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□ Case Report □

Intracranial Immature Teratoma with Syncytiotrophoblasts and Tumor Marker Positive Intestinal Lining Cells

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Intracranial teratomas are rare entities that can present as a pure type or as mixed germ cell tumor. Cases of mixed germ cell tumor composed of immature teratoma and choriocarcinoma have been reported. Also, immature teratoma can be mixed with only syncytiotrophoblasts. We report a case of immature teratoma with syncytiotrophoblasts of the brain discovered in a 3-year-old male baby. Serum human chorionic gonadotrophin (hCG) was normal and serum alpha-fetoprotein (AFP) was elevated. The tumor was mainly composed of intestinal glands, and neither endodermal sinus tumor nor embryonal carcinomatous elements were found. The cells lining the intestinal glands were positive for hCG and AFP. These findings suggest that the syncytiotrophoblasts are differentiated from the endoderm and AFP is not necessarily a marker exclusive to endodermal sinus tumor or embryonal carcinoma.

Key Words: Teratoma, brain, syncytiotrophoblasts, endoderm, AFP

As intracranial tumors, teratomas are rare constituting about 0.5 per cent of the total. It can present as a pure type or as mixed germ cell tumor (Russel and Rubinstein, 1989; Burger and Scheithauer, 1994). Cases of mixed germ cell tumor composed of immature teratoma and choriocarcinoma have been reported (Inamo *et al.* 1991). Also, cases of immature teratoma mixed with only syncytiotrophoblasts have been found (Rueda-Pedraza *et al.* 1987; Russel and Rubinstein, 1989; Burger and Scheithauer, 1994). However, no case of immature teratoma with syncytiotrophoblasts has been reported as yet in Korea.

There has been many reports on the diagnostic utility of the immunohistochemical demonstration of alpha-fetoprotein (AFP) in areas of mixed germ cell tumors that contain endodermal sinus tumor or embryonal carcinomatous elements and of human chorionic gonadotrophin (hCG) in areas of syncytiotrophoblastic differentiation (Russel and Rubinstein, 1989). This case is, thus, unique in that neither endodermal sinus tumor nor embryonal carcinomatous elements were found despite elevated serum AFP level. Another unusual feature includes immunohistochemical positivity of intestinal lining cells for hCG and AFP.

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CASE REPORT

A 3-year-old male baby was admitted due to incidentally found scrotal enlargement. Cranial MR image showed an irregular, partially cystic heterogenous mass located in the pineal gland. Physical and neurological examinations revealed to be normal. The level of serum AFP and hCG were 36.2 IU/ml and 5.23 mIU/

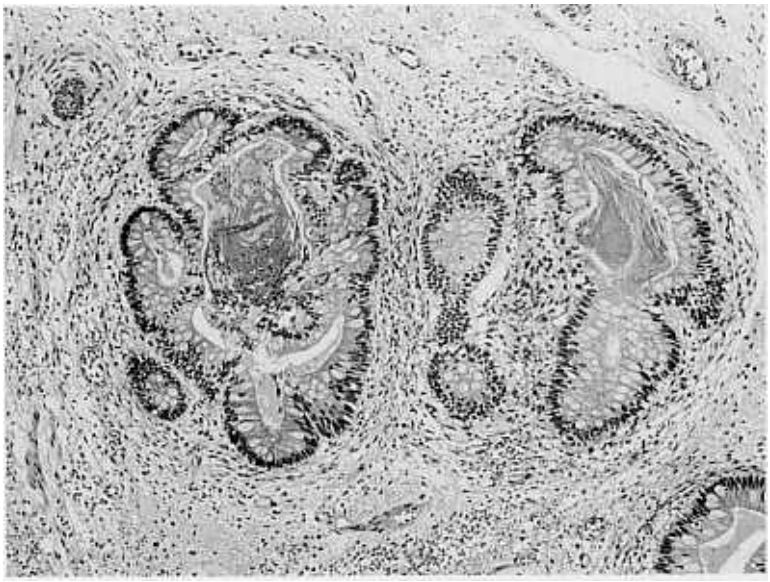

Fig. 1. *The tumor is mainly composed of intestinal glands.*

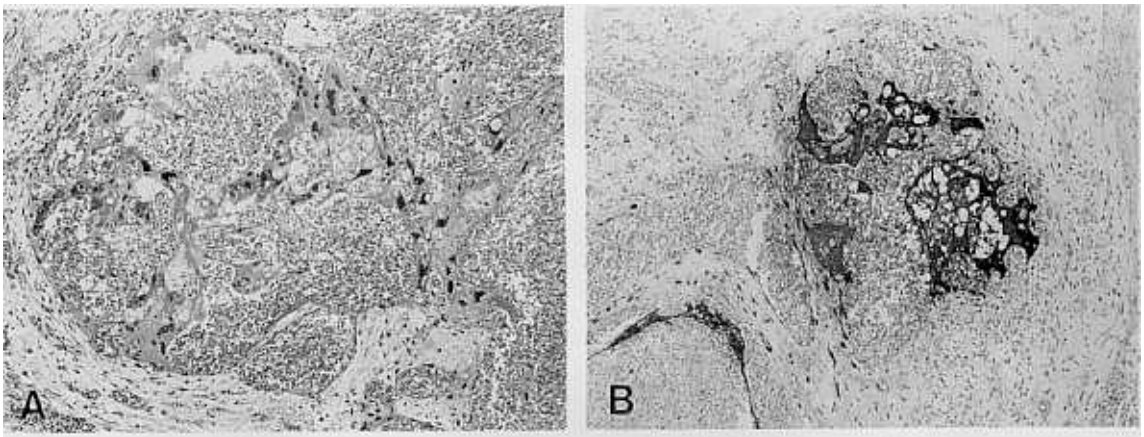

Fig. 2. *A cluster of multinucleated giant cells having basophilic cytoplasm is found within the tumor (left), and these cells are strongly positive for hCG (right).*

ml, respectively. Complete removal of tumor was done. Histologically, the tumor was mainly composed of intestinal glands lined by columnar cells and goblet cells (Fig. 1). In addition, stratified squamous epithelium, sebaceous gland, immature cartilage and adipose tissue were found within the tumor. In one area, multinucleated giant cells having abundant basophilic cytoplasm were noted (Fig. 2). These cells were positive for hCG (Fig. 2) and human placental lactogen. Thus, they were considered as syncytiotrophoblasts. The cells lining the intestinal glands were also positive for hCG (Fig. 3A) and AFP (Fig. 3B). The tumor was diagnosed as grade 1 immature teratoma by the criteria of Norris (Norris *et*

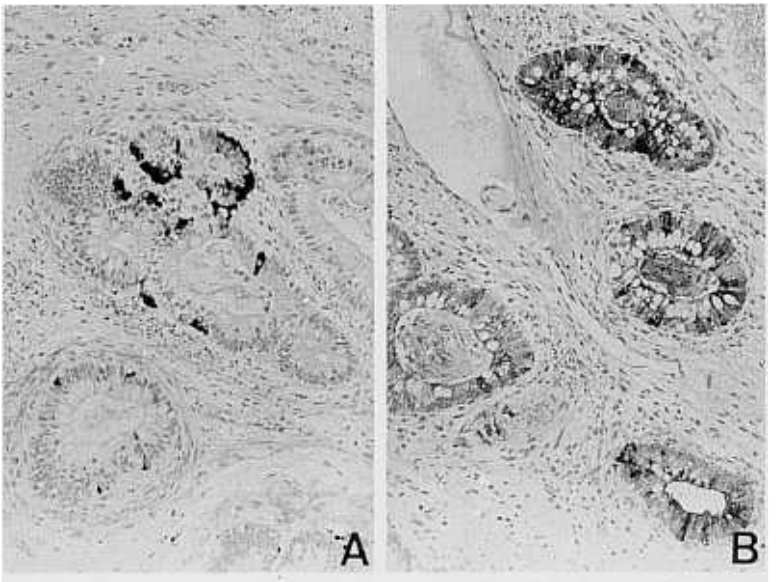

Fig. 3. The intestinal lining cells are positive for hCG (A) and AFP (B).

al. 1976).

After removal of the tumor, the level of serum AFP and hCG fell to 3.5 IU/ml and 2.60 mIU/ml.

DISCUSSION

As intracranial tumors, teratomas are rare constituting about 0.5 per cent of the total. The pineal body or its immediate neighbourhood is the most common site and other less frequently involved midline sites include the suprasellar region, pituitary fossa and the region of the fourth ventricle (Russel and Rubinstein, 1989). The extragonadal teratomas including intracranial teratoma arise from early embryonic cells or primordial germ cells in the course of migration during embryogenesis and they are considered as misplaced conjoined twin pregnancy (Saiga *et al.* 1991). According to the theory of fetus-fetus interaction, some germ cell tumors result from the incorporation of one member of a zygotic twin set into the body of the other. Subsequent destruction and involution of one twin result in a spectrum from conjoined monsters to fetus in fetu to fetus with teratoma (Rueda-Pedraza *et al.* 1987; Burger and Scheithauer, 1994).

Histologically, intracranial teratoma displays similar features as compared to other extragonadal teratomas. The identification of immature elements in teratomas is of prognostic importance, as it seems to herald an unfavorable course (Russel and Rubinstein, 1989). However, the presence of high grade immature neuroectoderm in congenital intracranial teratomas may simply reflect the tumor's presence in a more primitive host in contrast to ovarian teratomas which occur in an older age group (Rueda-Pedraza *et al.* 1987).

The elements of germinoma, embryonal carcinoma and choriocarcinoma may be present in the teratoma, and conversely, other elements of a teratomatous nature may be found on serial examination of intracranial germinoma or embryonal carcinoma (Russel and Rubinstein, 1989; Burger and Scheithauer, 1994). Mixed germ cell tumor consisting of choriocarcinoma and teratoma was reported (Inamo *et al.* 1991). Also cases of immature teratoma mixed with only syncytiotrophoblasts without cytotrophoblast were observed (Rueda-Pedraza

et al. 1987; Russel and Rubinstein, 1989; Burger and Scheithauer, 1994). Syncytiotrophoblast without cytotrophoblast must be distinguished from choriocarcinoma, which includes both, because the prognosis may be better for tumors containing syncytiotrophoblast only (Rueda-Pedraza *et al.* 1987). However, the effect of syncytiotrophoblastic component on the prognosis is not certain due to frequent presence of other tumor components (Rueda-Pedraza *et al.* 1987).

The diagnostic utility of the immunohistochemical demonstration of AFP in areas of mixed germ cell tumors that contain endodermal sinus tumor or embryonal carcinomatous elements and of hCG in areas of syncytiotrophoblastic differentiation have been firmly established. In conjunction, the diagnostic and prognostic value of raised levels of these proteins in the serum and cerebrospinal fluid harbouring germ cell tumors has been discussed (Russel and Rubinstein, 1989). Discrepancies occasionally arise when attempt are made to correlate the biochemical serum and cerebrospinal fluid data with the histological features of surgically removed tumor specimens. The reasons for discrepancy may be attributed to incomplete sampling of the tumor resulting in exclusion of other germ cell elements that were in fact present (Russel and Rubinstein, 1989).

Nogales et al discovered focal differentiation of the primitive endodermal epithelium into trophoblast-type cells which are capable of secreting hCG (Nogales *et al.* 1993). They attributed this phenomenon to three facts. First is the early spatial relationship between endoderm and trophoblasts. That is, in preimplantive stages the endodermal layer is found in close apposition under the trophoblasts and in the earliest postimplantation embryos the endoderm is represented by a thin layer within the trophoblast constituting a bilaminar omphalopleure. Second, simultaneous expression of both chorionic (hCG) and endodermal (AFP) markers may be found in the lung and stomach tumors. Third is the existence of intermediate cells that are found between the endoderm and trophoblasts in rhesus monkey.

In this case, identification of hCG positive intestinal lining cells suggets the origin of syncytiotrophoblasts as endoderm. The absence of endodermal sinus tumor or embryonal carcinomatous elements despite eleveted serum AFP level, and immunohistochemical positive reaction for AFP in the intestinal lining cells with subsequent return to normal level after removal of tumor, suggest that AFP is not necessarily a marker exclusive to endodermal sinus tumor or embryonal carcinoma, and the elevated level of AFP in immature teratoma may clinically be misinterpreted as a more malignant tumor.

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