



Case Report

Primary ependymoma of round ligament of ovary masquerading as ovarian neoplasm: A rare case report with review of literature

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ABSTRACT

Ependymomas predominantly develop from neuroectodermal organs. Extra-neural ependymomas are uncommon tumors that occur in the sacrococcygeal region, pelvis, and meso-ovarium. Ependymomas of the round ligament/broad ligament are extremely rare, and only a few cases have been reported in the literature so far. Here, we report a rare case of primary uterine round ligament ependymoma in a 28-year woman presenting as a pelvic mass which clinically and radiologically mimicked a primary ovarian neoplasm. On histopathology and immunohistochemistry, the diagnosis of this typical primary extra-CNS ependymoma was supported by the presence of ependymal rosettes, perivascular pseudorosettes, and Glial fibrillary acidic protein immunoreactivity, along with Estrogen receptor and Progesterone receptor expression.

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INTRODUCTION

Extraneural ependymomas are uncommon tumors that occur in the sacrococcygeal region, pelvis, and meso-ovarium.¹ Ependymomas of the round ligament/ broad ligament are extremely rare; only a few cases have been reported in the literature so far, as per the PubMed database.^{2,3} Kleinman et al. provided the first complete description of the pelvic ependymoma with their report on three cases of ovarian ependymomas, and since then, five additional cases have been published with origins including the broad ligament, mesovarium, and omentum.⁴⁻⁸

Primary uterine broad ligament ependymoma is an extremely rare extra-CNS neoplasm with only seven previously reported cases in the literature worldwide. We are reporting this case because of its rarity.

CASE REPORT

A 28-year woman presented with abdominal distension and pelvic discomfort to the gynecology outpatient department of Gujarat Cancer & Research Institute. On imaging, the patient revealed a large pelvic solid cystic mass involving the right adnexa, and the right ovary was not seen separately from the lesion. Her routine serum tumor markers were only slightly elevated, and LDH levels were normal. The patient underwent primary staging laparotomy, and the specimen was sent to the oncopathology department of the institute for frozen section. On gross examination, the mass measured 28x20x16 cm, and the outer capsule was intact. No surface deposits were seen. The right ovary and fallopian tube were adherent to the mass and were grossly unremarkable. On cutting open, the mass was solid and cystic with fleshy and cystic areas filled with clear fluid (fig. 1). A squash preparation was done, and bits were taken. The squash preparation revealed round to oval cells with fibrillary processes and bland vesicular nuclei with occasional mitotic

figures (fig. 2a, b). Frozen section showed monotonous cells arranged in pseudo-rosettes, and a frozen diagnosis of embryonal-type neuroectodermal tumour with a possibility of ependymoma was rendered, with final diagnosis deferred to paraffin section and immunohistochemistry.



Figure 1: Gross examination of mass showing solid, fleshy and cystic cut surface

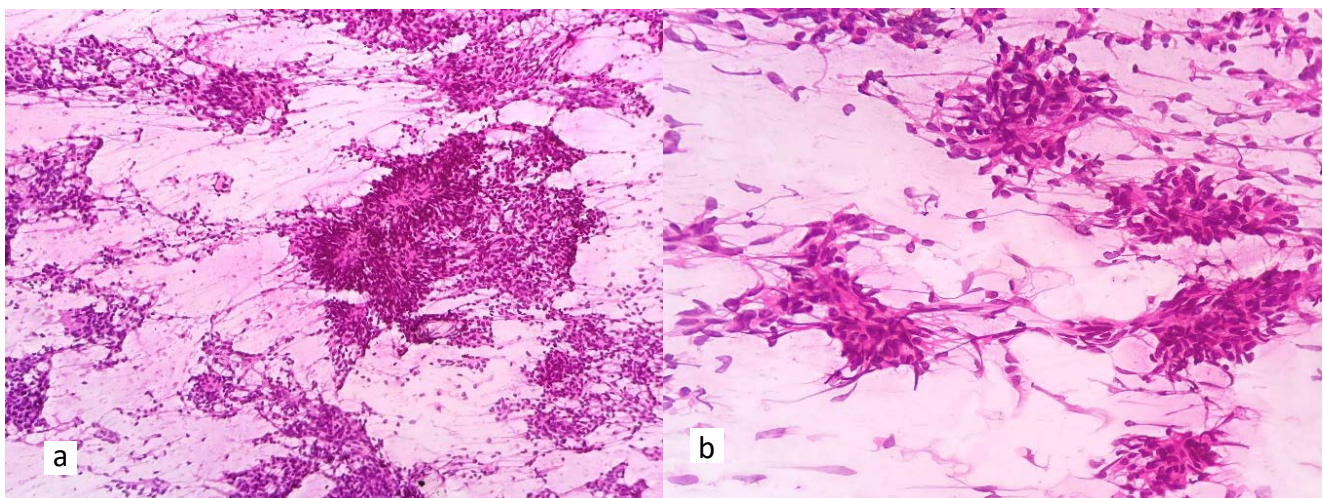


Figure 2a: Squash preparation showing cells forming rosettes having round to oval nuclei and moderate eosinophilic fibrillary cytoplasm (HE stain, X100) Figure 2b: Photomicrograph showing higher power view with bland nuclear features (HE stain, X200)

Microscopic examination of formalin-fixed & paraffin embedded sections revealed a cellular tumor composed of true ependymal and perivascular pseudorosettes. The neoplastic cells were cuboidal to columnar with round

vesicular nuclei and a moderate amount of fibrillary eosinophilic cytoplasm (fig. 3a, b). Occasional mitotic figures were noted. Metastatic deposits were seen in the pouch of Douglas and paraaortic lymph nodes (fig. 3c, d).

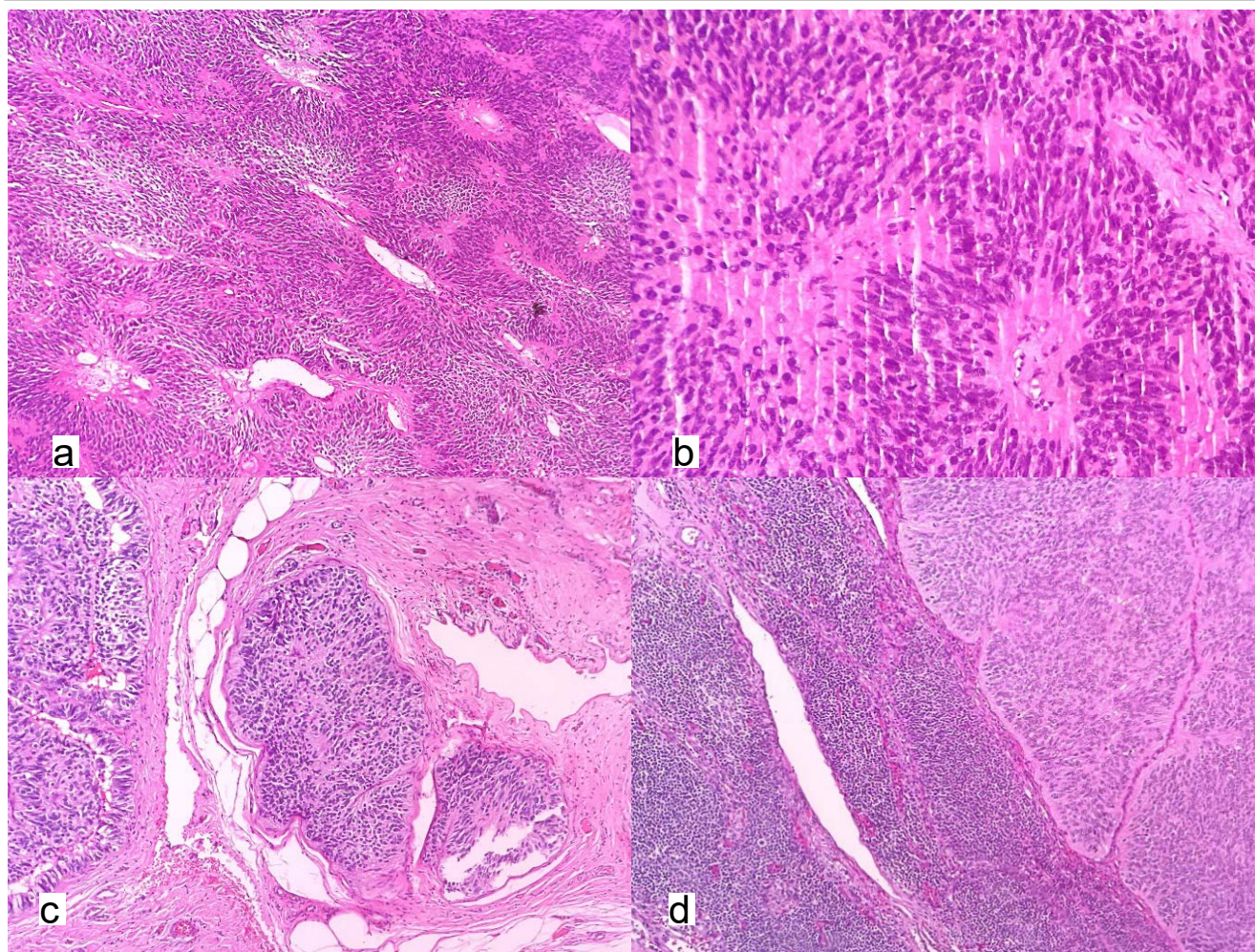
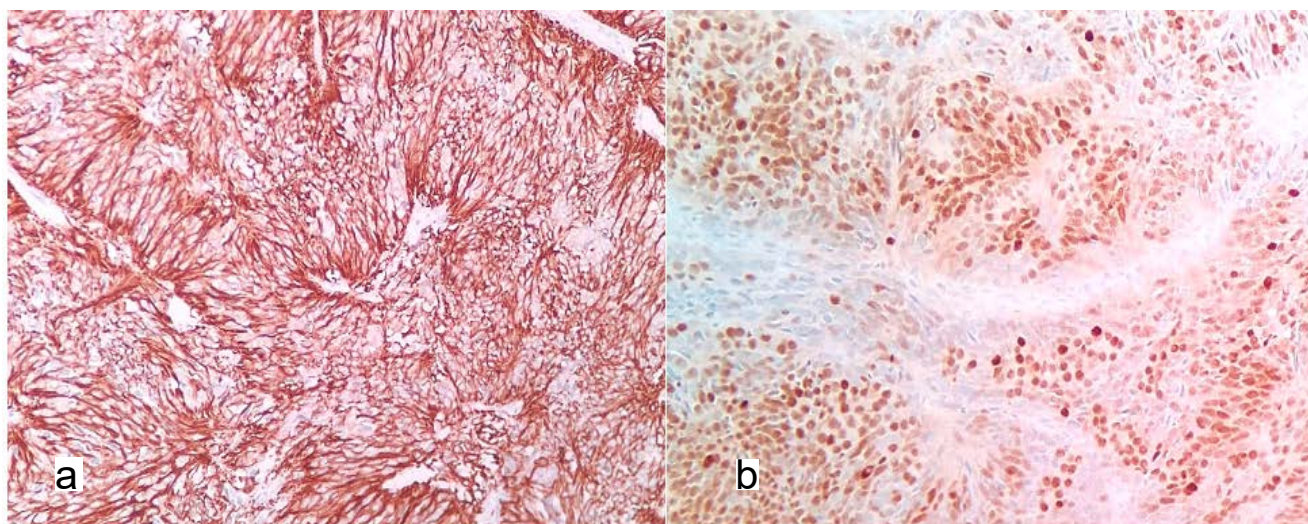


Figure 3a, b: Characteristic perivascular pseudo-rosettes of ependymoma (HE stain, X100, X400) Figure 3c: Metastatic deposit in pouch of Douglas (HE stain, X200) Figure 3d: Para-aortic lymph node deposits. (HE stain, X 200)

On immunohistochemistry, the neoplastic cells were immunoreactive for GFAP (diffuse and strong), ER, PR, EMA (dot positive), CD99 (membranous), and Oligodendrocyte transcription factor 2 (focal positive) and negative for SALL 4, pan-cytokeratin (AE1/AE3), and SOX10. MIB index was

approximately 18-20% (fig. 4a, b, c, d, e). Based on classical morphology and immunohistochemistry findings, a final histopathological diagnosis of Primary ependymoma NOS of the round ligament was given.



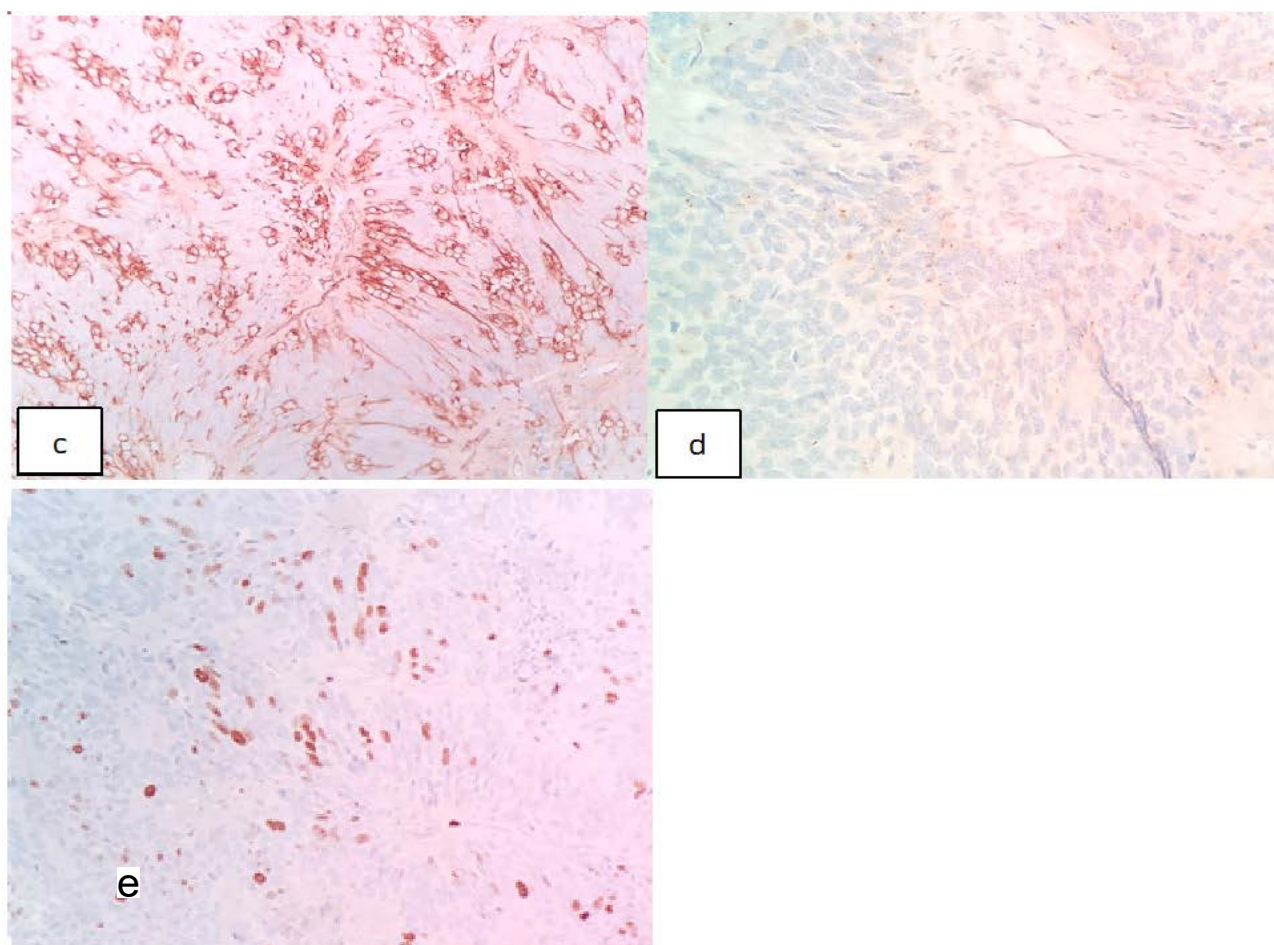


Figure 4a. Photomicrograph showing diffuse cytoplasmic GFAP positivity, 5b. ER nuclear immunoreactivity in tumor cells, 5c. CD99 positivity in tumor cells, 5d. Characteristic focal dot-like immunoreactivity for EMA, 5e. Photomicrograph showing MIB index of 18-20% at hot-spots.

DISCUSSION

Primary ependymoma of the round/broad ligament of the ovary is an extremely rare entity with only seven cases reported in the literature. The average age at presentation in the previous studies was 38 years, although it ranged from 13 to 61 years.² The exact histopathogenesis of the origin of these neoplasms is elusive, although many hypotheses have been proposed. One of the hypotheses considers neo-metaplasia of the peritoneal epithelium to neuroectoderm, which is not normally present at this site. In addition to ependymoma, under the hypothesis, other CNS tumors like astrocytoma and glioblastoma can also occur in fat and are considered examples of neo-metaplasias. Other theories state the presence of ectopic glial tissue or fetal neural tissue deposited at these sites because of prior aborted pregnancies; however, none of them is fully explanatory.⁷

Pelvic ependymomas predominantly occur in the reproductive age group and present as large, solid-cystic masses. Larger tumors are associated with the risk of metastasis and recurrence.

Mitotic activity is a poor indicator of prognosis.⁷

Due to the young age of the patient, germ cell tumors, especially yolk sac, was excluded by the absence of characteristic histomorphological features and normal AFP levels. Other differentials excluded were serous carcinoma, endometrioid carcinoma, granulosa cell tumor, and Sertoli-Leydig cell tumors by using appropriate immunohistochemistry panels.^{8,9}

Due to the extreme rarity of this entity, no consensus guidelines are available for therapeutic management. In all the previously reported cases, surgical excision was the first approach, followed by neoadjuvant therapy, as all cases revealed metastasis at the time of presentation.²⁻⁴

GnRH analogue therapy revealed good clinical response in some cases and may be considered in young females desiring fertility-sparing treatment.¹

Extra-axial ependymomas are immunoreactive for GFAP as well as ER and PR, thus may potentially respond to hormonal agents.⁹

Due to the paucity of data, the appropriate treatment guidelines, biological behaviour, and overall survival prediction are elusive.

CONCLUSIONS

Primary round/broad ligament ependymoma is an extremely rare entity with different therapeutic and prognostic implications. Nonspecific clinical and radiological findings make the preoperative diagnosis challenging. This case highlights the decisive role of histopathology in conjunction with immunohistochemistry in elucidating the accurate diagnosis. Complete surgical excision is the treatment of choice, along with long-term surveillance, given the uncertain biological behaviour and potential for late recurrences.

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Conflict of interest: None.

REFERENCES

1. Zhou F, Song J, Mikolaenko I, Rosenblum M, Shukla PS. Pelvic ependymoma with clinical response to GnRH analog therapy: a case report with an overview of primary extraneural ependymomas. *Int J Gynecol Pathol*. 2015;34(5):450-58. [Crossref](#)
2. Matsuyama A, Hisaoka M, Yamamoto I, Toyoshima S, Hashimoto H. Extraspinal ependymoma of the broad ligament. *Pathol Int*. 2010;60(3):241-44. [Crossref](#)
3. Bell DA, Woodruff JM, Scully RE. Ependymoma of the broad ligament: a report of two cases. *Am J SurgPathol*. 1984;8(3):203-9. [Crossref](#)
4. Kleinman GM, Young RH, Scully RE. Ependymoma of the ovary: report of three cases. *Hum Pathol*. 1984;15:632-38. [Crossref](#)
5. Grody WW, Nieberg RK, Bhuta S. Ependymoma-like tumor of the mesovarium. *Arch Pathol Lab Med*. 1985;109:291-93. PMID: 3838461
6. Dekmezian RM, Sneige N, Ordonez NG. Ovarian and omental ependymomas in peritoneal washings: cytologic and immunocytochemical features. *DiagnCytopathol*. 1986;2:62-8. [Crossref](#)
7. Duggan MA, Hugh J, Nation JG, Robertson DI, Stuart GC. Ependymoma of the uterosacral ligament. *Cancer*. 1989;64(12):2565-71. [Crossref](#)
8. Idowu MO, Rosenblum MK, Wei XJ, Edgar MA, Soslow RA. Ependymomas of the central nervous system and adult extra-axial ependymomas are morphologically and immunohistochemically distinct: a comparative study with assessment of ovarian carcinomas for expression of glial fibrillary acidic protein. *Am J SurgPathol*. 2008;32(5):710-18. [Crossref](#)
9. Carr KA, Roberts JA, Frank TS. Progesterone receptors in bilateral ovarian ependymoma presenting in pregnancy. *Hum Pathol*. 1992;23:962-65. [Crossref](#)