PostScript

LETTERS TO THE EDITOR

Extraventricular unusual glioma in a child with extensive myxoid change resembling chordoid glioma

A 5-year-old boy was referred with a history of headache, vomiting, abnormal body movements and altered sensorium for 2 days. The past and family history was not significant. Neurological examination revealed features of raised intracranial tension—that is, intermittent tonic posturing of the body, asymmetric non-reacting pupils and blurring of the nasal margin of the fundus. In addition, he had increased tone in the right upper and lower limbs and upgoing plantar reflex. The child was intubated and treatment with mannitol and phenytoin was commenced. The provisional diagnosis was acute febrile encephalopathy; ceftriaxone and acyclovir were commenced. CT scan showed a mass lesion (7×5.5×4 cm) in the left parieto-temporal region with haemorrhage inside it. There was midline shift and the lesion was abutting the trigone of the left lateral ventricle (fig 1). He underwent surgery and the lesion was examined by histopathology.

Light microscopy revealed multiple fragments of a tumour admixed with blood clots. The tumour cells were arranged in cords and small clusters and were lying in a mucinous vacuolated background. The cells were relatively uniform, oval to polygonal, and had abundant eosinophilic cytoplasm (fig 2). No mitotic figures were seen. A few fragments showed limited glial differentiation in the form of coarsely fibrillar processes. No histological features of meningioma such as whorls, psammoma bodies or nuclear pseudoinclusions were identified. For immunohistochemistry, sections were treated with monoclonal antibodies (Dako Corp., Carpinteria, California, USA). Staining was carried out for glial fibrillary acidic protein (GFAP) (1:100), vimentin (1:200), S-100 (1:400), CD 34 (1:100), cytokeratin (CK) (1:150) and epithelial membrane antigen (EMA) (1:80) using the peroxidase–antiperoxidase method. The tumour cells were positive for GFAP, S-100 and vimentin, focally positive for CD 34 but negative for CK and EMA. A sample for electron microscopic examination was taken from the paraffin embedded tissue, therefore the ultrastructural findings could not be defined fully. However, the tumour cells showed cytoplasmic intermediate filaments, intermediate junctions and focal basal lamina. There were no cilia, complex interdigitations of cell membrane or well-formed desmosomes. Based on the histomorphology, immunohistochemistry and ultrastructural findings, unusual glioma with extensive myxoid change resembling chordoid glioma was diagnosed. The child had a cardiac arrest 2 days after the surgery and could not be revived.

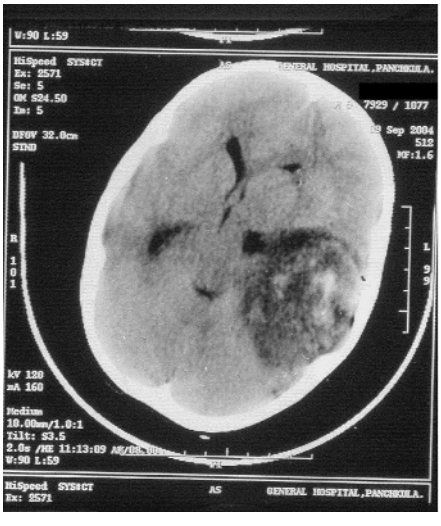

Figure 1 CT scan showing a large mass lesion in the left parieto-temporal region with midline shift.

Discussion

The features of the index case resembled chordoid glioma (CG) of the third ventricle, which was described in 1998 by Brat *et al* as a novel clinicopathological entity based on a series of eight cases.[1] Since the initial description, only 37 cases have been documented in the literature that all highlight the unique clinical, neuroradiological, and pathological characteristics of this lesion.[2] The immunohistochemical and ultrastructural studies indicate that CGs are glial in nature; they are placed in the category of glial tumours of uncertain origin in the latest World Health Organization classification.[3 4] CGs usually occur in adult patients (mean age 44.9 years, range 24–70 years), with a 1.7:1 female predominance.[5] To the best of our knowledge, only one case in a child (12 years) has been described previously.[6]

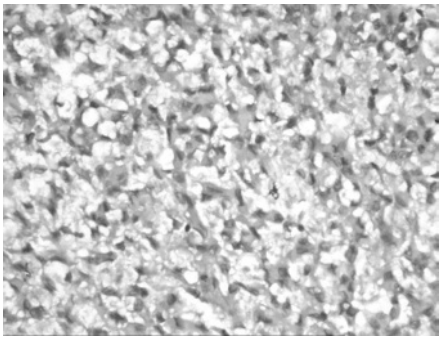

Figure 2 Photomicrograph of the tumour showing cords of tumour cells in mucinous vacuolated background (H&E, original magnification ×20).

Chordoid glioma shows a strikingly stereotypical anatomic localization in the hypothalamic/suprasellar/third ventricular region.[5] However, an unusual and multicystic component extending within the right temporal lobe and sella turcica was mentioned on MRI in only one of the cases by Brat *et al*.[1] In another exceptional case, the autopsy findings confirmed a tumour connection not only with the roof and floor of third ventricle but also with the right thalamus as well as a tumour extension within the hypothalamus and chiasmatic cistern.[7] CG has never been reported in cerebral hemispheres. In the index case, the tumour is located in the left parieto-temporal region and was abutting the trigone of the left lateral ventricle.

The microscopic findings were typical for chordoid glioma but there was no lymphoplasmacytic infiltrate. Strong lymphoplasmacytic infiltrate with numerous Russell bodies is a feature in CG, which was lacking in our case. However, an occasional case lacking lymphoplasmacytic infiltrate and Russell bodies has been described in the literature.[1] Another feature that is usually present is reactive astrocytes and Rosenthal fibres in the adjacent non-neoplastic tissue.[5] In the present case, no adjacent non-neoplastic tissue was included. However, the histomorphology, GFAP positivity and ultrastructural findings led us to make the diagnosis of unusual glioma with extensive myxoid change resembling CG.

The strikingly ''chordoid'' appearance of the neoplasms, with their eosinophilic clustered tumour cells in a blue mucinous matrix is distinctive among other regional tumours including pituitary adenoma, craniopharyngioma, pilocytic astrocytoma, and meningioma. The presence of GFAP reactivity and the absence of synaptophysin staining are inconsistent with pituitary adenoma. Pilocytic astrocytoma and craniopharyngioma bear even less of a morphological resemblance to chordoid glioma. Although there are histological similarities between chordoid gliomas and chordoid meningioma such as clustering of epithelioid cells, these meningiomas are typically dura based, and have a more prominent lymphoplasmacytic component often featuring germinal centres.[4] In the index case, the lesion was not dura based. The other features against chordoid meningioma were GFAP positivity, EMA negativity and absence of complex interdigitations of cell membrane or well formed desmosomes. Also, chordoid meningioma invariably contains foci of identifiable meningioma.[8] Another differential diagnosis is chordoma, which usually has a close relationship with the bone, but it is possible to find it in the intradural space without any apparent bone connection.[9 10] A strong inflammatory infiltrate can be seen in the stroma, but physaliphora cells are usually prominent. Moreover, intense immunoreactivity for CK, especially CK 8 and 18[10] and mitochondria–rough endoplasmic reticulum complexes on ultrastructural examination confirms the diagnosis of chordoma.[11]

Reifenberger *et al* showed that the genetic anomalies characteristic of astrocytomas, meningiomas and chordomas are not features

of CG of the third ventricle.[1,2] Ultrastructural study of chordoid gliomas lends support for their glial derivation. The presence of focal basal lamina formation and of microvilli in most cases suggests the possibility of ependymal derivation. The more definitive ultrastructural features of ependymoma, such as cilia and desmosomal junctions are usually lacking.[4] However, an occasional case with few tumour cells showing isolated abnormal cilia in the vicinity of the nucleus has recently been reported.[5] In the present case, based on negativity for ependymal features (EMA negativity and lack of microvillous structures on electron microscopy), GFAP positivity and a large number of intermediate filaments on ultrastructure, the possible origin of the tumour is astrocytic.

In summary, the index case is a 5-year-old boy with an unusual extraventricular glioma with extensive myxoid change resembling chordoid glioma, occurring in left parieto-temporal region.

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Metastatic placental site trophoblastic tumour in the duodenum presenting with a gastrointestinal bleed

A 31-year-old woman presented with altered bowel habits, black stools and profound weakness. Abdominal ultrasonography performed elsewhere had shown hepatic metastases. Oesophago-gastroduodenoscopy had been reported as normal. A red blood cell labelled blood pool study had shown a slow bleed at the ileocaecal junction, caecum and/or ascending colon. Except for pallor, her general condition was good. There was no supraclavicular adenopathy or ascites. She underwent lower gastrointestinal endoscopy at our hospital and was detected to have a rectal polyp. Because the size and appearance of the polyp did not correlate with the symptoms, the possibility of a separate pathology was considered. Consequently, the patient underwent an upper gastrointestinal endoscopy, which showed friable ulcerated lesions in the third part of the duodenum. The endoscopic differential diagnosis included carcinoma, lymphoma and tuberculosis.

The rectal lesion was a juvenile polyp. The duodenal biopsy specimen showed ulceration with neutrophilic infiltrates. The lamina propria contained a neoplasm composed of confluent masses of polygonal to round cells with large, vesicular nuclei. Some of the cells possessed smudged nuclei; a few multinucleate giant cells were also present. There were moderate amounts of eosinophilic cytoplasm; some cells contained clear cytoplasm.

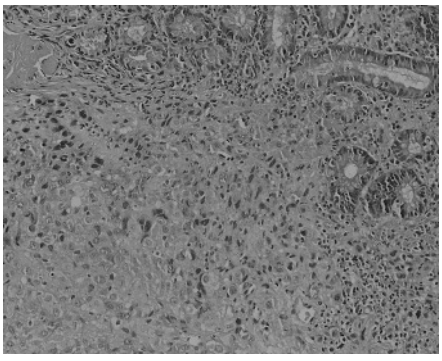

Figure 1 Duodenal neoplasm composed of sheets of polygonal to round cells with vesicular/smudged nuclei.

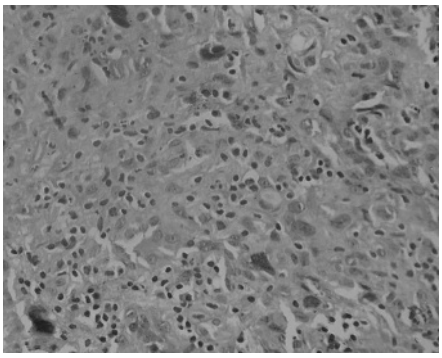

Figure 2 High power view of the tumour cells.

Mitotic figures were scarce. There was no desmoplasia, haemorrhage, necrosis or calcification. The tumour cells expressed cytokeratin (1:50) strongly and diffusely and were negative for CD45 (1:50) and CD30 (1:20) (all antibodies from Dakocytomation, Carpinteria, CA, USA).

Initially, a diagnosis of a high grade undifferentiated carcinoma involving the duodenum was considered. However, the unusual clinical features of a massive gastrointestinal bleed occurring in an otherwise healthy woman and the presence of hepatic metastases without abdominal adenopathy on ultrasonography were contradictory to a histological diagnosis of a high grade epithelial neoplasm. Moreover, there were some unusual morphological features: the bulk of the neoplasm was deep in the lamina propria and there was no dysplasia in the overlying epithelium. Further, the smudged cells raised the possibility of a trophoblastic neoplasm (figs 1 and 2). Further investigation showed a serum β-human chorionic gonadotropin (β-HCG) level of 11 000 mIU/ml. The slides were re-evaluated and a diagnosis of placental site trophoblastic tumour (PSTT) was made. The tumour cells expressed β-HCG focally and human placental lactogen diffusely (both Dakocytomation, Ontario, Canada; predilute, kindly done by Dr R Chetty). The patient was re-interviewed and we learnt that she had had an abortion recently. However, she refused further investigation or treatment and died a month later.

Primary duodenal choriocarcinoma as seen at autopsy and PSTT involving the duodenum and retroperitoneum in a known case of PSTT have been reported earlier.[1,2] However, we are unaware of any reports of metastatic PSTT presenting primarily as a duodenal lesion. Choriocarcinoma usually shows haemorrhagic areas, with focal expression of human placental lactogen and strong expression of β-HCG in the tumour cells. PSTTs possess sheets of tumour cells with diffuse human placental lactogen and focal β-HCG expression. The moderate rise in serum β-HCG levels in patients with PSTT, as in our case, correlates with the focal expression of β-HCG in the tumour cells.[3,4] The cells of epithelioid trophoblastic tumour are smaller than those of a PSTT and form cords and nests but no sheets; moreover, epithelioid trophoblastic tumour cells express human placental lactogen only focally. Differentiating between choriocarcinoma and PSTT is of importance because the former are exquisitely chemosensitive. PSTTs are relatively resistant to chemotherapy,