Pilomyxoid astrocytoma in the adult cerebellum

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ABSTRACT

Pilomyxoid astrocytoma (PMA) is a recently recognised World Health Organization (WHO) Grade II tumour that was previously characterised as a subtype of the WHO Grade I pilocytic astrocytoma (PA). PMA has a histological appearance distinct from PA and a poorer prognosis due to its greater propensity for local recurrence and cerebrospinal dissemination. Although originally considered a paediatric tumour involving mainly the hypothalamic and chiasmatic region, reports of the lesion occurring in the adult population and other areas of the neuroaxis are emerging. We review the literature on PMA within the adult population and present the first case of PMA in the cerebellum of an adult female.

1. Introduction

The recently described pilomyxoid astrocytoma (PMA) is primarily a paediatric tumour with a predilection for the hypothalamic and chiasmatic region [1]. We present a case of PMA located in the cerebellum of an adult female and a review of the literature on PMA in adults.

2. Case report

A 20-year-old woman presented with 2 months of headache, nausea, vomiting and papilloedema characteristic of raised intracranial pressure. She was otherwise neurologically intact.

MRI (Fig. 1) demonstrated a large well-circumscribed intra-axial solid and cystic tumour within the left cerebellar hemisphere and vermis and extending down to the cervicomedullary junction. The remaining neuroaxis was normal.

A posterior fossa craniotomy was performed with macroscopic resection of the tumour (Fig. 2). Paraffin sections revealed a moderately hypercellular tumour on a myxomatous background, comprising a mixture of plump piloid cells with moderate nuclear pleomorphism. There were no Rosenthal fibres, no mitoses, no microvascular proliferation nor was there any necrosis. Immunohistochemistry was positive for glial fibrillary acidic protein, Nestin, ATRX (not mutated), MGMT (likely unmethylated), and negative for isocitrate dehydrogenase-1 and p16. The topoisomerase index was 2% (Fig. 3).

There was no adjuvant therapy and the patient was tumour free and well 5 months postoperatively.

3. Discussion

PMA was for a long time considered an infantile subtype of the World Health Organization (WHO) Grade I tumour, pilocytic astrocytoma (PA). Whilst the term "pilomyxoid astrocytoma" was first coined in 1999 by Tihan et al. [2], it was not until 2007 that the tumour was recognised as a WHO grade II tumour that was distinct from PA [1]. Both are abundant in piloid cells but PA is biphasic in appearance whilst PMA is monophasic with a rich myxoid background. PMA does not usually demonstrate the Rosenthal fibres and eosinophilic granular bodies that are characteristic of PA.

Komotar et al. compared 21 PMA cases against 42 PA cases and found higher recurrence in the former (76% PMA *versus* 50% PA, *p* value not specified) [3]. When matched for age, there was also significantly reduced mean progression-free time (25 months PMA *versus* 163 months PA, $p < 0.01$) and mean survival time (60 months PMA *versus* 23 months PA, $p < 0.001$). Cerebrospinal dissemination was found in three PMA patients but not seen in any of the PA patients. Higher recurrence rates of both PA and PMA have been found with nestin positivity of greater than 50% ($p = 0.047$) but not with tumour location or degree of resection [4].

PMA is reported to have an average age at presentation ranging from 10–18 months [1,3]. To our knowledge, there are only 21 adult patients reported in the English language literature (Table 1) [4–22]. A report of two adult cases did not specify age [11]. Of the remaining 19 patients, most (six patients, 32%) were in the third decade of life [5–7,13–15] with more even distribution across other age groups. Three patients (16%) were aged under 20 years [8,12,20], four (21%) were in the fifth decade of life [13,16,19,22] and three (16%) were in the fourth decade of life [4,10,21]. Three cases (16%) of patients older than 60 years have also been published [9,17,18].

In a paediatric series, there was a 57% incidence of PMA in the hypothalamic/chiasmatic (HC) region with less common locations being the cerebrum, basal ganglia/thalamus, cerebellum and fourth ventricle [13]. Of the adult cases, six have been reported in the HC region [12,13,18,20,22] and four in the spine [14,16,19,21]. It would appear that PMA has a predilection for the HC region in children, which is less evident in adults.

PMA in the cerebellum is uncommon. There have been 12 reported patients and only one of these was in an adult [4,13,17,23–29]. This was a 72-year-old man who underwent subtotal resection and died 4 months postoperatively [17].

To our knowledge our case is the first report of cerebellar PMA in an adult female. The location of the tumour in this patient afforded gross total resection. The influence of surgical resectability on tumour prognosis is yet to be fully determined. Further reporting of this uncommon tumour particularly within

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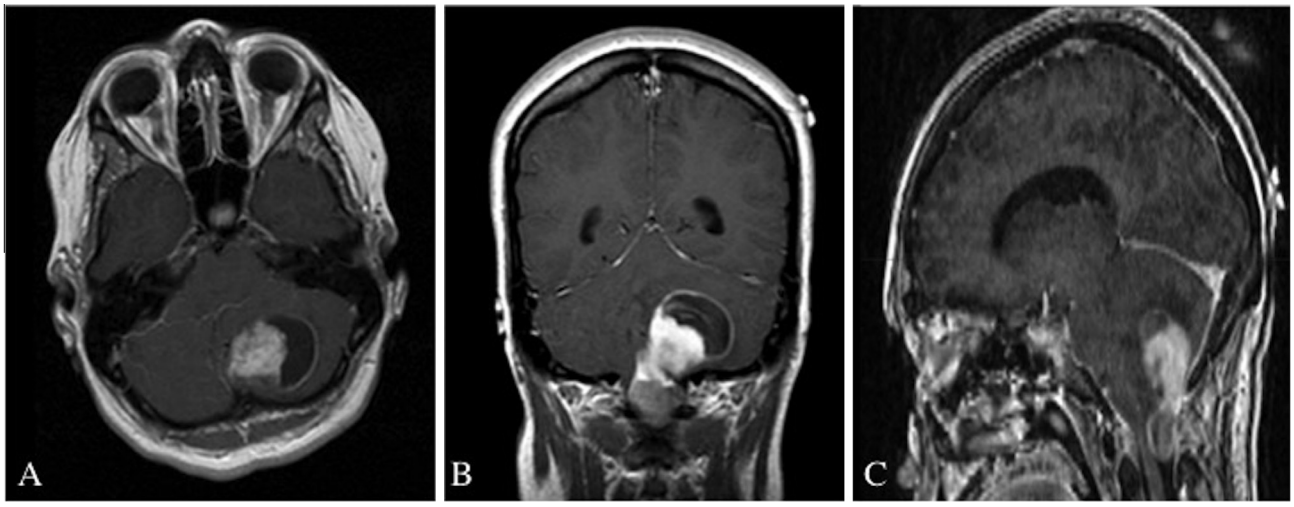

Fig. 1. (A) Axial, (B) coronal and (C) sagittal gadolinium-enhanced T1-weighted MRI demonstrating a large intra-axial solid and cystic tumour in the left cerebellar hemisphere and vermis with extension down into the cervicomedullary junction. Histopathology showed pilomyxoid astrocytoma.

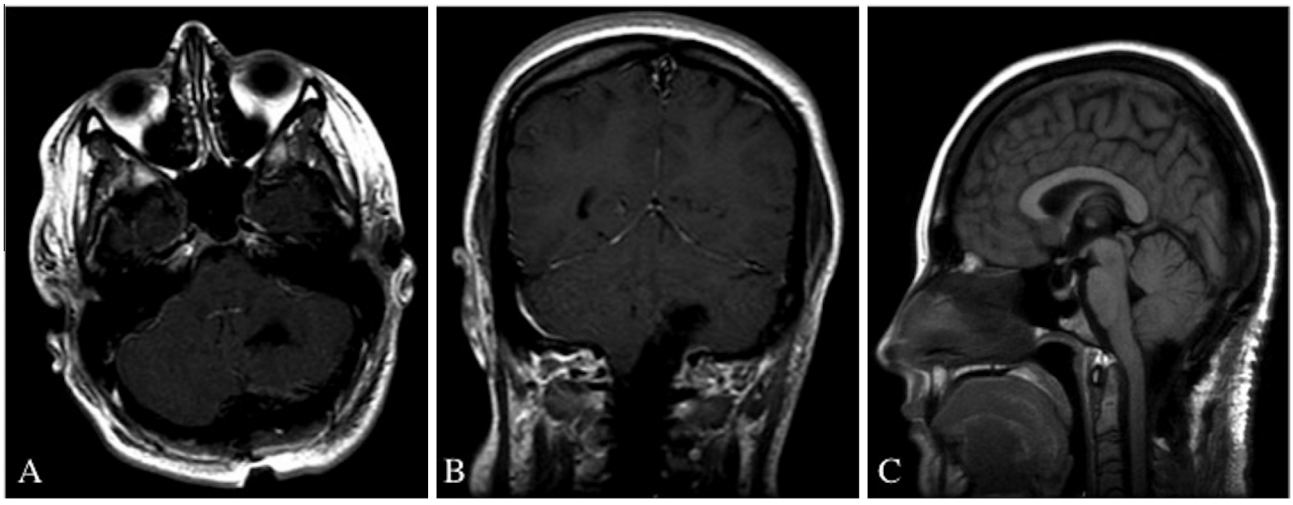

Fig. 2. (A) Axial, (B) coronal and (C) sagittal gadolinium-enhanced T1-weighted MRI demonstrating gross total resection of the pilomyxoid astrocytoma.

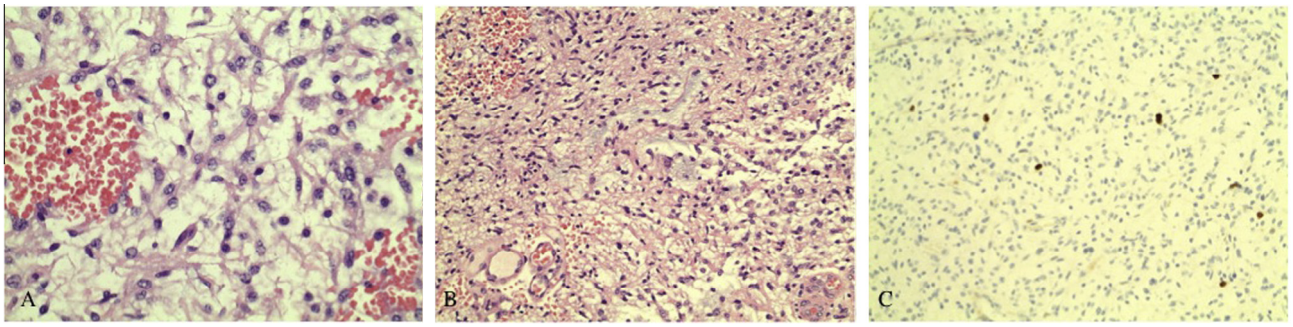

Fig. 3. Pilomyxoid astrocytoma. (A, B) Haematoxylin and eosin staining showing bipolar tumour cells in a diffusely myxoid background (original magnification × 200 in A; original magnification × 100 in B). (C) The topoisomerase index is about 2% (topoisomerase immunostain, original magnification × 100).