

## CASE REPORT

# Neurofibroma of the Ovary

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We report an unusual example of ovarian neurofibroma in a woman with neurofibromatosis (von Recklinghausen's disease). The neoplasm caused ovarian enlargement which surgically simulated malignant neoplasm. Ovarian neurofibromas are extremely rare and may arise from small myelinated nerves in the ovarian hilum. © 1990 Academic Press, Inc.

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### INTRODUCTION

Neurofibroma involving the ovary is rare, having been reported only once before in the English literature [1]. We report a second case occurring in a woman with von Recklinghausen's disease.

### CASE REPORT

A 51-year-old woman with von Recklinghausen's disease sought medical attention because of spotty vaginal bleeding of 6 months' duration. A 2-cm vaginal apical ulcer was found and biopsy of this lesion showed adenocarcinoma. Total abdominal hysterectomy and right salpingo-oophorectomy had been performed 20 years previously for cervical squamous cell carcinoma *in situ*. In the light of this history and the histologic appearance of the vaginal neoplasm, the possibility of metastasis from a primary ovarian carcinoma was considered. At laparotomy, the left ovary was enlarged and firmly adherent to the adjacent peritoneum lateral to the rectum and was removed along with adjacent soft tissue. Small neurofibromas were noted along the pelvic sidewalls and in the periaortic areas.

The resected ovary was 3.5 cm in diameter and had a finely convoluted surface. With the exception of a simple cyst, the parenchyma was uniform. The hilum was expanded by soft, tan tissue which extended into the perihilum. Microscopically, the hilum contained well-circumscribed nodules of clustered fascicles of spindled

cells. These cells possessed elongated tapered nuclei and dense eosinophilic cytoplasm (Fig. 1). A myxoid matrix containing delicate collagenous fibrils separated the spindled cells. Mast cells were in evidence; mitotic figures were absent. Immunohistochemistry demonstrated that the nuclei and cytoplasm of the spindled cells contained S-100 protein (Fig. 2).

### DISCUSSION

Neurogenous neoplasms of the female reproductive organs, particularly of the ovary, are extremely rare [2]. To our knowledge, only one other example of an ovarian neurofibroma has been reported in the English literature [1]. In that report, Smith described a 5.4-kg well-encapsulated ovarian tumor. The diagnosis of neurofibroma was based upon the histologic finding of interlacing whorls of spindled cells and the patient's history of von Recklinghausen's disease.

While the histologic features present in our case support the diagnosis of neurofibroma, the presence of S-100 protein corroborates the diagnosis. S-100 protein is widely distributed in the central and peripheral nervous systems, and is present in virtually all neurofibromas [3]. In view of the origin of neurofibromas from peripheral myelinated nerves [3] and the widespread distribution of nerves to the ovarian stroma [4], it is surprising that ovarian neurofibromas are not more frequently found in women with von Recklinghausen's disease. It is conceivable in our case that the ovarian neurofibroma had its origin from a neurofibroma of the ipsilateral pelvic sidewall. However, the ovary was excised because its enlargement simulated malignant neoplasm. This case is a reminder that ovarian involvement by neurofibroma should be considered in the differential diagnosis of ovarian enlargement in women with von Recklinghausen's disease.



**FIG. 1.** Small clusters and fascicles of spindled cells interspersed by a myxoid matrix expand the hilum. Normal ovarian stroma is on the upper right. Hematoxylin and eosin,  $\times 125$ .

#### REFERENCES

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**FIG. 2.** Immunoperoxidase staining demonstrates that nuclei and cytoplasm of cells in the hilum contain S-100 protein. Note immunostaining of small nerves on the right. Immunoperoxidase,  $\times 250$ .