Malignant Oligodendroglioma in a Child: A Case Report

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Abstract:

Neonatal brain tumours are a rare clinical finding. Only few such cases are reported so far in the literature. The primary presentation in an affected child is usually discrete, non-specific and focal neurological symptoms. Clinically the development of the child was normal except for the macrocephaly. We describe a 9-month-old girl with a congenital anaplastic oligodendroglioma and a fatal outcome. In our case the diagnosis was made after the child died. The autopsy found a tumour formation 8/8/5 cm in size and 130g in weight in the frontotemporal lobe of the right brain hemisphere, which compressed the lateral ventricle. The microscopic examination revealed an anaplastic oligodendroglioma. Besides the routine hemathoxyline-eosine staining histochemical and immunohistochemical staining were used to prove the tumor diagnosis.

Keywords:oligodendroglioma, congenital, anaplastic

Introduction

Intracranial tumours are among the most prevalent in pediatrics. More frequent are only neuroblastoma in infancy and the neoplasms of the lympho-haemopoietic system in children[1]. The brain tumors arising in the neonatal period are different in etiology, histopathological type, topographical localization and prognosis in comparison with older children[2,3]. Teratoma is the most frequent brain tumor in infancy and together with astrocytoma, chorioid plexus pappiloma, ependymoma and meduloblastoma is responsible for about 80% of the neonatal brain tumours[1-3]. Oligodendroglioma is rare in newborns and only few cases have been described in the literature[4-7]. We describe a patient with an anaplastic oligodendroglioma, which presented clinically in the first months after birth with slight muscle tone disturbances and resulted in fatal outcome.

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Case Report

We present a case of a 9-month-old girl with congenital anaplastic oligodendroglioma with fatal outcome. According to the mother the infant had normal developmental milestone. The infant did not present with any neurological symptomes in the sense of vomiting or seizures and therefore no specific clinical examinations concerning a brain tumor were performed.



Fig. 1: Macroscopic view of the tumour

At autopsy it was observed that the fontanel was bulging and tense. The head circumference was bigger for the age by 3-4cm. The right hemisphere of the brain showed a tumor measuring 8/8/5 cm in size and weighed 130g, with a denser consistency than the surrounding brain tissue and well differentiated from it (Fig.1). The tumour was gray-white in color. On slice regions with necroses, hemorrhages and gelatinous areas were seen.

The microscopic examination after hemathoxylin-eosin staining showed a hypercellular

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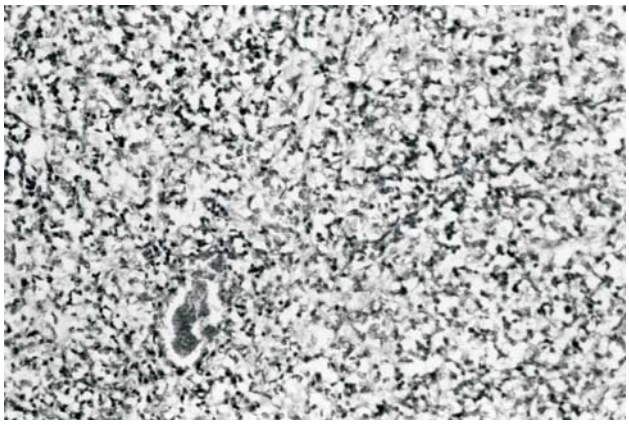


Fig. 2: Histopathology, H&E stain, original magnification x 100

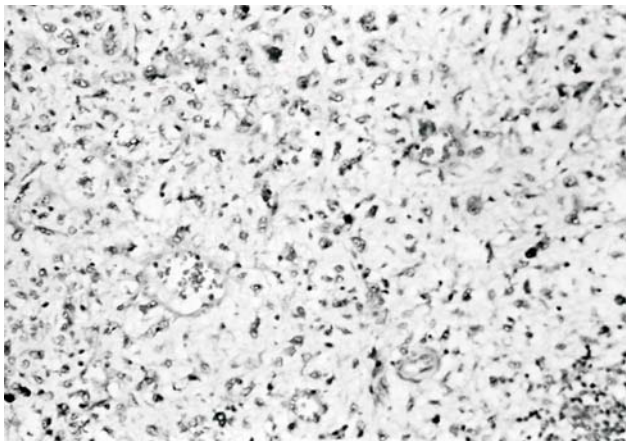


Fig. 3: Immunohistochemistry, S-100, original magnification x 200

tumor, consisting of tumor cells with hyperchromic round or oval nuclei, light cytoplasm and a gentle vascular net of branching capillaries (Fig. 2 H&E stain, original magnification x 100). Mitoses were seen, with areas of focal necrosis and endothelial vascular proliferation. The histochemical stain for mucopolysacharides with Schiff was negative and the stain for gliofi-brillas after PTAH was positive and outlined a typical "honey-cell" type picture. Immunohisto-chemically the tumor cells were positive for GFAP (+) and S-100 protein (++) (Fig. 3 S-100, original magnification x 200). According to the last revision of the classification of the nervous system tumors, WHO 2000, Lyon, France, the anaplastic oligodendroglioma is histologically rated as WHO grade III[8].

Discussion

Neonatal brain tumors are defined as those that manifested in the first two months of life[2]. They are rare and comprise only 0.5-1.9% from all brain tumors in pediatrics[1-3]. Congenital oligodendroglioma is rarer and only few cases have been described so far in the literature[4-7]. In a series of 165 diagnosed oligodendrogliomas in adults, the tumors were most often localized in the cerebral hemispheres and no sex predominance was observed[9]. Dohrmann et al[10] in a systematic study of the Tumor Register in Kynektikyt for a 41-year period, found only 12 verified oligodendrogliomas cases in children. The youngest patient in the series was 2-year-old and a male predominance was marked. Most of the oligodendrogliomas in these series also originated from the cerebral hemispheres except one, which originated from the brainstem. In previous reports there is no sex predominance and the tumor can be localized practically in any part of the brain including the medulla oblongata and the cerebellum[4-7].

Symptoms of intracranial neoplasm usually depend on the location of the tuomour. In older pediatric patients non-specific headache, generalised or focal epilepsy seizures and symptoms of upper motor neuron are observed in supratentorial lesions and the patients with infratentorial tumors usually present with symptoms of increased intracranial pressure, cerebellar symptoms and cranial nerves deficit[1]. The clinical manifestation of the neonatal brain tumors is usually discrete, non-specific and rarely includes focal neurological symptoms[2-3].

Wakai et al[2] reported two patients with congenital brain tumours and reviewed 200 patients with neonatal brain tumours at the age up to 2 months reported in the literature. Macrocephaly is the most frequent primary sign, which is observed in 62% of the symptomatic patients. Increased intracranial pressure with vomiting is reported in 14% of the patients. Seizures, irritability, facial palsy, muscle tone disturbances and other focal

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neurological symptoms are rarely reported. Buetow et al[3] reported 18% frequency of the tumour haemorrhage in 45 reviewed cases of congenital brain tumours. This might be due to the fast growth of the neonatal brain tumours. Congenital oligodendroglioma of medulla oblongata with unusual clinical manifestation (repeating apnea attacks), was first described by Koeppen et al[4]. In older patients the congenital oligodendroglioma presented with indirect hyperbilirubinaemia[11]. The cause of the jaundice might be secondary, due to the hemolysis of the intracranial haematoma, which is not rare in neonatal brain tumours. Most of the affected children presented with neurological syndrome including muscle tone disturbances, opistotonus and stupor in the first days of life. Few of these children might develop seizures[2-3]. Transcranial ultrasonography is of great importance for the early differential diagnosis. Computer tomography and magnetic resonance imaging are also useful tools for finding the intracranial tumours in newborns[2-3].

The diagnosis oligodendroglioma is made microscopically on Haematoxylin-Eosin stained slices. Immunohistochemical examination of the oligodendroglioma with different markers like S-100 protein, GFAP and Leu-7 is not reliable in this kind of tumour[6-11].

The survival rate in newborns with brain tumors is usually poor[2,3]. Chorioid plexus pappilomas and the astrocytomas have best prognosis. No optimal treatment in the neonatal brain tumors is established. Surgical intervention with total resection or stage surgery is the main goal. These methods are applicable but the risk of life-long injury should be considered. Chemotherapy and radiotherapy are not largely used in newborns[3]. In children less than 36 months of age irradiation is not recommended because of the risk of endocrine dysfunction[8].

In conclusion, oligodendrogliomas in children are extremely rare. The presentation is almost asymptomatic. The macroscopic and histological findings are typical. Research on the effect of the conservative therapy may be rewarding.

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