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Case Report

Pilocytic astrocytoma with angiocentric arrangement: pathological features and differential diagnostic

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Abstract: Pilocytic astrocytoma is a low-grade glioma that occurs mainly in cerebellum of children, and young adults. The article is a rare case report of pilocytic astrocytoma with angiocentric arrangement in the supratentorial region of a 39-year-old female. Immunohistochemical markers, including GFAP, Vimentin, S-100, CD56, NeuN and Olig2, showed positive expression in tumor cells. The proliferation index of Ki-67 was about 1%. And we will analysis the clinicopathological features, diagnosis and differential diagnostic of pilocytic astrocytoma with angiocentric arrangement.

Keywords: Pilocytic astrocytoma, angiocentric arrangement, differential diagnosis, prognosis

Introduction

Pilocytic astrocytoma (PA) is a well circumscribed tumor with a low growth and has good prognosis and the 10-year survival of over 90%. It is classified as a grade I tumor by the World Health Organization [1]. Pilocytic astrocytoma occurs most commonly brain tumor in children and adolescents, can occurs in all of the neural axis, especially in the optic nerve, optic chiasma/hypothalamus, thalamus and basal ganglia, cerebral hemisphere, cerebellum and brain stem areas. The case occurred in the left occipital of adults, and has obvious angiocentric arrangement. Combined with the clinic pathological features, we herein report a typical case of pilocytic astrocytoma with angiocentric arrangement with pathological features and differential diagnostic.

Case report

On November 7, 2014, a 39-year-old female presented to the Third Affiliated Hospital of Soochow University suffering from headache and lags in response and progressive for two days. The patient had a headache, was swelling pain, especially in the left occipital, headaches persist and increased gradually nearly a week. The patient was lags in response nearly two days, and vomited two times, the vomitus was gastric contents.

Gross examination

The brain tissue was quasi-circular, off-white, about 3×3×2 cm in size. The section had nodules in the central, 2.5×2.3 cm in size, relatively well-circumscribed, off-white or off-red, reversible focal hemorrhage, light-tumor texture, no cystic degeneration, the section of focal had sense of gravel.

Neuroimaging

The computed tomography (CT) scan of the chest showed lumpy and iso- or slightly hypodense and markedly enhance with contrast media in left occipital, and calcification in fringe area, obvious compression in left ventricle, midline slightly right (**Figure 1A**). On MRI, PAs are typically slight hypo- or iso-intense, hypointense or mixed on T1 weighted images and hyperintense or mixed T2 weighted images. They are typically strongly and diffusely enhancing. They may contain cysts or consist of a tumor nodule in a cyst, mainly solid tumor (most with irregular contrast enhancement) (**Figure 1B**, **1C**). In this case, in contrast to diffuse intrinsic pontine gliomas, which infiltrate and expand primarily

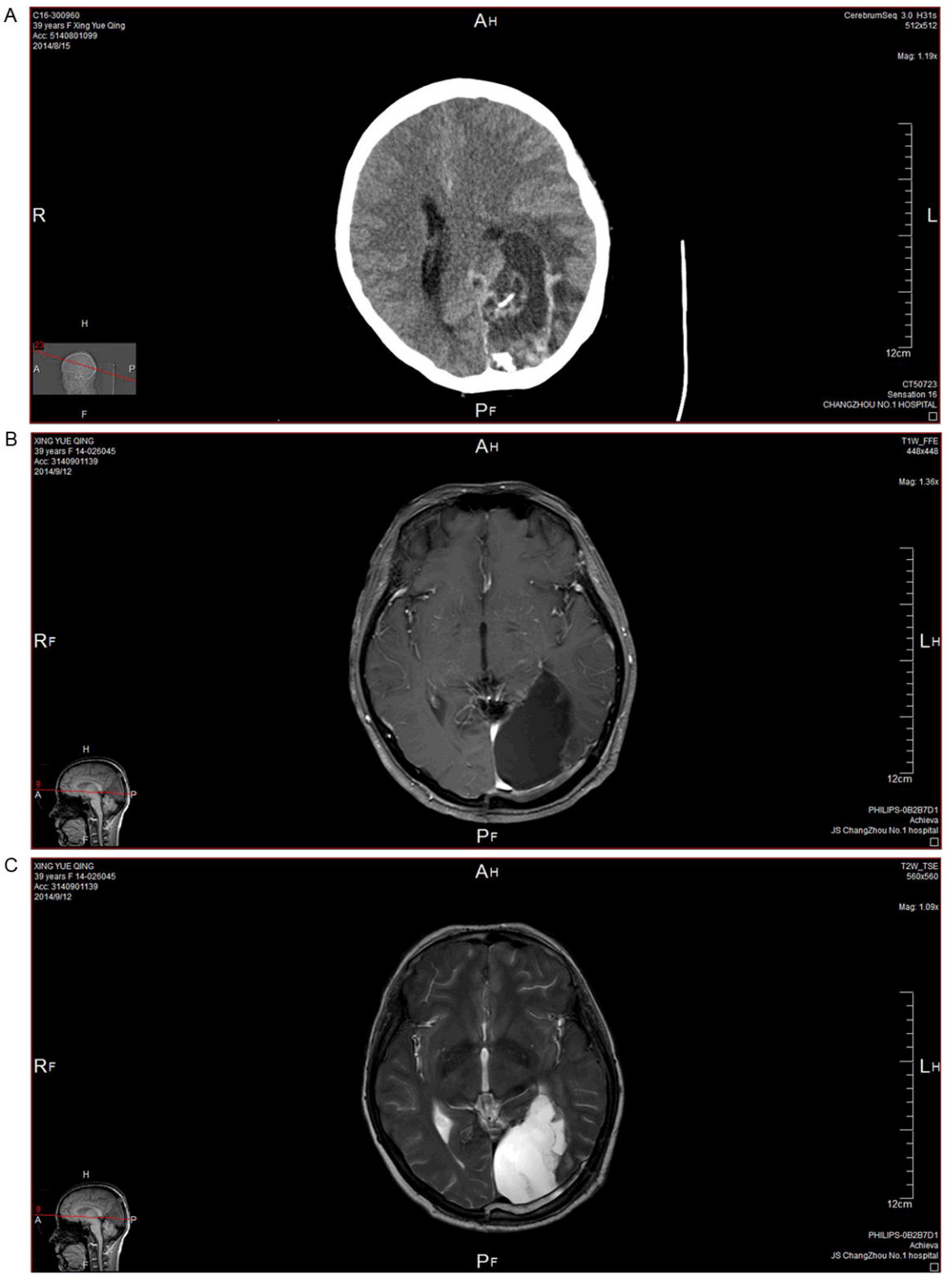

Figure 1. A: CT scan of the chest showed lumpy and iso- or slightly hypo-dense and markedly enhance with contrast media in left occipital, and calcification in fringe area, obvious compression in left ventricle, midline slightly right; B: Hypointense or mixed on T1 weighted images and hyperintense or mixed T2 weighted images; C: PAs are typically slight hypo- or iso-intense, hypointense or mixed on T1 weighted images and hyperintense or mixed T2 weighted images.

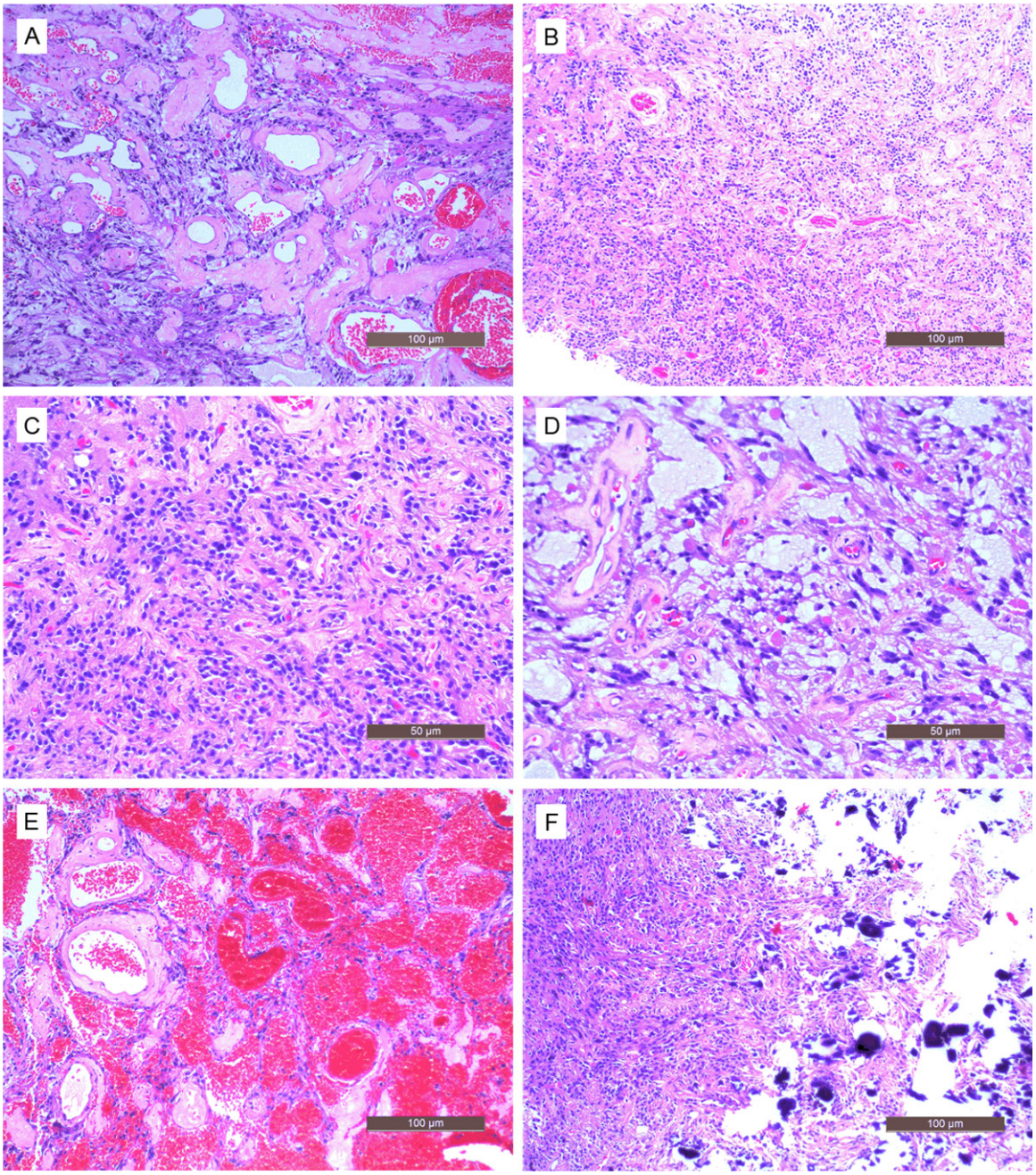

Figure 2. A: The tumor cells with round or oval nuclei with the characteristics of angiocentric arrangement, some vessel walls were glassy degeneration; B: Tumor cells revealed oval or short-fusiform; C: Cytoplasm showing slender hair-like protuberance of both ends, abundant cytoplasm and eosinophilic; D: The tumor of loose reticulum revealed mucoid degeneration, distinct micro-cystic structure formation, the nuclei showed round or ovoid, small soma, relatively short cytoplasmic process, arachnoid, and eosinophilic; E: Focal cavernous hemangioma; F: The tumor edge was with small calcification (magnification, ×100).

within the pons, PAs are generally located dorsally and have an exophytic pattern of growth (**Figure 1B**, **1C**).

Pathology

Optical microscopy revealed the tumor cells with round or oval nuclei with the characteristics of angiocentric arrangement (**Figure 2A**). Some tumor cells had two-way, showed compact fascicles and loose stellate reticulum. The tumor of fascicles reticulum were abundant, tumor cells revealed oval or short-fusiform (**Figure 2B**), cytoplasm showing slender hair-like protuberance of both ends, abundant cytoplasm and eosinophilic (**Figure 2C**), and

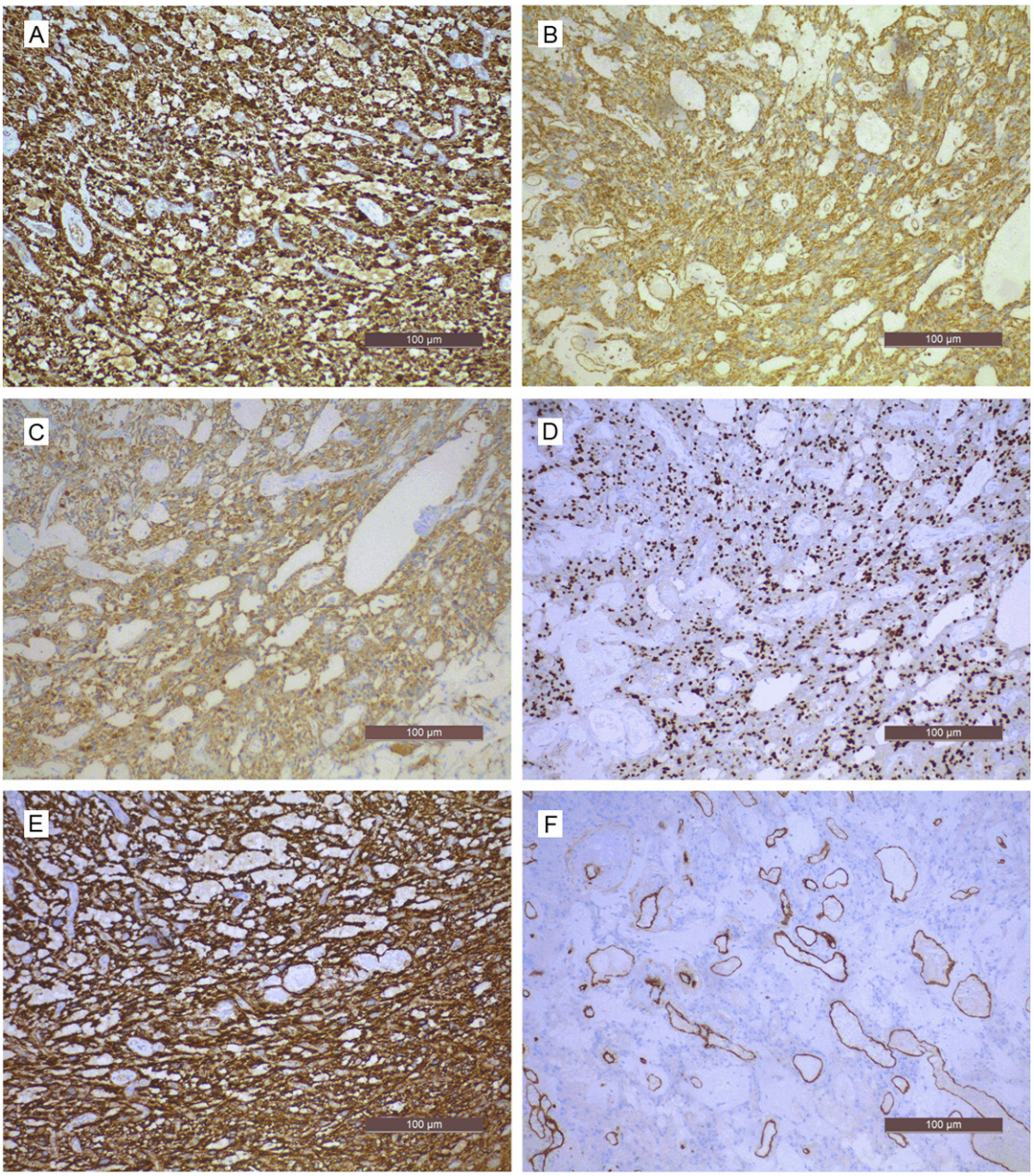

Figure 3. A: Immuno-staining showing tumor cells strongly positive S-100; B: Positive for Vimentin; C: Positive for GFAP; D: Positive for Olig-2; E; Positive for CD56; F: Positive for CD34 (magnification, ×100).

absence of Rosenthal fibers. The tumor of loose reticulum revealed mucoid degeneration, distinct micro-cystic structure formation (**Figure 2D**), the nuclei showed round or ovoid, small soma, relatively short cytoplasmic process, arachnoid, and eosinophilic granular bodies was easy to see. Blood vessels were rich in interstitial, and some vessel walls were glassy degeneration, focal cavernous hemangioma (**Figure 2E**). The tumor edge was with small calcification, no necrosis, no pathological fission and vascular invasion (**Figure 2F**). According to the immunohistochemical staining, the tumor cells were diffusely positive for GFAP, S-100, Vimentin, NeuN, Olig2, CD56 and CD34 (blood vessels) (**Figure 3A-F**), while the tumor cells

were negative for INA, Syn, IDH1, P53, EMA, and the proliferation index of Ki-67 was about 1%. Follow-up of about 9 months, the tumor did not relapse without postoperative radiotherapy or chemotherapy.

Molecular genetics

There were a number of reports about fusion gene between KIAA1549 fusion and BRAF, and mutations of BRAF V600E in some cases [2] and fusions between SRGAP3and RAF1 have also been found in rare cases. And Pilocytic astrocytomas carry duplication at chromosome band 7q34 containing a BRAF-KIAA1549 gene fusion in the majority of cases. IDH1 mutations are observed very frequently in adult astrocytomas and IDH2 mutations have been reported in some astrocytomas [3]. In this case, we did not find 12- exon, 13 exon of Kras, and BRAF V600E, and ROS mutations, also did not find P13K mutation.

Discussion

The term "pilocytic" to describe astrocytoma variants has been used since the 1937 to indicate cells with hair-like, bipolar processes. Almost all PAs are considered WHO grade I [2]. PA is the most frequent primary brain tumor below 20-year olds, no significant differences of gender. Pilocytic astrocytoma accounts for 10% of brain astrocytoma and 85% of the cerebellar astrocytoma [4]. The case was in the left occipital of adults. The patients who suffer from pilocytic astrocytoma usually have some nonspecific signs including focal neurological dysfunction, giant cranial disease, headache, nausea, vomiting, walking instability, blurred vision, and endocrine disorders and so on. All of these are due to the placeholder effect of tumor and ventricle. Tumors rarely involve the cerebral cortex, so rare epilepsy.

The main morphological features of pilocytic astrocytoma are with large number of myxoid background, bipolar spindle tumor cells arranged with radial around tumor cells vascular center. Tumor cells arrange in unipolarity, lack of density and osteoporosis bipolar organization form of pilocytic astrocytoma, and absence of Rosenthal fibers or eosinophilic granular bodies [5]. Compared with the pilocytic astrocytoma, some of pilomyxoid astrocytomas express Syn, and the proliferation index of Ki-67 is 2%-20%, the range is wide [6].

The histologic characteristics of *angiocentric glioma* are single bipolar spindle tumor cells arranged with single or multi-layer around the cortex blood vessels, rich blood vessels, but the vessels wall have no obvious hyaline degeneration. Immumohistochemical staining showed that spindles cells are positive for EMA (dot expression) [7]. This case can see large number of blood vessels with hyaline degeneration in the tumor tissue, show obvious dual-phase structure, Immumohistochemical staining was negative for EMA.

The tumor tissues of *diffuse astrocytoma (grade 2)* contain neoplastic-fibrous or gemistocytic astrocytoma, osteoporosis, loose mesenchyme, and can see micro-capsule structure, no dual-phase structure and absence of Rosenthal fibers or eosinophilic granular bodies [8].

The interstitial blood vessels of pilocytic astrocytoma is abundant, and can see cavernous hemangioma area, if the cavernous hemangioma region is dominant in the tumor and PA is a small amount located in the peripheral, may be misdiagnosed as a cavernous hemangioma. But the cavernous hemangioma has no spindle-shaped tumor cells or reticular tumor cell with bipolar tenuous hair structure around the blood vessels, main in the brain, rare in the cerebellum [9].

The preferred treatment of PA is Surgery. If the anatomical site allowed, total removal of tumors was necessary, and didn't use radiotherapy or chemotherapy drugs after removal. The adult pilocytic astrocytoma is easy recurrence, but follow-up of 9 months, the patient didn't use postoperative radiotherapy and chemotherapy, and had no recurrence so far.

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Disclosure of conflict of interest

None.

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References

[1] Alimli AG, Oztunali C, Okur A, Akarca O and Boyunaga OL. Intramedullary pilocytic astrocytoma. Spine J 2015; 15: e25-6.

[2] Collins VP, Jones DT and Giannini C. Pilocytic astrocytoma: pathology, molecular mechanisms and markers. Acta Neuropathol 2015; 129: 775-788.

[3] Korshunov A, Meyer J, Capper D, Christians A, Remke M, Witt H, Pfister S, von Deimling A and Hartmann C. Combined molecular analysis of BRAF and IDH1 distinguishes pilocytic astrocytoma from diffuse astrocytoma. Acta Neuropathol 2009; 118: 401-405.

[4] De Ioris MA, Carai A, Valente P, Angioni A, Randisi F, Cozza R, Romanzo A, Marras CE and Mastronuzzi A. Sporadic retinoblastoma and pilocytic astrocytoma: a rare association of two tumors. Pediatr Blood Cancer 2015; 62: 2245-6.

[5] Kibola AH, McClelland Iii S, Hlavin J and Friedman JA. Pilomyxoid astrocytoma in an adult woman: Case report. J Cancer Res Ther 2015; 11: 667.

[6] Kim S, Kang M, Choi S and Kim DC. Pilomyxoid astrocytoma occurring in the third ventricle. J Clin Imaging Sci 2015; 5: 41.

[7] Buccoliero AM, Castiglione F, Degl'innocenti DR, Moncini D, Spacca B, Giordano F, Genitori L and Taddei GL. Angiocentric glioma: clinical, morphological, immunohistochemical and molecular features in three pediatric cases. Clin Neuropathol 2013; 32: 107-113.

[8] Lind-Landstrom T, Habberstad AH, Sundstrom S and Torp SH. Prognostic value of histological features in diffuse astrocytomas WHO grade II. Int J Clin Exp Pathol 2012; 5: 152-158.

[9] Matsuno A, Nagashima T, Tajima Y and Sugano I. A diagnostic pitfall: Angiosarcoma of the brain mimicking cavernous angioma. J Clin Neurosci 2005; 12: 688-691.