THE MEDICAL BULLETIN OF
SISLI ETFAL HOSPITAL

DOI: 10.14744/SEMB.2023.74340
Med Bull Sisli Etfal Hosp 2024;58(2):237–240

Case Report

Choroid Plexus Papilloma Tumor of the Ovary: A Case Report

Merve Aldikactioglu Talmac,[1] Yuksel Ulu,[2] Hilal Serap Arslan,[2] Ilkbal Temel Yuksel[1]

[1]Department of Gynecological Oncology, University of Health Sciences Türkiye, Basaksehir Cam and Sakura City Hospital, Istanbul, Türkiye
[2]Department of Pathology, University of Health Sciences Türkiye, Basaksehir Cam and Sakura City Hospital, Istanbul, Türkiye

Abstract

This is a case report of a 19-year-old nulligravid patient with a choroid plexus papilloma tumor in a mature cystic teratoma in the right adnexal area.

The patient, who had abdominal pain and dyspepsia, showed a 9 cm diameter mass with a solid/cystic component, initially interpreted as a dermoid cyst in the right adnexal region. Mature cystic teratoma is a benign germ cell tumor and is common in women during the reproductive period. However, choroid plexus papilloma is a rare brain tumor. The diagnosis of ovarian choroid plexus papilloma can be made with imaging tests such as magnetic resonance imaging or computed tomography, and treatment is usually by surgical removal.

Only four cases of ovarian teratoma with choroid plexus papilloma have been informed in the English literature, and this issue is the fifth.

Keywords: Choroid plexus papilloma, cystic teratoma, dermoid cyst, ovary, teratoma

Please cite this article as "Aldikactioglu Talmac M, Ulu Y, Arslan HS, Temel Yuksel I. Choroid Plexus Papilloma Tumor of the Ovary: A Case Report. Med Bull Sisli Etfal Hosp 2024;58(2):237–240".

Mature Cystic Teratoma (MCT) is a benign germ cell tumor originating from two or three germ sheets, usually unilaterally seen in women of childbearing age. Its incidence among all ovarian tumors is 20%.[1] Macroscopically, it is primarily cystic but can also be seen in solid form. Sebaceous material, hair, bone, and cartilage tissue can be observed in the cyst lumen. In solid areas, squamous epithelium of ectoderm origin, skin appendages, neuroectodermal tissue; adipose tissue, bone, cartilage, smooth muscle from mesoderm; gastrointestinal, respiratory bronchial epithelium, thyroid and salivary glands originating from endoderm can be seen.[2] Rarely, prostate, pituitary, adrenal, and parathyroid tissue can be observed. Malignant transformation is extremely rare (1.5-2%).[3] It is usually seen in the postmenopausal period. In the differential diagnosis, there are other ovarian masses, especially endometrioma.[4] Carcinoid tumors and adenocarcinoma follow the most usual squamous cell carcinoma.

Choroid plexus papilloma (CPP) is a rare brain tumor and usually arises from epithelial cells of the choroid plexus, where cerebrospinal fluid (CSF) is produced in the brain. The incidence of CPP among all brain tumors is 0.3-0.6%. While 10-20% of these cases are seen in the first year of life, 2-4% are seen under the age of 15.[5] These tumors occur primarily in the lateral ventricle in children. In adults, it appears in the 4th ventricle.[6] CPPs usually grow slowly and are rarely cancerous. However, large tumors can block CSF circulation and cause severe brain damage. While it is a tu-

Address for correspondence: Merve Aldikactioglu Talmac, MD. Department of Gynecological Oncology, University of Health Sciences Türkiye, Basaksehir Cam and Sakura City Hospital, Istanbul, Türkiye
Phone: +90 532 560 23 66 **E-mail:** drmrve@hotmail.com

Submitted Date: April 27, 2023 **Accepted Date:** August 18, 2023 **Available Online Date:** June 28, 2024

mor that commonly occurs in the brain, in rare cases, it can also occur in other regions, such as an ovarian teratoma. The diagnosis of ovarian CPP can be made with imaging tests such as computed tomography (CT) or magnetic resonance imaging (MRI). Treatment of CPPs in the ovary is usually done by surgical removal. Only four cases of ovarian teratoma have been described in the English literature. [7] Our case is the 5th CPP case in ovarian teratoma in the literature.

Case Report

A 19-year-old nulligravid patient complained of abdominal pain and dyspepsia for about four months. There was no specificity in his medical, family, or psycho-social history. In the abdominal ultrasonography and MRI, a 9 cm diameter mass through solid/cystic components in the right adnexal area, interpreted as a dermoid cyst, was observed in the foreground (Fig. 1). Tumor markers were detected as CEA: 0.975 ng/mL, CA 19-9: 15.9 U/mL, and CA 125: 33.7 U/mL. The patient underwent laparoscopic right ovarian cystectomy with the preliminary diagnosis of mature cystic teratoma.

Macroscopically, the tumor was 11x8x4 cm in size and contained hair, sebaceous material, and cartilage tissue. Microscopically, multilocular cyst, keratinized squamous epithelium, mucinous epithelium, skin appendages, hair shafts, adipose tissue, mature glial foci, chondroid and osteoid structures were observed. An insular focus was observed, consisting of monomorphic cells with a 0.9 cm diameter salt-and-pepper chromatin structure, round nuclei, eosinophilic cytoplasm, and monomorphic cells (Figs. 2, 3). Immunohistochemically, S100 (focal+), Synaptophysin (+), Chromogranin (+), CD56 (+) staining were observed. Ki 67 proliferation index was 6-7%, and mitosis was 3/2 mm^2. Necrosis was not observed. All findings were evaluated as 'Atypical Carcinoid Tumor (WHO Grade 2). Non-invasive papillary structures with multifocal mild-moderate nuclear atypia lined with focal stratified cuboidal epithelium, some with fibrovascular cores, were observed around this focus (Figs. 4, 5). Necrosis was not observed. Immunohistochemically, S100 (+), GFAP (focal+), CK7 (focal+), PR (focal, weak+), p16 (focal+), p53 (-), CK20 (-), AMACR (-), Calretinin (-), and WT1 (-) immunoreactivity were detected. Mitosis was 2-3 /10 BBA. Ki 67 proliferation index was 1-2%. The present findings were interpreted as 'Atypical CPP (WHO Grade 2)'.

Since the case was interpreted as 'Atypical Carcinoid Tumor, Atypical CPP Followed,' the patient was informed, and laparoscopic right salpingo-oophorectomy was performed as a complementary surgery. No abnormal cells were detected in the salpingo-oophorectomy material. Ga-68 Dot-

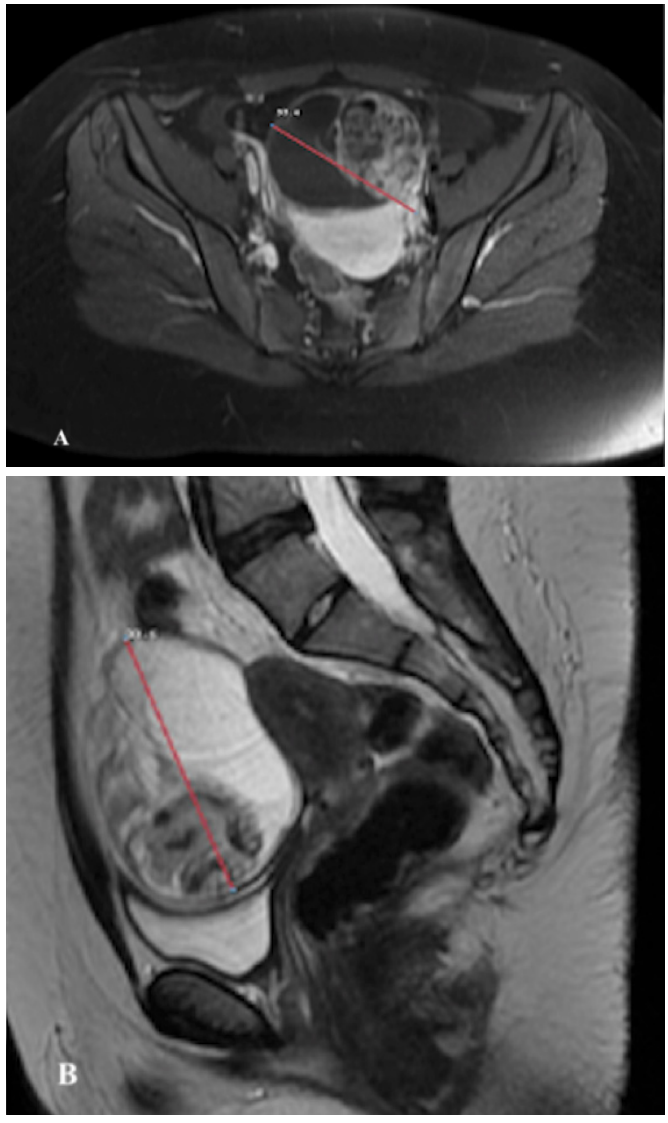

Figure 1. Preoperative MRI images of right side adnexal mass A. Transverse section B. Sagittal section.

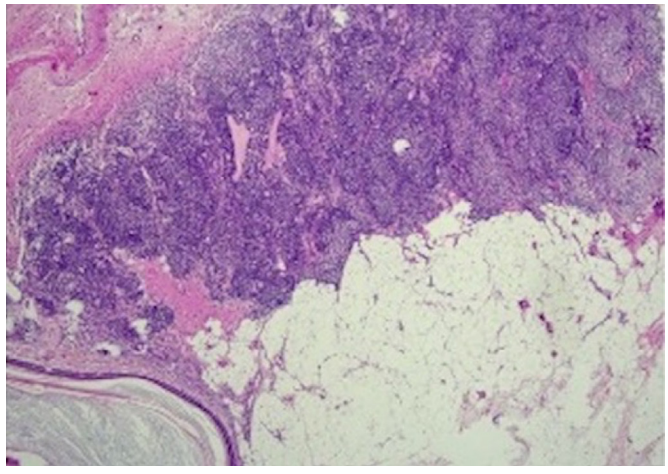

Figure 2. (H&E X4): Fat tissue, a focus of monomorphic cells next to a keratinous cyst.

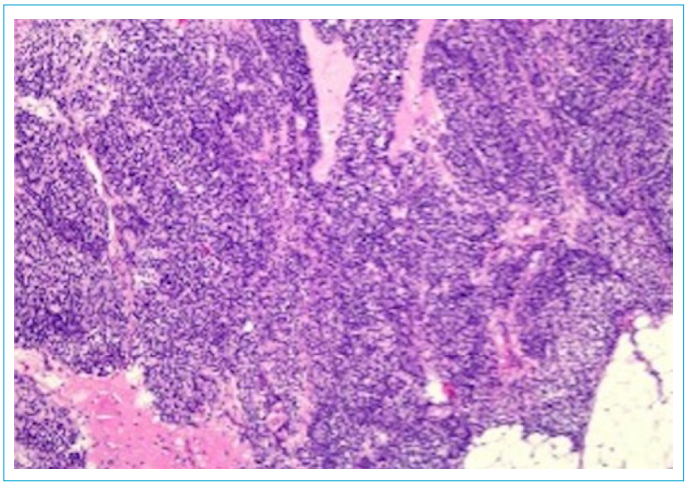

Figure 3. (H&E X10): Focal focus consisting of monomorphic cells with round nuclei, eosinophilic cytoplasm, with salt pepper chromatin structure.

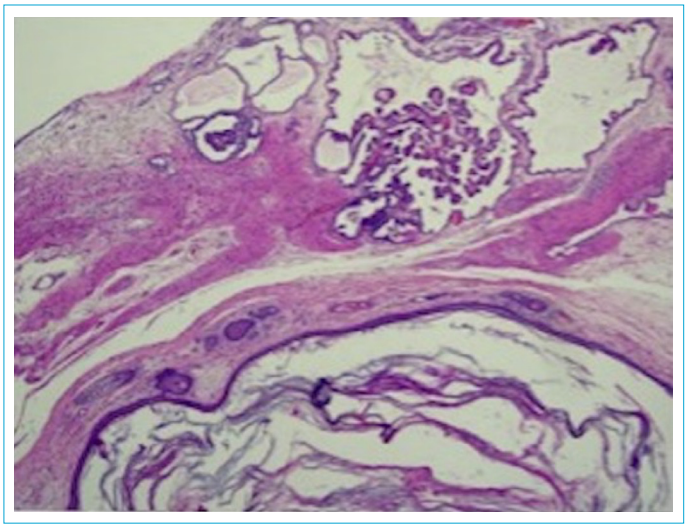

Figure 4. (H&E X4): Keratinous cyst, skin appendages, papillary structures.

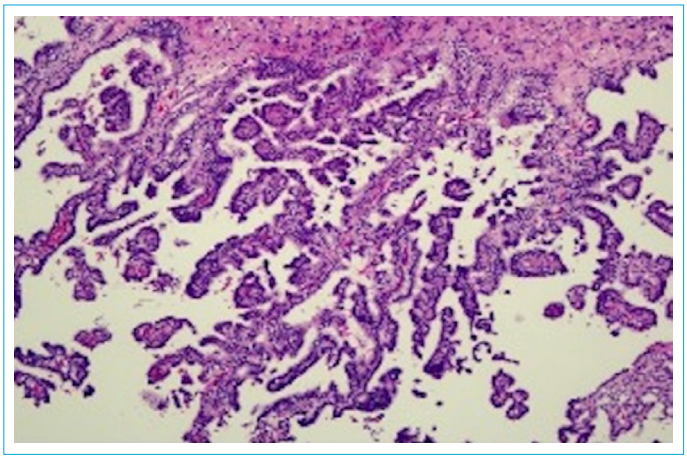

Figure 5. (H&E X10): Papillary structures lined with focal stratified cuboidal epithelium, some with fibrovascular cores.

atate Positron Emission Tomography showed no evidence of malignancy holes containing any somatostatin deposits at the sixth postoperative month. The patient continues to be followed at 6-month intervals.

Discussion

Ovarian primary carcinoid tumors are extremely rare and account for <5% of all carcinoid tumors. On the other hand, it is seen in less than 0.1% of all ovarian neoplasms.[8] It is separated into four subtypes: mucinous, insular, mixed, and trabecular type. CPP is a rare primary tumor of the central nervous system. Sixty neuroectodermal ovarian neoplasms were reported by Morovic and Damjanov in 2008.[9] These cases between 1983 and 2006 were classified as astrocytoma, ependymoma, primitive neuroepithelial tumor, or glioblastoma but were not included in the choroid plexus papillomas.

There are 4 cases published in the English literature that were diagnosed with CPP in the ovary. It was first reported by Von Gunten et al.[10] in Japan in 2006. However, no report was given regarding the size of the mass and its treatment method. The second case in the literature is the treatment of a 9 x 7 x 7 cm mass with cystectomy, similar to our case published by Quadri et al.[11] in 2011. In 2013, Dessauvagie et al.[12] published the third case of oophorectomy for an 8 x 3 cm mass. Kihara et al.[7] published the fourth case in the literature, choroid plexus tumor in the ovary, in 2015. The size of the mass, in this case, was quite large, 22.5×20.5 cm and the patient was treated with salpingo-oophorectomy. The subjects were between 14-32 years old, and the mean age was 23.75. The lesions had a maximum diameter that varied between 8 and 22.5 cm. Among the ovarian masses of which we know the dimensions, the mean tumor diameter was 11.5 cm. Our case is similar in terms of both age and tumor size. It also provides additional information regarding preoperative mass imaging and postoperative follow-up.

Usual WHO grade I CPPs are typically benign brain tumors considered curable through complete surgical removal. The successful removal of teratomas through oophorectomies and other reported cases suggests that these procedures can also be curative. It's important to note that ovarian papillary epithelial neoplasms, such as serous, clear cell, or endometroid tumors, can sometimes be confused with choroid plexus papillomata, particularly if psammomatous calcifications accompany them. Also, metastases from malignant mesothelioma, papillary thyroid carcinoma, lung adenocarcinoma, and urothelial carcinoma may be considered part of the differential diagnosis.[12]

Kleinman et al.[13] reported a neuroectodermal tumor series consisting of 25 cases. The prognosis of the CPP of the ovary is uncertain. According to this report, there was no choroid

plexus papilloma in them. The prognosis of people with 10 stage I tumors was better than those with stage II and III tumors. On the other hand, one patient with stage IA glioblastoma died of the disease within two years. Therefore, it remains unclear whether the clinical stage or tumor grade of an ovarian neuroectodermal tumor is relevant to prognosis. There is a need for statistical forecast analysis studies in more extensive case series on this subject. Our case was WHO Grade I in clinical stage IA. Therefore, we expected a good prognosis. However, due to the uncertainty of the forecast, long-term observation will be required so that we can continue the follow-up process at 6-month intervals.

Conclusion

This article provides valuable information about a rare ovarian teratoma case involving an atypical carcinoid tumor and choroid plexus papilloma. We emphasize the importance of a comprehensive pathological examination of ovarian teratomas and the need for appropriate management and follow-up in such rare cases. The presented case adds to the limited literature on ovarian teratomas, including choroid plexus papillomas as case 5, and provides insight into the management and prognosis of such claims. The presented case contributes to the limited literature as the fifth case with an atypical choroid plexus tumor in ovarian teratoma. It provides insight into the management and prognosis of such patients.

Disclosures

Acknowledgments: The authors thank the hospital pathology laboratory for their support. There is no funding from anywhere.

Informed Consent: The patient provided written informed consent to publish the case report and accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

Authorship Contributions: Concept – I.T.Y; Design – M.A.T; Supervision – I.T.Y; Fundings – Y.U.; Materials –H.S.A; Data collection &/or processing – Y.U.; Analysis and/or interpretation – M.A.T; Literature search – M.A.T; Writing – M.A.T.; Critical review – I.T.Y.

Use of AI for Writing Assistance: Grammarly premium program was used for language proofreading.

References

1. Kim MJ, Kim NY, Lee DY, Yoon BK, Choi D. Clinical characteristics of ovarian teratoma: age-focused retrospective analysis of 580 cases. Am J Obstet Gynecol 2011;205:32. [CrossRef]
2. Lee KH. 'Sack of marbles' in mature cystic ovarian teratoma. Abdom Radiol (NY) 2017;42:1616–7. [CrossRef]
3. Atwi D, Kamal M, Quinton M, Hassell LA. Malignant transformation of mature cystic teratoma of the ovary. J Obstet Gynaecol Res 2022;48:3068–76. [CrossRef]
4. Aşıcıoğlu O, Temizkan O, Arıcı B, Aşıcıoğlu BB. Clinical characteristics and outcomes of laparoscopic surgery in ovarian endometrioma cases treated at a gynecology clinic. Sisli Etfal Hastan Tip Bul 2018;52:1–5. [CrossRef]
5. Saygın İ, Çakır E, Ercin ME. Choroid plexus papilloma in extragonadal teratoma with predominantly neurogenic elements: a case report and review of the literature. Turk J Pediatr 2020;62:509–13.
6. Khade S, Shenoy A. Ectopic choroid plexus papilloma. Asian J Neurosurg 2018;13:191–4. [CrossRef]
7. Kihara A, Iihara K, Murata K, Kitagawa M, Horiuchi H. Choroid plexus papilloma arising in a mature cystic teratoma of the ovary: a short case report and literature review. Pathol Int 2015;65:563–5. [CrossRef]
8. Sharma A, Bhardwaj M, Ahuja A. Rare case of primary trabecular carcinoid tumor of the ovary with unusual presentation. Taiwan J Obstet Gynecol 2016;55:748–50. [CrossRef]
9. Morovic A, Damjanov I. Neuroectodermal ovarian tumors: a brief overview. Histol Histopathol 2008;23:765–71.
10. von Gunten M, Burger H, Vajtai I. Choroid plexus papilloma developing in a dermoid cyst of ovary. Histopathology 2006;49:204–5.
11. Quadri AM, Ganesan R, Hock YL, Karim SN, Hirschowitz L. Malignant transformation in mature cystic teratoma of the ovary: three cases mimicking primary ovarian epithelial tumors. Int J Surg Pathol 2011;19:718–23. [CrossRef]
12. Dessauvagie BF, Ruba S, Robbins PD. Choroid plexus papilloma arising in a mature cystic teratoma of a 32-year-old female. Pathology 2013;45:88–9. [CrossRef]
13. Kleinman GM, Young RH, Scully RE. Primary neuroectodermal tumors of the ovary. A report of 25 cases. Am J Surg Pathol 1993;17:764–78. [CrossRef]