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Case Report

Central Neurocytoma Arising Within a Mature Cystic Teratoma of the Ovary

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Summary: Tumors of central nervous system elements are uncommon in cystic teratomas and tend to be derived from glial or primitive neuroectodermal cells. We describe the case of a 23-year-old woman with a central neurocytoma arising in an otherwise mature cystic teratoma of the ovary. Histologically, the neurocytoma was composed of collections of oligodendroglioma like cells in a fibrillary matrix. Cytologically the tumor was identical in appearance to central neurocytomas occurring within the adult brain. It expressed synaptophysin and neuron-specific enolase, confirming its neuronal lineage, but not glial fibrillary acidic protein. The site of the tumor recapitulated the typical location of neurocytomas adjacent to the lateral ventricle in that it abutted onto an ependyma-lined cyst within the teratoma. The patient remains well and free of tumor 1 year after cystectomy, in keeping with favorable follow-up data for surgically excised central neurocytomas within the brain. **Key Words:** Central neurocytoma—Cystic teratoma.

A wide variety of tumors, either benign or, more usually, malignant, may arise within cystic teratomas of the ovary. The most common of these is squamous cell carcinoma (1). Tumors of central nervous origin are uncommon and have included neuroblastoma, glioblastoma, and primitive neuroectodermal tumours (2–6). Ovarian ependymomas (6,7) have been described but they do not appear to arise within teratomas. We report the histological and immunohistochemical findings in a central neurocytoma arising within an otherwise mature cystic teratoma of the ovary.

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CASE REPORT

A 23-year-old nulliparous woman presented with acute onset of severe, spasmodic pain in the right side of the abdomen. On examination, a large, tender pelvic swelling was palpable 4 cm above the pubic symphysis. An ultrasound scan showed bilateral multiloculated ovarian masses; a torted ovarian cyst was diagnosed. Laparotomy confirmed torsion of the right ovary, which contained a cyst ~10 cm in diameter. A left ovarian cyst 9 cm in diameter was also present. The right ovary seemed to be viable after relief of the torsion. In view of the patient's age and the absence of any clinically malignant features, bilateral ovarian cystectomies were performed. The patient has had an uneventful follow-up of 1 year with no clinical or sonographic evidence of recurrent tumor.

The right ovarian specimen measured 10 × 7 × 3.5 cm, weighed 95 g, and contained two cysts 6 cm

and 4 cm in diameter. Both were filled with clear, straw-coloured fluid and had a smooth inner lining. The left ovarian cyst measured 8 × 9 × 5 cm, weighed 90 g, and contained hair and the typical sebaceous contents of a cystic teratoma, a small Rokitansky tubercle with bone and a well-formed tooth projected into the lumen. The right ovarian specimen contained a benign, simple serous cyst and a follicular cyst, whereas that from the left ovary contained mature adult tissues derived from all three germ layers. Ectodermal elements were represented by skin and adnexal structures. Respiratory epithelium and underlying mucus-secreting glands constituted the endodermal component, and the mesodermal tissues consisted of smooth muscle, adipose tissue, lamellar bone, cartilage, and a tooth.

A moderate amount of mature central nervous tissue elements were also identified in the wall of the teratoma. These included an ependyma-lined cyst with choroid plexus projecting into the lumen and several nodules of small, monomorphic tumor cells lying just beneath the ependyma (Figs. 1 and 2). The cells were embedded in a delicate fibrillary matrix and had fairly uniform, round nuclei with dispersed chromatin and perinuclear cytoplasmic clearing that imparted an oligodendroglioma-like appearance (Fig. 3). No mitotic activity, vascular hyperplasia, or necrosis was identified.

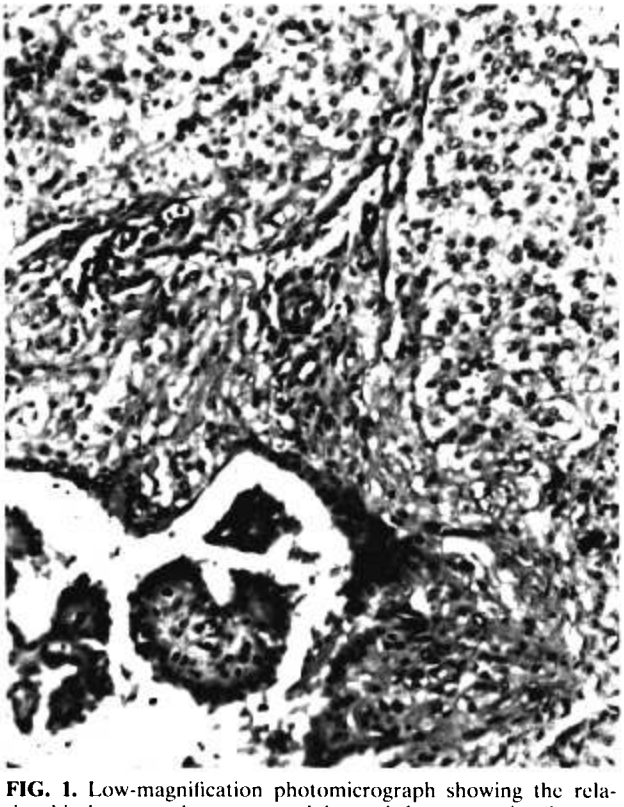

FIG. 1. Low-magnification photomicrograph showing the relationship between the tumor nodules and the cyst cavity (bottom left).

FIG. 2. At higher magnification, the cyst cavity is seen to be lined by ependymal cells.

Immunohistochemistry showed that the tumor cells expressed neuron-specific enolase (NSE) and synaptophysin (Fig. 4), indicating mature cells of neuronal lineage and excluding the possibility of oligodendroglioma. There was no immunoreactivity for glial fibrillary acidic protein (GFAP), confirming the lack of glial differentiation within the tumor nodules.

The immunohistochemical staining pattern, cytomorphology of the tumor cells, and anatomical relationship of the neoplastic cells to the ependyma-lined cavity were typical of a central neurocytoma. No undifferentiated or immature neuroectodermal elements were seen.

DISCUSSION

To our knowledge, there are no reports in the literature of a central neurocytoma arising in the ovary, either de novo or within a teratoma. Central neurocytomas are mature neuronal tumors that tend to occur adjacent to the lateral ventricles in young

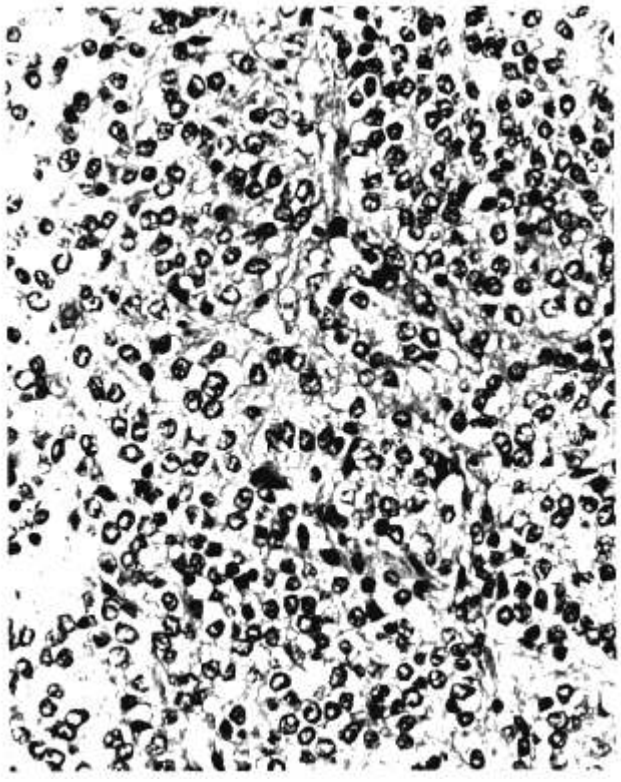

FIG. 3. The central neurocytoma consists of uniform cells with round nuclei and perinuclear cytoplasmic clearing, simulating an oligodendroglioma. Note the delicate fibrillary matrix separating some of the tumor cells.

FIG. 4. The tumor cells show cytoplasmic synaptophysin immunoreactivity. Note the lack of synaptophysin expression in the blood vessel toward the top right corner of the figure.

adults and to produce signs and symptoms of raised intracranial pressure as a result of obstructive hydrocephalus (8,9). Histologically, these tumors resemble oligodendrogliomas but have a fibrillary matrix and a rich, neuroendocrine-like vasculature. Typically, immunohistochemistry shows synaptophysin expression (9,10) but no GFAP immunoreactivity. NSE expression is also described (8,9). Synaptophysin is a glycoprotein of M_T38000 and spans the membrane of presynaptic clear vesicles. Its expression by the tumor cells indicates a differentiated, mature neuronal neoplasm. Electron microscopy has confirmed the neuronal origin of these tumors by showing the presence of dense core granules, clear vesicles, and synaptic membrane differentiation. Central neurocytomas usually grow relatively slowly, are well circumscribed, and may be successfully treated by surgical resection.

The location of these tumors adjacent to the lateral ventricle suggests that they originate from remnants of the subependymal germinal matrix, which retain proliferative potential. It is of interest that the central neurocytoma in this case occupied the same relative position in relation to the "ventricle" as it would have done in the brain and presumably arose from immature neural elements at this site within the teratoma.

In view of the good prognosis associated with excision of these tumors in their usual location and that they are terminally differentiated, mature central nervous neoplasms, we anticipate that excision of the tumor in the present case has been curative.

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