Melanotic neuroectodermal tumour of the pineal region

Abstract We describe CT and MR findings in a 23-month-old infant with a melanotic neuroectodermal tumour of the pineal gland. The tumour has been stereotactically biopsied and surgically resected. The pathological diagnosis was made on the resected piece. Embryology of the pineal gland and the histology of melanotic neuroectodermal tumour of infancy are discussed.

Keywords Pineal · Melanotic neuroectodermal tumour · Imaging · Embryology · Histology

Introduction

The melanotic neuroectodermal tumour of infancy (melanotic progonoma) (MNTI) is a rare tumour that most frequently involves the maxilla. The other most common sites of origin are the skull, the mandible and the brain. Most of the tumours occur in children under 1 year of age. Electron microscopy and histochemical studies have established the neural crest as the most likely origin [1]. Peripheral MNTIs are considered to have a benign course, while MNTIs occurring in the brain have a much less-favourable prognosis [2]. The treatment of MNTI consists of total surgical excision, eventually followed by radiation therapy and chemotherapy when the behaviour is malignant [3].

Case report

A 23-month-old child was hospitalised for assessment of a first generalised clonic convolution in a hyperthermic context. This child had been born prematurely (26 amenorrhoeal weeks) and had presented a grave hyaline membrane disease. The clinical and neurological examinations revealed a walking and psychomotor retardation without neurological focal signs.

A CT scan was performed during the infant’s hospitalisation, and demonstrated a heterogeneous midline mass associated with moderate hydrocephalus. The tumour was rounded, moderately hyperdense with grey matter and with a peripheral isodensity. No calcification was noted within the tumour (Fig. 1a). Its diameter measured 2 cm. Following contrast administration (Fig. 1b), only the central part of the tumour enhanced intensely.

An MR imaging was realized (Figs. 2, 3) and showed a well-outlined, round, pineal lesion. The central part of the tumour was hyperintense compared with grey matter on T1-weighted images, isointense on proton density, and hypointense on T2-weighted images. The peripheral component was hypointense compared with grey matter on T1-weighted images, and hyperintense on T2-weighted images and proton density. The gradient-echo sequence showed no susceptible magnetic artefacts within the tumour. After contrast administration, only the central part of the tumour showed intense and homogeneous enhancement. The tectum was discretely compressed and the splenium of corpus callosum superiorly displaced. Ventrices were moderately dilated.

Mesurement of tumour markers (alpha-fetoprotein, human chorionic gonadotropin-beta, carcinoembryonic antigen) in the serum before stereotactic biopsies was negative.
Two months after the initial hospitalisation, the child underwent stereotactic biopsies of the pineal region tumour. The pathological diagnosis was that it was a ganglioglioma, stage I, in the international classification. One month later the tumour was resected surgically. The tumour was located at the posterior extremity of the third ventricle, in the pineal region. It was very firm, and well-outlined. The cleavage was easy and the resection was macroscopically complete. The centre of the tumour contained a black pigment. The pathological diagnosis was of a ganglioglioma in the tumoral superficial tissues, and high content of melanin in the central tumour, and indicated a melanotic neuroectodermal tumour (Fig. 4).

The child is monitored every year, clinically and by MRI. The last cerebral MRI was in March 1999, and there was no sign of recurrence.

Discussion

We describe the sixth case of melanotic neuroectodermal tumour located in the pineal region. In three cases [4, 5, 6] the pineal gland itself could not be identified at the necropsy: the tumour was located in the site of the anatomic precursor of this gland. In one case [2] CT, angiography and ventriculography showed an extensive midline tumour of the third ventricle, originating from the pineal gland.

Melanotic neuroectodermal tumour of infancy is a rare, benign, pigmented lesion which most frequently involves the maxilla. In a review of 158 cases of MNTI, the most common sites of origin were the maxilla (68.8%), the skull (10.8%), the mandible (5.8%) and the brain (4.3%) [1]. The most frequent extracranial site was the genital organs.

Primary melanotic neuroectodermal tumours of the brain are rare. Rickert et al. [2] reported twenty-five, mostly located in the cerebellum. In five cases the tumour was sited in the pineal region. Brain parenchymal involvement results more frequently from extension of skull lesions [1, 7, 8] that are located in the anterior fontanelle [8].

The overwhelming number of lesions (95%) occurred in children under 1 year of age. There is no gender predilection,[1]. The local recurrence rate of this tumour was reported to be 10–15%. The malignancy rate in this review was 3.2%.[1].

In our patient, and in accord with previous reports [7, 9], CT and MR aspects revealed characteristics related to melanin contained in the central part of the tumour.

The formation of melanin is complex [10]. The mechanism is by phenolic oxidation into variations of quinoid structures that then autopolymerise into melanin. Melanin presents free-radicals, which possess non-paired electrons. This produces a paramagnetic effect, responsible for a shortened T1 relaxation time; this shortened time is correlated with the amount of melanin in the tumour [9]. It results in a hyperintensity on T1-weighted images, and hypointensity on T2-weighted images.

Two cases of melanotic neuroectodermal tumour (MNET) are described with an isointense signal on T1-weighted images and hyperintense signal on T2-weighted images; factors other than paramagnetic properties and the absolute amount of melanin might have been involved [11]. In two cases of intracranial meningeal melanocytoma [12] the signal was isointense on T1-weighted images and low on T2-weighted images; the pathological specimen showed tumour cells containing scattered areas of cytoplasmic deposition. The low signal on T2-weighted images was attributed to either fibrous content of the neoplasm or the paramagnetic effect of the melanin.

Hyperintense lesions on T1-weighted images suggest some differential diagnosis. On high magnetic field images, subacute haematoma, fat, proteinaceous effusions, calcification and tumours with high degree of cellularity (classically, cerebral lymphoma and glioma) have a hyperintense signal to brain on T1-weighted images.

In our patient, CT and MRI findings indicated that subacute haemorrhage, fat, high level of protein content and calcification were absent. Indeed the magnetic susceptible artefact on gradient-echo sequence was absent, no chemical displacement artefact or negative density on CT imaging were present and there was an intense enhancement after contrast administration.

The characteristics of signal of the tumour were correlated with its histology. The superficial tissues consisted of ganglioglioma, and the central tumour presented cord cells containing black pigment which was identified by the Fontana coloration method as melanin. According to the histological results after stereotactic biopsies, biopsied tissue might probably concern only the peripheral tumour. Histological studies revealed striking similarities between the pineal gland of foetus and infant, and pigmented neuroectodermal tumours [13]. Clarke and Parsons, in 1951 [14], suggested that the tumour origin might have a phylogenetic origin related to the median or pineal eye seen in certain lower vertebrates (lampreys). They used the term tumour of retinal anlage. Stowens in 1957 [13] selected the term progonoma to define this tumour due to misplacement of tissue as the result of foetal atavism to a stage which occurs in the ancestral forms of the species. He postulated that the precursor of this tumour might be some transiently existing embryonic structure of neuroectodermal origin because of the presence of melanin.

Recently others authors [1] suggested that the tumour was believed to be of neural crest origin. They based this hypothesis on studies of the ultrastructure and histochemistry, and in the observation of elevated urinary vanillylmandelic acid levels in a limited number of cases.