Clinical Reports from Memorial Hospital, New York City

NEUROFIBROMA OF THE OVARY ASSOCIATED WITH RECKLINGHAUSEN'S DISEASE

By FRANK R. SMITH, M.D.

Solid fibromas of the ovary constitute a well defined pathologic entity with a definite clinical course. The purpose of this paper is to report a case of ovarian fibroma presenting certain clinical and pathologic features which appear to separate it from the usual ovarian fibroma.

The patient was fifty years of age, a virgin. Her past and family history offered nothing of interest save as detailed below. At the time
of the menopause, some twelve years before admission to the Memorial Hospital, multiple small tumors appeared in the skin of the entire body, especially over the trunk. The number of such lesions increased, although the individual tumors remained unchanged in size. The patient complained of itching of the skin, loss of weight, and dyspnea, and consulted a physician, who referred her to the Memorial Hospital, where she was first seen July 11, 1930.

Examination revealed a fairly well nourished woman. Her entire body was studded with numerous small nodules ranging from pin head size to 3.0 cm. in diameter. Blotches of brownish pigmentation were present over the chest and abdomen (Fig. 1). The abdomen was filled by a partially movable cystic mass, which rectal examination revealed as separate from a small senile uterus. Two small myomas were palpable in the uterus. Further examination showed nothing of interest, and a chest skiagraph was negative. The clinical diagnoses were (1) cutaneous neurofibromatosis (von Recklinghausen's disease) and (2) carcinoma of the ovary with possible lung metastases (on account of the dyspnea). A biopsy from one of the skin nodules was reported "neurofibroma" (Fig. 2).

On August 19 a celiotomy was performed. The right ovary was found practically free and filling most of the abdominal cavity. Solid masses were palpable within a cystic ovary, and the operator felt that he
was dealing with a papillary adenocarcinoma. The uterus with its small myomas, a calcified left ovary, the cervix, and the large tumor of the right ovary were removed en bloc. Figure 3 shows the tumor. It weighed 12 lbs. Examination showed it to be encapsulated. The surface was smooth. On section approximately two-thirds of the mass was made up of a smooth-walled cyst containing clear straw-colored fluid. One pole, or about one-third of the tumor, was solid, hard, yellowish, and grossly made up of interlacing fascicles. A trace of lutein pigment occurred on the cyst wall.

Histologically the tissue did not resemble the usual fibroma of the ovary. The interlacing whorls of spindle cells were very suggestive of neurofibroma or low-grade neurogenic sarcoma. This opinion was expressed by the pathologist, although, being unaware, at the time, of the coexistent cutaneous neurofibromatosis, he was unwilling to make a definite diagnosis of visceral von Recklinghausen's disease. When this history became known, it, together with the structure of the solid portion of the ovarian mass, appeared to justify the diagnosis of ovarian neurofibroma, probably arising in sympathetic nerves of the ovary.

The patient was discharged September 6, 1930, with her dyspnea relieved but, of course, with the skin lesions unchanged.

Visceral neurofibromatosis is, of course, well known. In certain instances of neurofibromatosis the cutaneous tumors are complicated with tumors of deeper nerve trunks and multiple or single tumors which project into the pleural or peritoneal cavities,
giving the superficial impression of being metastatic. The complicating feature of the case is the presence of lutein pigment in blotches along the wall of the cystic portion of the tumor. Experience with ovarian cystomas, however, has convinced us that this offers no bar to the neurogenic interpretation of the tumor, since in many cystic tumors of the ovary which cannot be considered to be of lutein cyst origin, pigment occurs over the cyst wall. This pigment is probably the result of the incorporation of incidental lutein tissue during the growth of the major cyst.

No effort has been made to review the literature on ovarian tumors to ascertain the frequency of probable neurogenic origin. Stewart, in a personal communication, informs us that in a recent examination of some 80 articles on neurofibromatosis he has not seen reference to ovarian neurofibroma, and the apparent rarity of the condition seems sufficient justification for this brief report.