Mature ovarian cystic teratoma containing well-differentiated cerebellar tissue

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Summary

A 38-year-old female patient experienced groin pain; ultrasound imaging revealed a dermoid cystic mass in the right ovary and a cystectomy was then performed. Unusually, a mature cerebellum is found in the cyst wall. The pathological diagnosis was 'mature cystic teratoma with well-differentiated cerebral and cerebellar tissue'. Glial tissue is a common neural component of teratomas, but a cerebellum is extremely rare in mature ovarian cystic teratomas. The authors report the case because of its rare component; they acknowledge that a cystic teratoma is the most common neoplasm of ovarian germ cells.

Key words: Dermoid mass; Mature cystic teratoma; Cystectomy; Reproductive age.

Introduction

Benign cystic teratoma is the most common tumor in females of reproductive age [1]. The teratoma has one or more embryonic germ layers [2]. Ectodermal tissues, especially skin and skin appendages, are commonly present [3]. Another common ectodermal element is brain [2], but mature cerebellar tissue is very rare. Only a few cases have been reported [4].

Case Report

A 38-year-old female patient, gravida 2, para 2, presented with groin pain. An ovarian cystic mass similar to a dermoid cyst was evident upon ultrasonic examination. A right ovarian cystectomy was performed. The uterus and left ovary were normal. A 65×55×35-mm cystic mass was detected. Oily, tan-colored, sebaceous, hairy material was present in the cyst cavity, and bone and cartilage were found in the cyst wall. A 17×12×9-mm-sized grey-white region (glial tissue) was noted; several samples were taken for microscopic examination. Histopathologically, tissues of all three germ layers were present. Most of the cyst was lined by keratinized squamous epithelium, as is the skin (Figure 1A). Desquamation and a foreign body giant cell reaction were evident in some areas. The cyst wall contained ectodermal derivatives, including skin appendages: hair follicles and sebaceous and sweat glands (Figure 1A), mature brain, and well-differentiated cerebellar tissue. Mesodermal tissues included fibrous and fatty tissues, smooth muscle, bone, and cartilage (Figure 1B). Additional endodermal tissue included gastrointestinal, bronchial epithelium, and glands. The cerebellar tissue was large (ten mm in diameter) and well-differentiated, thus exhibiting normal histological features of the mature cerebellum (Figure 1C). The tissue contained molecular-Purkinje cells and inner granular layers (Figure 1D). All tissues were mature and there were no immature neuroglial tissue despite extensive sampling. The final diagnosis (upon microscopic examination) was 'mature cystic teratoma with well-differentiated cerebral and cerebellar tissue'.

Discussion

Mature cystic teratomas are the most common ovarian tumors (20-40%) [5]. A teratoma is a germ cell tumor and develops most commonly during the reproductive years. However, unlike other germ cell tumors, the age distribution is wide; a tumor may develop at any age. Half of all cases occur in patients aged 20-40 years [4, 5]. Approximately half of all patients present with abdominal pain [5]. The present patient experienced groin pain only. Benign teratomas are nearly always cystic in nature and are frequently < ten cm in diameter; the average diameter is seven to eight cm [2, 5]. Abundant hair and sebaceous material are present in the cyst cavity [1, 5]. The gross findings can be classical (as in the present case) or variable [1]. The tumors contain well-differentiated derivatives of the three germ layers: ectoderm, mesoderm, and endoderm. The most common derivatives are ectodermal structures such as skin, hair follicles and sebaceous and sweat glands [2, 3]. Neural tissues, including glia and neurons, are frequently present in benign cystic teratomas. A retina and choroid plexus may also be encountered [2, 3]. A well-differentiated cerebellum is extremely rare in teratomas; only 14 cases have been reported [4].

Histopathologically, tissues derived from all three germ layers were evident in the present slides, but surprisingly, the mature cerebellum had molecular Purkinje cells and

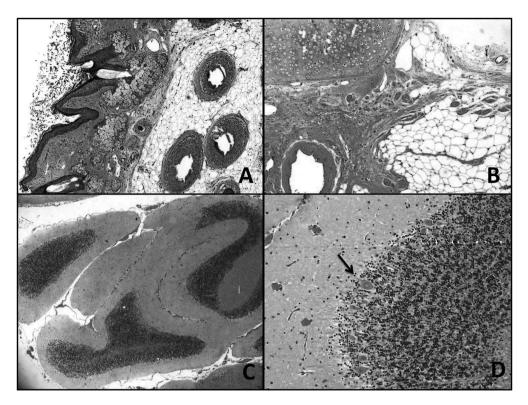


Figure 1 — A. The cyst cavity is lined by keratinized squamous epithelium, and skin appendages are present in the cyst wall, HE, ×40. B. Mature fatty and fibrous tissues and cartilage are evident in the cyst wall, HE, ×40. C. Well-differentiated cerebellar tissue is present within the tumor, HE, $\times 20$. D. The mature cerebellar tissue is composed of molecular-Purkinje cells with inner granular layers (black arrow indicated them, HE, ×100.

granular layers characteristic of the cortical layers of normal adult cerebellar tissue.

A foreign body giant cell reaction may also be present [3] and this reaction detected in certain areas of the cyst. Occasionally, the glial tissue may be cellular, and differential diagnosis among normal tissue, gliosis, and a lowgrade glial tumor developing within a teratoma, may be difficult [3]. One case of a primitive neuroepithelial tumor (similar to a medulloblastoma) within an ovarian teratoma has been reported [6]. The cerebral and cerebellar tissues of the present teratoma exhibited no suspicious features. Also, the authors were careful to examine many samples of the cyst wall and the glial tissue noticed upon gross examination. They are thus confident that no immature tissue was present.

The present patient experienced no complications. Possible complications of mature cystic teratomas include torsion, rupture, infection, and development of malignancy [5]. A case featuring torsion has been reported [7].

Mature teratomas can be treated conservatively by cystectomy or salpingo-oophorectomy [2]. The present authors completely resected the cyst. They report this case because, although the tumor itself is very common, the presence of well-differentiated cerebellar tissue is extremely rare in mature teratomas.

References

- [1] Medeiros F., Crum C.P.: "Germ cell tunors of the ovary". In: Crum C.P., Nucci M.R., Lee K.L., (eds). Diagnostic gynecologic and obstetric pathology. 2nd ed. Philadelphia: Elsevier Saunders, 2011, 904.
- [2] Zaloudek C.J.: "Pathology of germ cell tumors". *In:* Soslow R.A., Tornos C. (eds). *Diagnostic pathology of ovarian tumors*. New York: Springer, 2011, 168.
- [3] Baker P.M., Oliva E.: "Germ cell tumors of the ovary". In: Kurman R.J., Ellenson L.H., Ronnett B.M. (eds). Blaustein's pathology of the female genital tract. 6th ed. New York: Springer, 2011, 873.
- [4] Ishida M., Iwai M., Yoshida K., Kagotani A., Okabe H.: "Well-differentiated cerebellum within a mature cystic teratoma of the ovary". Int. J. Clin. Exp. Pathol., 2014, 7, 1255.
- [5] Talerman A., Vang R.: "Germ cell tumors of the ovary" In: Nucci M.R., Oliva E., Goldblum J.R. (eds). Gynecological pathology. London: Churchill Livingston, 2009, 520.
- [6] Boor P.J., Schoene W.C.: "Fetal cerebellar tissue associated with a primitive neuro-epithelial tumor in an ovarian teratoma". Can. J. Neurol. Sci., 1975, 2, 139.
- [7] Remadi S., Burkhardt K., Straccia A.T., Pizzolato G., Mac Gee W.: "Well differentiated cerebellar tissue within a mature cystic teratoma". *Pathol. Res. Pract.*, 1998, 194, 371.

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