THE PINEAL PARENCHYMAL NEOPLASMS - A CLINICOPATHOLOGICAL STUDY OF FOUR TUMOURS

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ABSTRACT

Tumours of pineocytes are rare intracranial tumours. They are suspected once the effects of compression of neighbouring structures manifest. Four pineocytic neoplasms are reported, of which one was a spinal metastasis. Clinical features and radiological appearances are described. Histopathological diagnosis was based upon cytoarchitectural patterns which are discussed. Surprisingly all four cases occurred in a period of one month.

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KEY WORDS : Pineoblastoma; Pineocytoma; Pineal tumours.

Introduction

Tumours of the pineal gland region comprise less than one percent of intracranial tumours [1]. Only 30 percent of these are derived from the pineal parenchymal cells, namely pinealocytes and the astrocytes. The remainder neoplasms of the pineal gland are of germ cell origin. The rarity of pineal gland neoplasms has for long caused controversy of classification, microscopic appearance and biological behaviour. Only 66 pineal tumours have been reviewed in literature [2,3]. Ultrastructure and immunoreactivity has rarely been described. Clinicopathological details of four pineal tumours are described.

Material and Methods

Four pineal gland tumours were detected among 432 surgical specimens processed at CPL(WC) in July 93. Case records were retrospectively analysed with respect to clinical presentation and mode of diagnosis. The specimen were fixed over night in 10 percent formal saline, processed by paraffin embedding and 5 micron sections were cut and stained by haematoxylin and eosin.

Results

Three pineoblastomas and one pineocytoma were studied. All three pineoblastomas were in males, while the solitary pineocytoma was in a female patient (Table 1). The clinical presentation of the primary tumours was uniform, comprising of headache, gait ataxia, papilloedema & superior gaze palsy. CT scan localized the tumour below the third ventricle, in the pineal region (Table 1).

The metastatic pineoblastoma was localised at the intradural extramedullary region at D-8 vertebral level. It presented as lumbar radiculopathy one year post surgery of the primary and caused paraparesis after another 2 years (Table 1).

Microanatomically pineoblastomas comprised of oval cells having scant cytoplasm and a hyperchromatic nucleus with fine chromatin. Architecturally arranged in sheets, in lobules or in parallel strands along the long axis of adjacent vascular channels. At places an admixture of small and larger oval cells occurred. Case 3, interestingly, had psammoma bodies and glial differentiation. It displayed ill-formed rosettes (Homer Wright type) but showed no neuronal differentiation.

Case 4 showed classical large pineocyto-

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TABLE 1
Clinical and peroperative details

Case	Clinical features	Computed tomography	Peroperative features
1.	50 years male. Pineoblastoma operated May 90. Lumbar radiculopathy July 91. Paraparesis July 93.	Intradural, extramedullary space occupying lesion D-8 level.	Well encapsulated, greyish pink tumour 1 x 0.5 cm attached to dura.
2.	5 years male. Headache, vomiting 6 mth. Gait ataxia 3 mth. Superior gaze palsy, truncal ataxia, papilloedema.	Pineal space occupying lesion with hydrocephalus (Fig. 3)	Quadrigenal arachnoid thickened, opaque and vascular. A small greyish pink space occupying lesion (in case 2,3,4)
3.	24 years male. Headache, vomiting for two months superior gaze palsy, truncal ataxia, papilloedema.	Pineal space occupying lesion with hydrocephalus.	
4.	58 years male. Giddiness, vomiting for 2 years. Aggravated past 2 mths. Superior gaze palsy, truncal ataxia, papilloedema, nystagmus.	Pineal space occupying lesion with hydrocephalus.

Fig. 1 Case 4. Pineocytoma - two cell population and pineocytomatous rosettes. H & E x 100.

Fig. 2 Case 2. Pineoblastoma - cells separated into lobules by glial bands. H & E x 100.

matous rosettes, composed of large round cells surrounding neurofibrillary tangles. Cells were small and large present either in rosettes or sheets between vascular channels. The cellular areas were separated into lobules by glial tissue.

Discussion

The current classification of tumours of pineal region clearly separates those arising from pineal parenchymal cells from those of germ cell origin. The former originates from the two main neuroepithelial cells types that make up the histological structure of the pineal gland, namely the parenchymal cells, the

pinealocytes and the astrocytes. The classification stresses two unique features of pineal tumours, one the transitional examples which bridge two tumour types and second, the possibility of divergent differentiation of neuronal and glial forms within a tumour [4]. Rarely, the 'trilateral retinoblastoma' comprising of a pineoblastoma and a bilateral retinoblastoma may occur [5]. A pineoblastoma may display features of retinoblastic differentiation at its primary site in the form of Flexner rosettes or fleurettes [6]. Transitional forms between pineoblastoma and pineocytoma may exist, but the metastatic deposit from the tumour will invariably have

Fig. 3. CECT Scan brain showing pineal space occupying lesion with hydrocephalus.

microscopic features of a pineoblastoma [7].

Pineoblastomas are highly primitive and malignant. They disseminate widely through the cerebrospinal axis and leptomeninges [2,8]. Lobular pineocytomas, without the pineocytomatous rosettes essentially behave as apineoblastoma. Neuronal differentiation such as large rosettes suggest benign behaviour [9]. Some are of the opinion that the malignant pineocytomas should be classified as pineoblastomas. We have adopted this view as it has important clinical relevance even though it may be an over simplification taxonomically.

Ultrastructurally pineoblastomas are bland. They reveal end bulbs as seen in the human foetal pineal gland and 9 + 0 axial filament complex [10]. In pineocytomas the cells are organelle rich, contain dense core and other vesicles and have the characteristic 9 + 0 neurosensory cell axial filament [11].

In the four neoplasms presented, the histological distinction between pineoblastoma and pineocytoma has been based upon biologically meaningful cytoarchitectural morphology i.e the presence of pineocytomatous rosettes. Case 1 exemplifies cerebrospinal axis metastatic ability of pineoblastoma. The extensive psammoma bodies in case 3 cannot be explained although microcalcification is commonly seen in pineoblastomas. In this case there was morphological evidence of glial differentiation. Case 2, was a classical pineoblastoma while case 4 distinctively a pineocytoma. Thus these four cases represent the spectrum expected of neoplasms of the pineocytes. Suprisingly four cases of this rare neoplasm presented in a short period of one month.

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