below the isthmus of the thyroid and bulging into the suprasternal notch, could be seen a tumor mass which extended into the mediastinum and apparently had no connection with the thyroid. Early in the process of freeing the tumor, a cyst was ruptured and approximately 150 to 200 cc. of thin, brownish fluid escaped. The character of the fluid suggested that the lesion was a dermoid cyst. The partially collapsed cyst-

wall was removed with considerable difficulty, on account of its intimate relationship to surrounding mediastinal structures. After the removal of the cyst, a large cavity, measuring approximately 12.5 cm. vertically and 7.5 cm. transversely, remained, bounded below by the arch of the aorta, anteriorly by the sternum, on the right by the innominate artery and vena cava, and on the left by the carotid and subclavian vessels. Posteriorly, the cyst-wall was in relation to the trachea and bronchi. After the cyst had been removed, the cavity was rinsed with saline and lightly packed with vaselinized scriflavine gauze. The operative wound was left open in order to allow free drainage. The patient was in excellent condition at the completion of the operation.

The postoperative reaction was very slight, and on the second postoperative day the cervical wound was closed around a small rubber catheter. The wound healed satisfactorily and the patient was discharged on the eighteenth postoperative day. At the time of discharge, abductor paralysis of the right vocal cord was present.

One year following the operation, a letter from the patient stated that she was in excellent health, had no evidence of recurrence and no hoarseness of the voice.

Macroscopically, the tissue received in the laboratory consisted of a ruptured, collapsed cyst-wall, weighing 45 grams and measuring 12 x 6 x 1.5 cm. It was irregularly pear-shaped. The cyst-wall was fibrous and varied from 3 mm. to 15 mm. in thickness. The inner surface was rough, somewhat trabeculated and had numerous, small masses of yellowish-brown granular material adherent to the lining. There were numerous small out-pouchings in the wall and several small, round, pouched-out areas, suggestive of ulceration, but without plastic exudate. In several areas, irregular masses of yellowish tissue were present in the wall, limited chiefly to the inner half. No hairs and no cartilage or bone could be recognized grossly. The outer surface of the cyst consisted of irregular, shaggy, fibrofatty tissue on the posterior aspect and a comparatively smooth, shiny, membranous layer on the anterior aspect.

Microscopically, the inner lining of the cyst consisted largely of fibrous tissue devoid of epithelium, but in some areas, particularly the thicker portions of the cyst-wall, there were fragments of stratified squamous epithelium, without keratohyaline material or hair shafts. Beneath this, there were a few lobules of sebaceous glands. No coil glands were recognized. In other areas, the inner lining consisted of simple, columnar mucus-secreting epithelium, and beneath this there was a large mass of non-encapsulated, lobu-

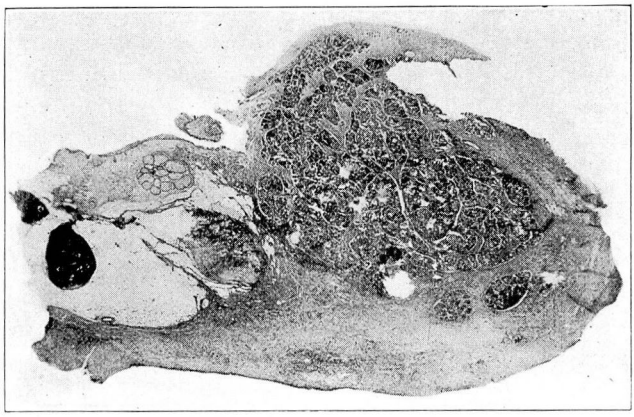

Fig. 2. Section of the cyst wall showing the mass of lobulated pancreatic tissue; x 6.

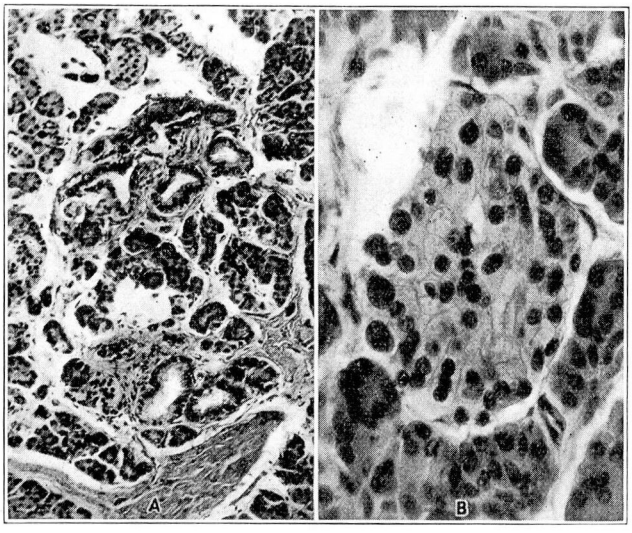

Fig. 3. (*a*) Pancreatic glandular tissue and ducts; x 150. (*b*) Pancreatic islet and glandular tissue; x 500.

lated, glandular tissue and ducts lined with cuboidal epithelium (Fig. 2). This tissue had the morphological characteristics of pancreas (Fig. 3). Scattered throughout this area were numerous small and large, solid masses of pale cells, morphologically similar to islet tissue. In other areas, the inner lining of the cyst was made up of stratified, columnar, ciliated epithelium, beneath which there were a few small, mucous glands in the stroma, suggestive of

pharyngeal mucosal structures. In this general neighborhood, there were several small islands of hyaline cartilage and fatty tissue in the cyst-wall, but no bone or dental structures were observed.

The middle zone of the cyst-wall consisted of collagenous, fibrous tissue showing extensive hyalinization and areas of mucoid degeneration. In numerous areas, there were irregular small and large collections of phagocytic cells containing yellowish-brown blood pigment and considerable lipoid material. A few scattered islands of perivascular lymphocytic infiltration were also found.

In the outer layers of the cyst-wall, several centimeters from the site of the pancreatic tissue, there were small masses of lobulated,

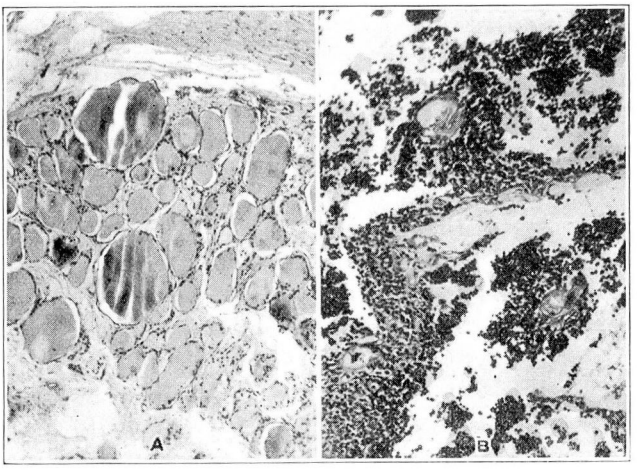

Fig. 4. From the outer layer of the cyst wall. (*a*) Thyroid tissue; x 100. (*b*) Thymic tissue; x 150.

colloid thyroid tissue (Fig 4), with vesicles lined by flattened epithelium, and considerable thymic tissue containing Hassall's corpuscles (Fig. 4). Some of the latter tissue extended well into the fibrous wall of the cyst and appeared to be somewhat compressed and atrophic. In such areas, Hassall's corpuscles were not present.

The most frequent location of intrathoracic dermoid cysts is the upper mediastinum. They are usually situated medially, but may extend laterally so that the major portion of the mass appears to the right or left of the midline. Generally, they are entirely within the thorax, but in about ten per cent of cases,[1] a swelling is apparent in the neck. The tumors vary greatly in size and consist of simple epidermoidal components or complex teratomatous growths. The symptoms are chiefly those due to mechanical ob-

struction, compression or irritation of surrounding structures, and are dependent largely upon the size of the mass. The principal complication is infection, which may terminate in the formation of an abscess, with spontaneous rupture, mediastinitis, perforation of the trachea, emphysema, pneumonia or pulmonary abscess. Malignant neoplastic transformation occurs in about 15 per cent of cases.[2]

The literature concerning intrathoracic dermoid cysts has been reviewed by Kerr and Warfield,[1] Williams,[2] Hertzler,[3] Harris,[4] Hale,[5] and others. To these articles, the reader is referred for a complete bibliography.

Discussion

The case reported illustrates how easily a mediastinal dermoid cyst may be mistaken for an intrathoracic goiter. The comparatively rare occurrence of the former as contrasted with the frequent occurrence of the latter lesion is probably one of the reasons why the differentiation is not made clinically more frequently. In cases of intrathoracic goiter, it is usually possible to detect a direct continuity between the thyroid itself and the mediastinal swelling. On the other hand, in ninety per cent of the cases reviewed in the literature, the mediastinal dermoid cyst is not in continuity with the thyroid. Therefore, demonstration of this continuity, or the absence of continuity, may prove to be a useful sign in making the correct diagnosis, although up to the present time we have not had occasion to confirm this suggestion.

Misinterpretation of thyroidal and dermoidal lesions, at operation and on gross examination of the removed tissue in the laboratory, may occur also in another direction. A broken down, necrotic, cystic adenoma of the thyroid, whether cervical, partially or completely intrathoracic, may be mistaken for a dermoid cyst on account of the nature of the contents and the character of the cyst-wall. Such tumors generally are a part of the thyroid, and microscopical examination after their removal suffices to make the distinction.

In the title the tumor is stated to be a dermoid cyst. Its very complexity, however, probably necessitates its being classified as a teratomatous cyst. Derivatives of three germ layers are represented, namely, stratified squamous epithelium and sebaceous glands; ciliated columnar epithelium and pharyngeal mucous glands; thyroid and thymus; pancreas; fat and cartilage.

The branchial apparatus would seem to be the most probable source for the particular combination of tissues present in the

cyst-wall and the tumor may be looked upon as an inclusion cyst. In this sense, it is not exactly comparable to those ovarian dermoids, in which a totipotential germ cell is capable of differentiation in all of the tissues of the body.

The thyroid tissue in the cyst-wall in our case is probably of lateral branchial rather than of median pharyngeal origin. Its intimate association with thymic tissue would seem to warrant this conclusion. The occurrence of tissue having morphological characteristics identical with those of the pancreas is of interest and has not been reported heretofore as occurring in a mediastinal dermoid, as far as we know. The presence of this tissue as a pharyngeal derivative is not so readily explained without invoking the theory of metaplasia. The salivary glands and the pancreas are physiologically related organs, but originate at widely separated points in the entodermal tract. If the primitive pharyngeal epithelium can differentiate into such diverse organs as the salivary glands, the thyroid and the thymus, it is not inconceivable that the pancreatic tissue in our case may likewise be of pharyngeal origin.

As was noted in the case report, no significant disturbance of sugar metabolism was detected.

Summary

A case of dermoid cyst of the anterior mediastinum extending into the neck and simulating a substernal goiter is presented. The tumor was removed through a low collar incision as in thyroidectomy. Recovery was complete and the patient reported that she was in excellent health, one year following the operation.

The presence of histologically normal pancreatic tissue in the cyst-wall makes this case unique.

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