YOLK SAC TUMOR IN THE CEREBELLUM : A CASE REPORT

TAKEDA, ZENJU

(Citation)
The Kobe journal of the medical sciences,31(2):63-72

(Issue Date)
1985-04

(Resource Type)
departmental bulletin paper

(Version)
Version of Record

(URL)
https://hdl.handle.net/20.500.14094/0100488757

Kobe J. Med. Sci. 31, 63/72, April 1985

YOLK SAC TUMOR IN THE CEREBELLUM
- A CASE REPORT -

ZENJU TAKEDA*, HIROMI MAEDA*, HIROSHI ITOH*,
KUNIO SHIRATAKI**, NORIO KOBAYASHI***,
AND KEIZOH SAKAMOTO***

*First Division, Department of Pathology
Kobe University School of Medicine

**Department of Neurosurgery
Kobe University School of Medicine

***Department of Neurosurgery
Kobe Children's Hospital
Takakuradai, Suma-ku, Kobe 654, Japan

INDEXING WORDS

yolk sac tumor; cerebellum; alpha-fetoprotein

SYNOPSIS

A 4-year and 7-month old boy with a cerebellar yolk sac tumor was treated with an enucleation of the tumor and post-operative irradiation. However, the titer of alpha-fetoprotein increased vigorously and a CT-scan showed an enhancing mass in the same primary location. He received the second operation by left hemi-cerebellectomy 6 months following the first operation. His clinical course is favorable until the age of 5 years and 3 months. Histological features revealed a typical yolk sac tumor with a reticular pattern with hyaline droplets in the stroma and the cytoplasm of tumor cells, and Schiller-Duval bodies like glomeruloid vessels. These hyaline droplets were positive in PAS

Received for publication : February 12, 1985
Authors' names in Japanese : 武田善樹　前田裕巳　伊東　宏
白瀧邦雄　小林憲夫　坂本敬三

and AFP staining. By transmission electron microscope, the droplets in the tumor cells were shown to be round dense bodies with a halo, less than 3 microns in diameter, with or without limiting membrane. Their cut surface revealed an irregularly mulberry like structure when observed by scanning electron microscope.

INTRODUCTION

Intracranial primary yolk sac tumors are very rare, 24 cases of primary tumor have been reported in the literature as far as we searched.[14] Most of cases originated in the pineal body and the para-pineal region of the brain.[1, 14, 16] This is the first cerebellar case ever to be reported. Furthermore, the tumor of this case was not located in midline, but in the left cerebellum. The presence of this tumor might be explained from the autonomous proliferation of heterotopic germ cells migrating to the cerebellum.[17] Higher alpha-fetoprotein titer in the serum and the cerebrospinal fluid decreased rapidly after the operation. Serological examination of alpha-fetoprotein can be helpful for diagnosis and detection of recurrence.[2, 7, 11]

CASE REPORT

The family and past history of the patient were not particular. He suffered from headaches, vomiting, and somnolence for several days at the age of 4 years and 1 month, and was admitted to Kobe Children's Hospital. The headache continued, sometimes vomiting occurred, and there was a slight impairment of consciousness but he had no focal sign. As the CT-scan revealed an enhanced mass in the left cerebellum, an emergency operation was performed for tumor enucleation with a left posterior-fossa-craniotomy. Post-operative CT revealed the disappearance of any enhancing foci. Then irradiation and clinical follow-up were performed, but the alpha-fetoprotein level increased at 4 years and 7 months. A CT-scan showed an enhancing mass in the same location at which the previous operation had been carried out. Upon a second surgical intervention a total tumor extirpation was performed along with one-third hemi-cerebellectomy. Post-operative irradiation was done, and his course is favorable at 5 years and 3

months old.

MATERIALS AND METHODS

Tumor masses obtained at operations were used for pathological examination. Histological sections were stained by HE, PAS, and immunohistochemical staining of AFP, CEA and HCG were performed by PAP method (DAKO Co., Ltd.), respectively. Specimens for the electron microscope were fixed in 2.5% buffered glutaraldehyde and 1% OsO_4 solutions. After the gradual dehydration, specimens for transmission electron microscope were embedded in Epon mixture. Their ultra-thin sections were stained by uranium acetate and lead nitrate, and observed by a Hitachi HS-8 electron microscope. On the other hand, specimens for the scanning electron microscope were preserved in isoamyl acetate after the dehydration. They were rapidly frozen by liquid nitrogen, and cut with frozen knife. After a drying process through critical point method, their cut surface was coated by gold ion sputtering, and was observed by Hitachi scanning electron microscope.

RESULTS

At the first operation, the enucleated mass weighing 15g showed grayish-white colour and elastic soft consistency. The mass obtained at the second operation showed grossly the same features with rather clear boundaries between the intact cerebellum, and weighed 6g (Fig. 1). Histologically, there were variously sized cystic areas with slightly mucinous fluid and loosely vacuolated meshworks. Papillo-tubular proliferation of tumor cells could be seen around capillaries, and subdivided cystic areas (Fig. 2). Cystic cavity channels communicated each other. Schiller-Duval bodies could be found in the myxoid stroma, covered with a lining of columnar of cuboidal tumor cells just like a glomeruloid structure (Fig. 3). Intra- and extra-cellular droplets were scattered in the vacuolated stroma, and were positively stained by PAS and AFP staining (Fig. 4). Slightly mucinous effusion in the cystic areas were also faintly positive by PAS and AFP staining, but not stained by CEA and HCG. Neither trophoblastic differentiation and structure resembling placental villi, nor epithelial differentiation forming glandular structure resem-

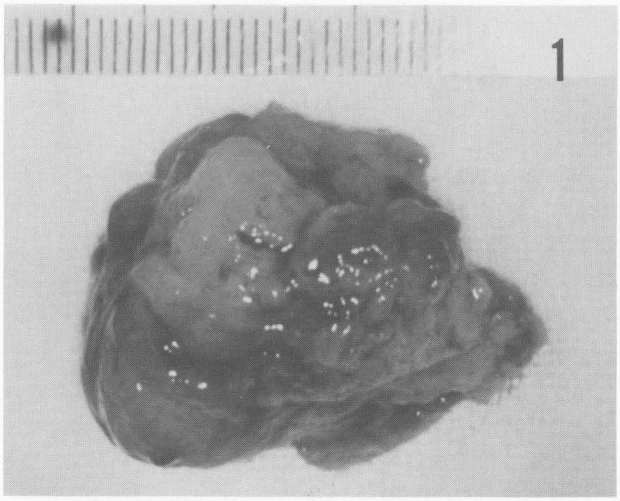

Fig. 1 A grayish white tumor embedded in the intact cerebellum, obtained at the second operation.

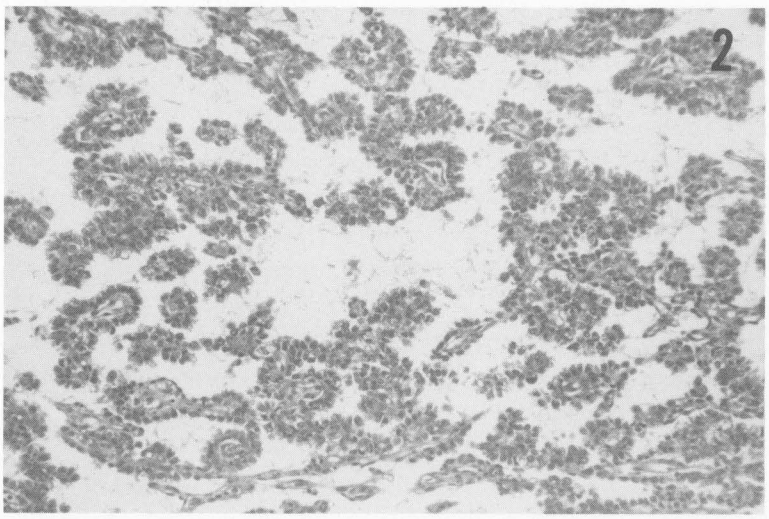

Fig. 2 Papillo-tubular arrangements of tumor cells in sheets or ribbons around vessels, subdivided into various-sized cystic areas. HE staining, x 173.

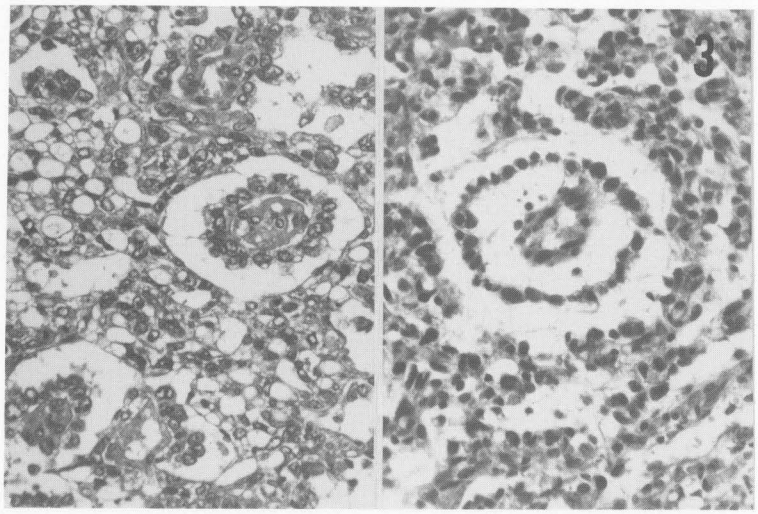

Fig. 3 Schiller-Duval bodies showing mantled vessels covered by cuboidal or columnar tumor cells. Left: HE staining, x 428, Right: PAS staining, x 428.

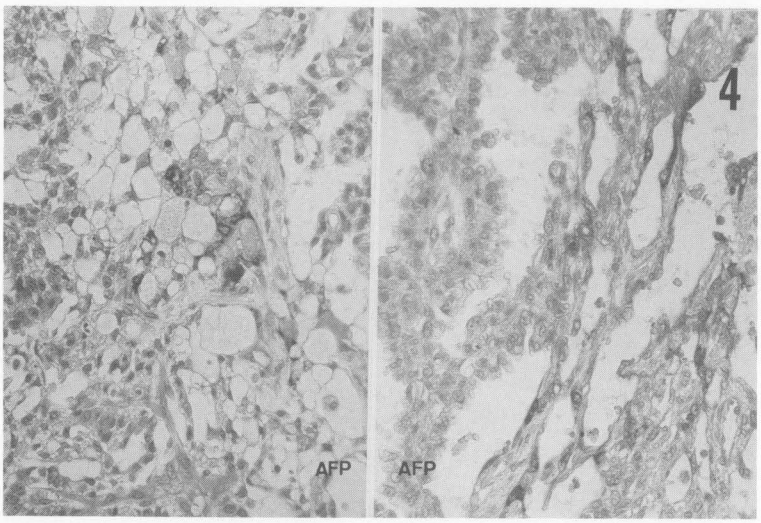

Fig. 4 Intra- and extra-cellular droplets showing positive alpha-fetoprotein scattered in the vacuolated myxomatous stroma (Left: alpha-fetoprotein staining, x 428) and cystic channel separated by strands lined by tumor cells (Right: alpha-fetoprotein staining, x 428).

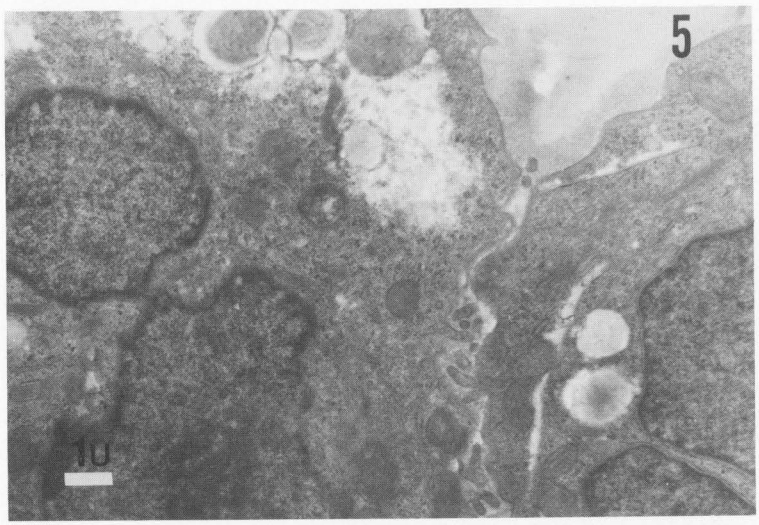

Fig. 5 Transmission electron micrograph showing electron dense bodies with halo and inter-cellular microvilli. Uranium acetate, and lead nitrate double staining, x 6075.

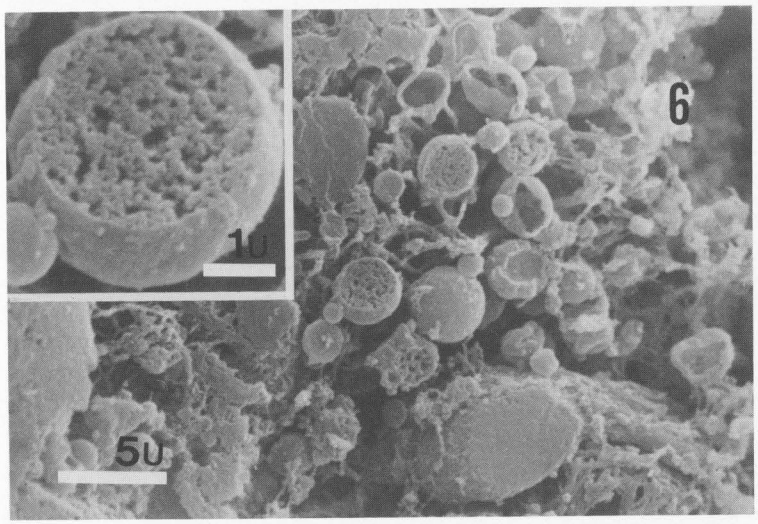

Fig. 6 Scanning electron micrograph showing many round bodies, with or without mulberry like materials at the cut surface. Ion gold sputtering, x 2850. Left upper frame, x 9600.

bling embryoid body, were seen in choriocarcinoma or in embryonal carcinoma. By transmission electron microscope, tumor cells had large and irregular nuclei with prominent nucleoli. They had many mitochondria and rough surfaced endoplasmic reticula, but few Golgi apparatuses. They showed micrivillous structure and occasional tight junctions between adjacent cells. Their cytoplasm contained round dense bodies with a halo, less than 3 microns in diameter with or without a limiting membrane (Fig. 5). Amorphous electron dense materials were also found in the loose cytoplasm or within dilated rough surfaced endoplasmic reticulum. Scanning electron micrographs revealed many round bodies covered by a capsule, with irregular mulberry like contents at their cut surface (Fig. 6). Furthermore, there were empty bodies without contents and mulberry like contents without a capsule in the loose cytoplasm.

DISCUSSION

Germinal cell tumors on gonads were classified into seminoma, embryonal carcinoma, choriocarcinoma and teratoma.[4] A group of embryonal carcinoma with a yolk sac structure was designated as endodermal sinus tumor by Teilum,[20] and yolk sac tumor by Huntington.[9, 10] The histological diagnosis of yolk sac tumor is based on:[5, 21] (1) Loose vacuolated network defined by flat mesothelioid cells, (2) Schiller-Duval bodies (endodermal sinuses), (3) Honeycomb pattern of microcysts (yolk sac vesicle), (4) Labyrinths of communicating cavities and channels, (5) Regularly arranged cysts lined by flat cells, (6) Polyvesicular vitteline pattern (supposed to recapitulate yolk sacs of blastocyst stage of embryogenesis), (7) Myxoid stroma (extra-embryonic mesoderm), (8) Compact aggregates of primitive cells, (9) Intra- or extra-cellular hyaline globules (alpha-fetoprotein positive). The extra-gonadal occurrence of yolk sac tumor is found in the saccrococcygeal region,[23] retroperitoneum,[15] anterior mediastinum,[22, 24] liver[8] and face, in addition to the pineal region.[1, 3, 16] However, a cerebellar origin of this tumor has not been reported to date.

The yolk sac membrane has the role of the encompassing and the protecting the fetus. Furthermore, it produces many serum

proteins at the fifth week of gestational age, such as pre-albumin, alpha-fetoprotein, alpha-1-antitrypsin and transferrin.[6] It fulfils a hepatic function during early embryogenesis. Histologically, alpha-fetoprotein positive droplets were variously sized in this case. Intra-cellular droplets showed amorphous low dense bodies covered with or without a limiting membrane. The contents of droplets were various morphologically, ranging from mulberry like solids to empty ones. Amorphous dense materials without a limiting membrane could be seen in the distended rough surfaced endoplasmic reticulum. From these findings, AFP droplets might be formed in the rough surfaced endoplasmic reticulum of tumor cells, and released after maturation into the interstitial and cystic lumina. Their secreted materials might be absorbed into draining capillaries, or mixed in cerebrospinal fluid, subsequently raised AFP titer in blood and cerebrospinal fluid.

Germ cells are located at the wall of the yolk sac during an early embryonal stage. They migrate toward the region of the genital ridge. Some of them failed to reach there, destined to survive as ectopic germ cells. Germ cell theory was hypothesized by Kleinsmith,[12] who suggested that their transplantation could induce multiple embryonal carcinoma associated with the potentiality for trophoblastic and teratomatous development.

Electron microscopic features of intracranial yolk sac tumor were occasionally reported in the literature, and were said to be similar to gonadal tumor. Stachura et al. reported characteristic basement membrane like materials in yolk sac tumor cells originating in the pineal region, but did not comment on the correlation with secretion of alpha-fetoprotein positive granules.[18]

REFERENCES

1. Albrechtsen, R., Klee, J.G., and Moller, J.E.: Acta Pathol. Microbiol. Scand. (Suppl.) 1972. 233. 32/38. Primary intracranial germ tumors including five cases of endodermal sinus tumor.
2. Allen, J.C., Nisselbaum, J., Epstein, F., Rosen, G., and Schwartz, M.K.: J. Neurosurg. 1979. 51. 368/374. Alpha-fetoprotein and human chorionic gonadotropin determination in cerebrospinal fluid.

3. Bestle, J.: Acta Pathol. Microbiol. Scand. 1968. 74. 214/222. Extragonadal endodermal sinus tumors originating in the region of the pineal gland.
4. Dixon, F.J., and Moore, R.A.: In:Dixon, F.J., and Moore, R.A., eds., Tumors of the Male Sex Organs (Armed Forces Institute of Pathology, Washington, D.C.). 1952. 55/104. Germinal tumors.
5. Everts, T.J., and Ransburg, R.C.: J. Neurosurg. 1979. 50. 246/252. Primary intracranial endodermal sinus tumor.
6. Gitlin, D., and Perricelli, A.: Nature 1970. 228. 995/997. Synthesis of serum albumin, prealbumin, alpha-fetoprotein, alpha-antitrypsin and transferrin by the human yolk sac.
7. Haase, J., and Nielsen, K.: Neurosurgery 1979. 5. 485/488. Value of tumor markers in the treatment of endodermal sinus tumors and choriocarcinomas in the pineal region.
8. Hart, W.R.: Cancer 1975. 35. 1453/1458. Primary endodermal sinus (yolk sac) tumor of the liver. First reported case.
9. Huntington, R.W., and Bullock, W.K.: Cancer 1970. 25. 1357/1367. Yolk sac tumors of the ovary.
10. Huntington, R.W., and Bullock, W.K.: Cancer 1970. 25. 1368/1376. Yolk sac tumor of extragonadal origin.
11. Itoh, T., Shirai, T., Naka, A., and Matsumoto, S.: Gann 1974. 65. 215/226. Yolk sac tumor and alpha-fetoprotein: Clinicopathologica study of four cases.
12. Kleinsmith, L.J., and Pierce, G.B.: Cancer Res. 1964. 24. 1544/1551. Multipotentiality of single embryonal carcinoma cell.
13. Loon, K.H., and Rassel Z.S.: Cancer 1979. 44. 1081/1085. Endodermal sinus tumor of the pineal region.
14. Murovic, J.A., Ongley, J.P., Parker, J.C., and Page, L.K.: J. Neurosurg. 1981. 55. 303/307. Manifestations and therapeutic considerations in pineal yolk sac tumors.
15. Norgard-Pedersen, B., Albrechtsen, R., and Teilum, G.: Acta Pathol. Microbiol. Scand. 1975. 83. 573/589. Serum alpha-fetoprotein as a marker for endodermal sinus tumor (yolk sac tumor) or a vitelline component of teratocarcinoma.
16. Prioleau, G., and Wilson, C.B.: Cancer 1976. 38. 2489/2493. Endodermal sinus tumor of the pineal region. Case report.
17. Rorke, L.B., and Fogelson, H.E.: Develop. Med. Child Neurol. 1968. 10. 644/650. Cerebellar heterotopia in infancy.

18. Stachura, I., and Mendelow, H.: Cancer 1980. 45. 2131/2137. Endodermal sinus tumor originating in the region of the pineal gland, ultrastructural and immunohistochemical study.
19. Tavcar, D., Robboy, S.J., and Chapman, P.: Cancer 1980. 45. 2646/2651. Endodermal sinus tumor of the pineal region.
20. Teilum, G.: Acta Pathol. Microbiol. Scand. 1965. 64. 407/429. Classification of endodermal sinus tumor (mesoblastoma vitellinum) and so-called "embryonal carcinoma" of the ovary.
21. Teilum, G.: In:Teilum, G., ed., Special Tumors of the Ovary and Testis and Related Extragonadal Lesions: Comparative Pathology and Histological Identification (Lippincott, Philaderfia, Tronto). 1976. 144/151. Endodermal sinus tumor.
22. Teilmann, I., Kassis, H., and Pietra, G.: Acta Pathol. Microbiol. Scand. 1978. 70. 267/278. Primary germ cell tumor of the anterior mediastinum with features of endodermal sinus tumor (mesoblastoma vitellinum).
23. Thiele, J., Castro, S., and Lee, K.D.: Cancer 1971. 27. 391/396. Extragonadal endodermal sinus tumor (yolk sac tumor) of the pelvis.
24. Vuletin, J.C., Rosen, Y., Brigati, D.J., and Gourin, A.: Chest 1977. 72. 112/114. Endodermal sinus tumor of the mediastinum, ultrastructure study.