Surgical Neurology International

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Letter to the Editor

A typical radiological presentation in a case of choroid plexus carcinoma

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Received: 03 November 13 Accepted: 27 March 14 Published: 07 May 14

This article may be cited as:
Salunke P, Sahoo SK, Madhivanan K, Radotra BD. A typical radiological presentation in a case of choroid plexus carcinoma. Surg Neurol Int 2014;5:63.
Available FREE in open access from: http://www.surgicalneurologyint.com/text.asp?2014/5/1/63/132103

Dear Sir,

Choroid plexus carcinomas are uncommon intracranial neoplasms accounting for 15-20% of choroid plexus tumors. About 80% of these are found in childhood.[3] The radiology usually shows a brilliantly enhancing heterogenous intraventricular mass invading the ependyma with edema.[9] However, the imaging may be atypical at times.[6] We describe here a case of choroid plexus carcinoma in an infant with atypical radiological features.

A 1-year-old male child presented with ataxia and intracranial hypertension since 2 months. The noncontrast computerized tomography (CT) scan showed a heterogenously hyperdense lesion in right posterior fossa region with gross hydrocephalus. The magnetic resonance imaging (MRI) revealed a posterior fossa lesion with the epicenter being located in the 4th ventricle, the presence of a solid and cystic component with extension into the right cerebellar hemisphere. The cystic component was uniformly hyperintense on MRIT1 and T2 sequences, and the fluid attenuation inversion recovery (FLAIR) sequence. The solid component appeared isointense on T1, T2 sequences and had a heterogeneous contrast enhancement [Figure 1]. Although diffusion weighted imaging (DWI) would provide us with added information, this was not available as the patient was referred to us with imaging. With the above image findings, we considered the cyst contents being high in protein or fat contents, and one of the differential diagnoses we kept in mind was an atypical teratoid tumor among other, which include ependymoma with cyst or bleed, ectopic cranipharyngioma, and infected dermoid. At surgery, the cyst had machine oil like fluid, usually characteristic of craniopharyngioma cysts.[1] The solid component of the tumor was soft, friable, and vascular with a clear margin separating the tumor from the brain stem with an absent interface and vermian infiltration at places. We achieved a gross total resection (GTR) and histopathology revealed a choroid plexus carcinoma [Figure 2].

The radiological imaging in our patient was unique in that the T1 and T2 sequences revealed a hyperintense signal suggestive of methemoglobin, high protein, or fat contents. However, in the absence of a fluid level we ruled out a prior hemorrhagic cyst. Furthermore, the CT scan images showed a slight hyperdensity within the lesion enabling us to rule out a high lipid containing lesion. The overall picture suggested a cystic solid lesion with high protein content. The differential in such cases would be craniopharyngiomas or atypical teratoid tumor and ependymoma with cyst.[1] Rare cases of ectopic fourth ventricular have been reported.[5] Craniopharyngiomas often appear isointense on T1, hyperintense on T2 with ring enhancement on contrast MRIs and, DWI shows no restriction.[1] Atypical teratoid tumor or teratomaand, ependymomas with a cystic component may share features with choroid plexus carcinoma. Atypical teratoid

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Quick Response Code:	Website: www.surgicalneurologyint.com
	DOI: 10.4103/2152-7806.132103

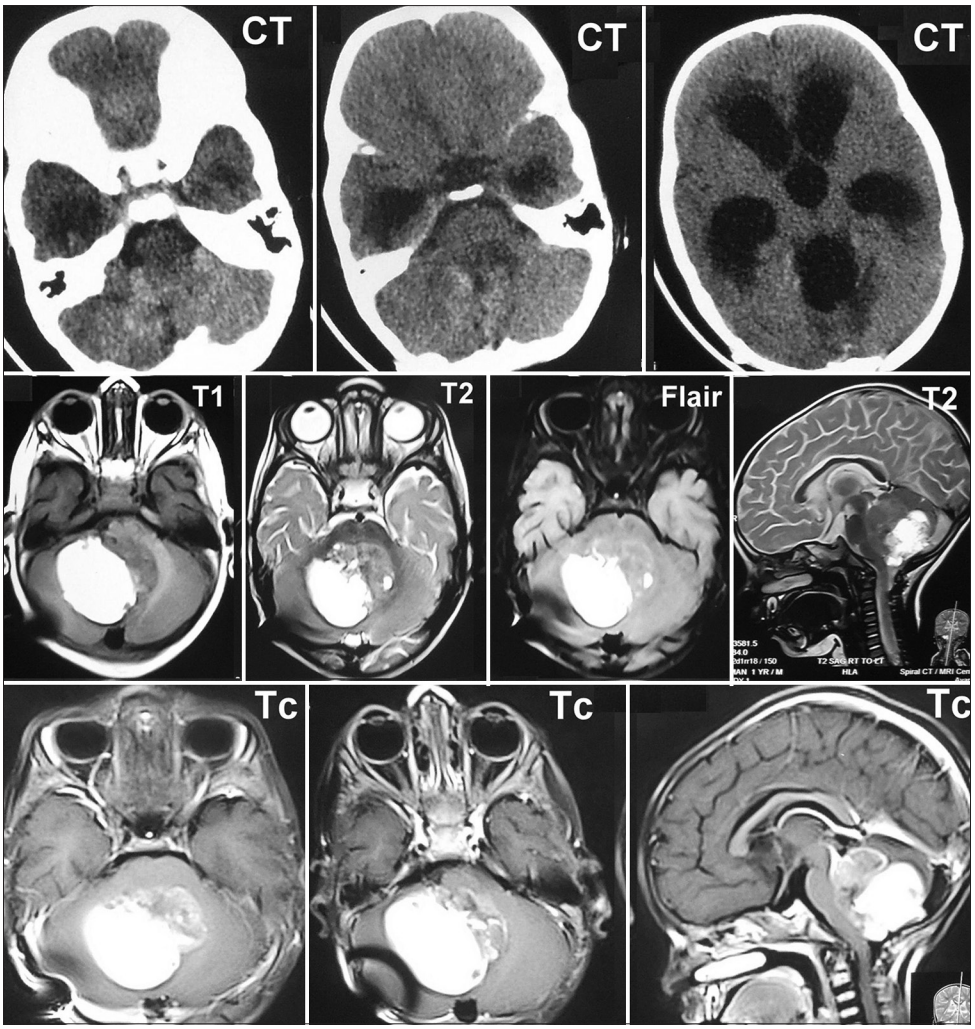

Figure 1: Noncontrast CT scan (First Row) showing heterogenously hypeerdense mass lesion in the posterior fossa compressing the fourth ventricle causing hydrocephalus. MRI showing posterior fossa lesion with cystic component hyperintense on T1, T2, and FLAIR (Second Row). Note the isointense solid component anterior to the cystic component seen on T2 sagittal image (second row extreme right). Solid component, ventromedial to the cystic componentiso to hypointense on T1, heterogenously hyperintense on T2 and FLAIR. Postcontrast MRI (Third Row) showing hyperintense cystic and contrast enhancing solid component without edema

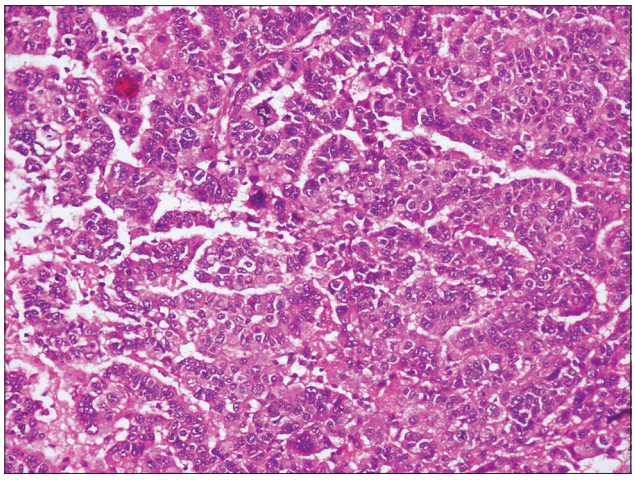

Figure 2: Micro-photograph shows solid as well as papillary architecture with nuclear atypia and many atypical mitosis. (H and E, ×20)

tumors are heterogeneously hypo-to isointense on T1, iso-to hyperintense on T2 with heterogenous contrast enhancement on MRI and shows mild to moderate diffusion restriction on DWI.[4] Ependymoma are heterogeneously hypointense on T1, hyperintense on T2 and FLAIR with homogenous contrast enhancement on MRI. DWI shows no restriction.[4]

Our case turned out to be a choroid plexus carcinoma. Tumors arising from the choroid plexus account for 0.4-0.6% of all intracranial tumors. Among these, 20-40% are choroid plexus carcinoma and 70% of them occur in younger than 2 years of age.[3] These lesions on MRI usually appear as contrast enhancing heterogenous large intraventricular mass that invades the ependyma or surrounding parenchyma with vasogenic edema. Usually they are isointense on T1 with areas of increased signal intensity due to hemorrhage, heterogenous hyperintense on T2 and homogenously

enhancing on contrast MRI. On FLAIR sequence, they appear heterogenous with surrounding edema. DWI show no restriction.[4] Heterogeneity is likely to be due to cysts, hemorrhage, necrosis, and calcification.[6,9]

Horská *et al*. studied MR Spectroscopy of childhood intraventricular lesions.[2] Both Choroid plexus carcinoma and papilloma was characterized by high levels of choline-containing compounds and a complete absence of creatine and the neuronal/axonal marker N-acetyl aspartate. However, the Choroid plexus carcinoma showed higher levels of choline compared with the choroid plexus papilloma, and it also had elevated lactate.[2] MRS was not performed in our patient. MRS may provide additional diagnostic information but is neither sensitive nor specific in these cases.

Treatment strategies include surgical resection followed by radiotherapy and/or chemotherapy depending on the extent of resection achieved. GTR of choroid plexus carcinoma increases the overall survival as well as progression free survival and is recommended if it can be safely performed. Five-year survivals are 58.1 ± 6.1% and 20.9 ± 5.1% for GTR and STR, respectively.[7] The role of adjuvant chemotherapy and/or radiotherapy in choroid plexus carcinoma is controversial, but increasing evidence suggests that combined radiation and chemotherapy should be given after subtotal resection.[8] Radiation is recommended following GTR in adults but not in children. Neoadjuvant chemotherapy (ifosfamide, carboplatin, and etoposide or ICE) regimen in choroid plexus carcinoma has shown to reduce vascularity, thereby increasing the chances of complete resection.[3] In case of subtotal resection, a reexploration to achieve a GTR may be attempted after completion of chemotherapy. This combined approach increases the survival rate.

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