CASE REPORT

Choroid Plexus Carcinoma in an Infant

W M Wan Muhaizan, MPath*, A A R Zurin, MSurg, M A Siti Aishah, MBBCh***

*Department of Pathology, **Division of Neurosurgery, Faculty of Medicine, Universiti Kebangsaan Malaysia, Kuala Lumpur

Summary

Choroid plexus carcinoma is a rare intracranial neoplasm, affecting mainly very young children. The commonest site is within the lateral ventricles and the prognosis is very poor. We report a seven month old baby boy who presented with raised intracranial pressure and seizures. Brain CT scan showed large intraventricular mass with calcification and hydrocephalus. Total macroscopic resection of the tumour was performed and diagnosis of choroid plexus carcinoma was made. However, the patient died 11 days after the tumour excision. The histopathology of this rare childhood neoplasm is discussed.

Key Words: Choroid plexus carcinoma

Introduction

The choroid plexus neoplasms - papilloma and carcinoma - comprise about 0.6 percent of all brain tumours[1,2] and 1-2 percent of all childhood brain tumours[3]. Choroid plexus carcinomas account for 17-30 percent of the choroid plexus tumour[1,2,4]. The incidence in childhood is much greater than adult. It is an agrresive tumour and the outcome in most cases are poor.

Case History

ZH, a seven month old baby boy who was previously well, presented with one week history of fever and projectile vomiting. He also had several episodes of generalized tonic clonic seizures which was resolved with valium and phenobarbitone. On examination, he was unconscious and extending to painful stimuli. All the limbs were stiff with brisk reflexes. The anterior fontanelle was tense and fundoscopy revealed bilateral papilloedema. He was intubated and ventilated for poor Glasgow Coma Score and control of intracranial pressure. Emergency CT Scan of the brain showed a large intraventricular mass occupying the posterior half of the left lateral ventricle (Figure 1). The tumour extended into the left parietal lobe and the brainstem with evidence of hydrocephalus and midline shift to the right. There was calcification seen within the tumour. A hyperdense lesion was also seen in the left parietal lobe suggestive of parenchymal haemorrhage. External ventricular drainage (EVD) was performed immediately to relieve the intracranial pressure. However, he developed a generalised subarachnoid and intraventricular haemorrhage subsuquent to the EVD. An emergency excision of the tumour via a craniectomy was performed the next day due to uncontrolled increase in the intracranial pressure. The tumour was soft, grey and extremely vascular. The tumour was totally excised and microscopic examination showed tumour cells mainly arranged in solid sheets as well as in papillary structures (Figure 2). The papillary fronds were lined by multiple layers of atypical cells. The cells exhibited round to oval mildly pleomorphic nuclei with moderate amount of cytoplasm. Areas with bizarre multinucleated tumour giant cells with numerous mitosis and some were aberrant was also seen. Mitotic counts were 37 per 10 high power field. Area of tumour necrosis and calcification was also noted. The tumour cells were positive for cytokeratin and negative for S100, carcinoembryonic antigen and glial fibrillary acidic

This article was accepted: 16 July 2003

Corresponding Author: Wan Muhaizan Wan Mustaffa, Department of Pathology, Faculty of Medicine, Universiti Kebangsaan Malaysia, Jalan Yaacob Latiff, Bandar Tun Razak, Cheras, 56000 Kuala Lumpur

(GFAP) protein. The diagnosis of choroid plexus carcinoma was made. The postoperative course was complicated by syndrome of inappropriate antidiuretic hormone secretion (SIADH) there was generalized cerebral swelling and persistently elevated intracranial pressure. Aggressive measures to control the increased intracranial pressure were instituted. Correction of hyponatraemia due to the SIADH was also done. Unfortunately, the patient succumbed to uncontrolled, elevated intracranial pressure 11 days after the tumour was excised.

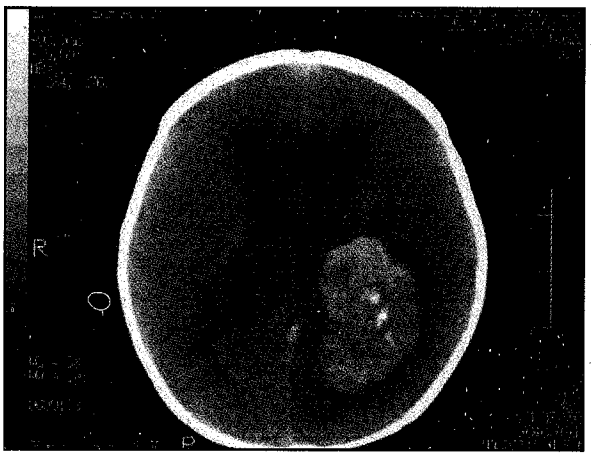
Discussion

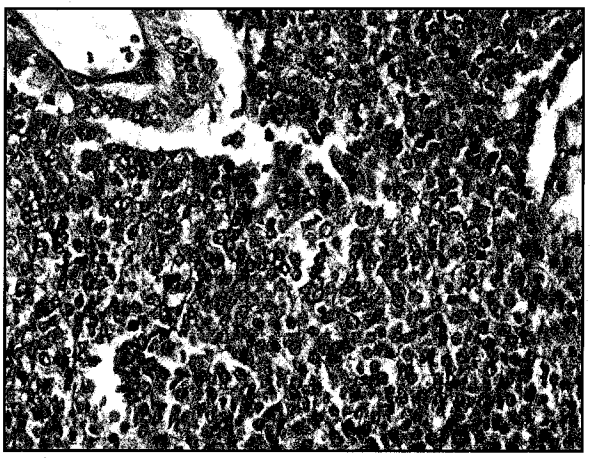
Choroid plexus carcinoma is a rare intracranial neoplasm that affects children in 91.8 percent of cases of which 44 percent are below the age of two years[4]. Occurrence in the neonatal period has also been reported as seen in our case[5]. There is moderate male predominance in many reported series. Lateral ventricle is the most common location of the choroid plexus carcinoma in paediatric age group followed by fourth ventricle and third ventricle. Posterior fossa location concur a worst prognosis as most of the tumour found in this location are poorly differentiated. Newbould et al reported 10 of his 12 choroid plexus carcinoma that occurred in the posterior fossa were of poorly differentiated category[6].

As the tumours are mostly located at the non-evocative site therefore they are not detected until they reach substanstial size. In our patient the tumour grow extensively involving ipsilateral parietal lobe and brain stem before he presented to the hospital with symptoms of raised intracranial which is the commonest presenting symptom in patients with this tumour[4]. Seizures or sign of hemisyndromes however are less common manifestations.

Neuroradiologic findings, although relatively stereotyped in patients with choroid plexus carcinoma, are not diagnostic. Its relation to the ventricles is a radiologic clue although other lesions such as papillary ependymoma, pilocytic astrocytoma and meningioma also can present at similar location. Choroid plexus carcinoma usually have irregular margins surrounded by hypodensities due to oedema with marked contrast enhancement. Accurate diagnosis of choroid plexus carcinoma on radiology may not be possible if there is extensive parenchymal invasion or the intraventricular origin of the tumour cannot be determined. Hydrocephalus, usually of obstructive type, is generally less severe than that found in the case of benign choroid plexus papillomas where hypersecretion of cerebrospinal fluid occurs. Tumour calcification which is present in this case is uncommon feature[4].

The diagnosis of choroid plexus carcinoma is based on histological examination; in typical cases as seen in our case, the tumour shows architectural dissarray with poorly formed papillae, high cellularity, severe cytonuclear atypia and a high mitotic index. It has great tendency to invade adjacent neural tissue. The distinction between choroids plexus carcinoma and papilloma is not always easy as atypical plexus papilloma may show significant cytonuclear atypia and scattered mitotic figures .





Choroid Plexus Carcinoma

Histologically, other malignant tumours such as malignant ependymoma and metastatic carcinoma may resemble a choroid plexus carcinoma. However, malignant ependymomas generally show clear-cut glial differentiation and GFAP staining. In adults, metastatic adenocarcinomas from the lung or other sites, are difficult to separate from choroid plexus carcinoma; therefore for many years the existence of choroid plexus carcinoma was questioned. However, because such metastatic lesions are rare in children the pathologic diagnosis is less confusing.

Poorly differentiated choroid plexus carcinoma occurring at posterior fossa can be difficult to distinguish from medulloblastoma as the location and the histology might appear similar[6]. Panels of antibody applied to the tumour tissue is helpful in the diagnosis[6]. The distinction is important as the behaviour of these tumours are different. These carcinoma usually shows immunohistochemical staining with epithelial markers such as keratin, CAM 5.2 and epithelial membrane antigen but compare to choroid plexus papilloma staining markers for glial differentiation such as S100 and GFAP is diminished or absent [4] in which these features were observed in our case. Herbert et al [10] recently described transthyretin is the excellent marker for primary choroid plexus carcinoma and is very helpful when the diagnosis is equivocal.

Some authors believed that CEA staining of the tumour heralds a poor prognosis in choroid plexus tumour[2] however this marker gave inconsistent findings in which Newbould et al[6] in his series of histologically confirmed moderately and poorly differentiated choroid plexus carcinoma, only three out of seventeen were positive for CEA and those that stained all were from moderately differentiated tumours.

The treatment for choroid plexus carcinoma varied from surgical resection alone to more aggressive therapy with surgery followed by radiotherapy and/or chemotherapy[4]. Most authors agreed that the tumour should be completely resected whenever possible. No difference was found between surgery alone and surgery followed by radiotherapy when complete excision was performed. Adjuvant therapy following surgery of choroid plexus carcinoma was useful when only partial resection of the tumour was obtained. There is no difference in disease-free interval between groups receiving either chemotherapy or radiotherapy after partial resection. As in most reported studies, the outcome of choroid plexus carcinoma was poor. Humphrey et al[8] in his series of six cases, only one was alive and disease free. In our case, the patient developed SIADH as a complication of subarachnoid haemorrhage (SAH) and intraventricular haemorrhage. Generalized cerebral oedema which ensued as a consequence of the SAH contributed to the persistent elevation of intracranial pressure and this led to the demise of the patient. This case illustrates a few important clinical points in which the clinical presentation of choroids plexus carcinoma is not unlike choroid plexus papilloma or other large space occupying lesions of the brain. The innocuous progression of the lesion can take on an accelerated clinical course if hydrocephalus develops. Rapid decompression of hydrocephalus can result in subarachnoid haemorrhage or intraventricular haemorrhage due to intratumoural bleeding.

References

1. Bohm E, Strang R. Choroid plexus papillomas. J Neurosug 1971; 18: 493-500.
2. Coffin CM, Wick MR, Braun F, Dehner LP. Choroid plexus neoplasms. Clinicopathologic and immunohistochemical studies. Am J Surg Pathol 1986; 10: 394-404.
3. Johnson Dl. Management of choroid plexus tumours in childhood. Pediatr Neurosci 1989;15: 195-206.
4. Geerts Y, Gabreels F, Lippens R et al. Choroid plexus carcinoma: A report of two cases and review of the literature. Neuropediatrics 1996; 27:143-48.
5. Ellenbogen RG, Winston KR, Kupsky WJ. Tumors of the choroid plexus in children. Neurosurgery 1989; 25: 327-35.
6. Newbould MJ, Kelsey AM, Arango JC et al. The choroid plexus carcinomas of childhood: histopathology, immunocytochemistry and clinicopathological correlations. Histopathology 1995; 26: 137-43.
7. Herbert J, Cavallaro T, JD Andrew. A marker for primary choroid plexus neoplasms. Am J Pathol 1990; 136(2): 1317-25.
8. Humphreys RP, Neomoto S, Hendrick EB et al. Chilhood choroid plexus tumours In: Marlin AE, ed. Concepts in Pediatric Neurosurgery. Basal: Karger,1987: 1-18.