Clinical case

Cerebellar liponeurocytoma: Case report

Liponeurocytome cérébelleux : un cas

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ABSTRACT

Liponeurocytoma (lipomatous medulloblastoma) is a rarely and recently described tumor. We report an additional case of this uncommon lesion in an adult and we describe its clinical, radiological and histological features. A 45-year-old woman presented with symptoms and signs of increased intracranial pressure and cerebellar dysfunction. Computed tomography (CT) and magnetic resonance imaging (MRI) scans showed a heterogenous poorly circumscribed mass situated within the cerebellar vermis. After complete tumour resection, pathologic examination with immunohistochemical study confirmed the diagnosis. The postoperative course after 18 months was favorable with no evidence of tumor recurrence.

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RÉSUMÉ

Le liponeurocytome (médulloblastome lipomateux) est une tumeur rare récemment décrite. Nous en rapportons un nouveau cas chez l’adulte et nous décrivons ses principales caractéristiques cliniques, radiologiques et histologiques. Une femme de 45 ans présentait une hypertension intracrânienne associée à un syndrome cérébelleux. Le bilan scanographique et l’imagerie en résonance magnétique révélaient une masse hétérogène, mal limitée, siégeant au niveau du vermis cérébelleux. Après l’exérèse complète de la tumeur, l’examen histologique et l’immunohistochimie ont confirmé le diagnostic de liponeurocytome. L’évolution postopératoire après un recul de 18 mois a été favorable sans récidive tumorale.

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1. Introduction

Liponeurocytoma (LNC) are rare and slow growing neurotectodermal tumors located predominantly in the cerebellum (Buccoliero et al., 2005; Montagna et al., 2002; Patel et al., 2009). They were recognized in the revised WHO classification of tumors of the central nervous system 2000 as a distinct clinicopathological entity separate from medulloblastoma, in terms of prognostic, epidemiological and clinical aspects (Pasquale et al., 2009; Patel et al., 2009). To our knowledge, less than 20 cases have been reported up to now. The term of “cerebellar liponeurocytoma” omits the word “medulloblastoma” clearly underlining a better prognosis (Montagna et al., 2002). Liponeurocytomas are characterized by both neuronal and glial differentiation as shown by immunohistochemical staining (Owler et al., 2005). Because of the rarity and the relatively short follow-up data of the tumor, the natural history of cerebellar liponeurocytoma has not yet been defined (Pasquale et al., 2009; Patel et al., 2009).

In this report we describe an additional case of this uncommon lesion with successful management by surgery without radiation therapy. Characteristic features of cerebellar liponeurocytoma are discussed in light of the recent literature.

2. Case report

A 45-year-old previously healthy woman presented with a 1-year history of progressive occipital headaches, vomiting and unsteadiness of gait. Three months prior to admission, she experienced horizontal diplopia and increasing headache. Her consciousness was clear and her neurological examination showed signs of cerebellar dysfunction including ataxia, wide based gait with a positive march test and right dysmetria on finger-to-nose testing. Computed tomography (CT) and magnetic resonance imaging (MRI) scans showed an enhancing heterogenous and poorly circumscribed mass located within the cerebellar vermis, displacing the fourth ventricle and causing active supratentorial
hydrocephalus (Fig. 1). After complete microscopic tumor resection, pathologic examination revealed monotonous population of highly cellular neoplasm composed of small round and ovoid cells with scanty and eosinophilic fibrillary cytoplasm. Isomorphic cells containing vesicular nuclei and clear cytoplasm resembling oligodendrocytes. The tumor cells, in areas, were mixed with lipomatous cells (Fig. 2 A and B). Mitoses were very rare. There was neither capillary endothelial proliferation nor necrosis. Immunostains for glial fibrillary acidic protein (GFAP), synaptophysin and neuron specific enolase (NSE) were positive (Fig. 2 C and D).

At 18-month follow-up from the operation, without additional therapy, the patient was asymptomatic and MRI showed no tumor recurrence (Fig. 3).

3. Discussion

We found less 20 cases of liponeurocytoma reported in the literature (Montagna et al., 2002). Bechtel et al. (1978) first described a 44-year-old man with an unusual cerebellar tumor, which was composed of mature adipose tissue, medullomyomatous, astrocytous, oligodendromatous and ependymatous areas. Several synonyms have been proposed so as to emphasize its resemblance to central neurocytoma.

Because of its rarity and the paucity of long-term follow-up data, prognostication is difficult. Initial reports stressed a favorable prognosis for cerebellar liponeurocytoma, particularly in comparison to classic medulloblastoma. In accordance with this, liponeurocytoma was considered a grade I tumor in the WHO 2000 classification (Kleihues and Cavenee, 2000). However, reported cases with longer follow-up have indicated a substantial rate of recurrence, leading to its reclassification as a grade II tumor in the 2007 WHO revision. In one collective series, six of 15 patients (40%) with available follow-up information had recurrence during a period ranging from 1 to 12 years (Horstmann et al., 2004).

The mean age at diagnosis in liponeurocytoma was 51 years (ranging from 36 to 66). The predominant clinical disturbance was related to posterior fossa symptoms, all patients experienced signs of cerebellar dysfunction (Akhaddar et al., 2003). The initial
Fig. 2. Histologie of the tumor showing (A) monomorphic population of small round cells admixed with areas of lipid-containing cells mimicking mature adipocytes, (B) cells with round nuclei and clear cytoplasm resembling oligodendrocytes (hematoxylin and eosin stain, original magnification × 400), (C) Immunohistochemical staining for GFAP and (D) Neuronspecific-enolase (NSE) (original magnification × 400).

L’histologie de la tumeur montre (A) une population à petites cellules rondes et monomorphes associée à des plages de cellules à contenu lipidique simulant des adipocytes matures, (B) des cellules à noyaux ronds et à cytoplasmes clairs ressemblant aux oligodendrocytes (hématoxyline–éosine, agrandissement × 400), (C) l’expression immuno-histochimique de GFAP (D) Enolase neuronale spécifique (NSE) (agrandissement × 400).

Fig. 3. Postoperative MRI sagittal T1-weighted image after gadolinium administration.
En postopératoire : coupe sagittale en T1 après injection de gadolinium.

complaint in decreasing order of frequency is headache, gait disorder and visual deficit. Surgical resection is recommended as the initial treatment modality in order to establish a diagnosis (Patel et al., 2009), while additional external beam radiation therapy (EBRT) and chemotherapy are controversial (Buccoliero et al., 2005; Cacciola et al., 2002; Jackson et al., 2001; Valéry et al., 2004).

Preoperative diagnosis of liponeurocytoma is difficult mainly because of its non-specific radiological features. Cerebellar liponeurocytoma is typically well circumscribed but may show mass effects on adjacent structures (e.g., fourth ventricle) as in our case. On T1-weighted magnetic resonance imaging images, the tumor appears isointense to hypointense, with patchy areas of hyperintensity corresponding to regions of high lipid content. Contrast enhancement is often heterogeneous. On T2-weighted magnetic resonance images, the tumor appears slightly hyperintense to the surrounding brain, with focal areas of more pronounced hyperintensity (Nishimoto and Kaya, 2012). Peritumoral edema is typically absent or minimal.

The diagnosis of liponeurocytoma can be made only pathologically. Microscopically, it is a neoplasm with two distinct cellular elements: a predominant poorly differentiated component of small round to ovoid cells, with a scanty eosinophilic cytoplasm, intermixed with another element indistinguishable from mature fat cells at optic microscopy examination (Montagna et al., 2002). Electron microscopy and immunohistochemistry confirm that the lipidised cells are not mature adipocytes.
but neurocytes containing coalescent vacuoles of lipid (Owler et al., 2005). Our case illustrates interesting histopathological features. The unusual aspect of this case consists in the evidence of patchy zones that are indistinguishable from mature adipose tissue and also evidence of divergent neuronal and glial differentiation in the tumor. This was supported by GFAP and synaptophysin immunoreactivity in the glial and neuronal component of the tumor. The absence of mitotic activity, necrosis and endothelial proliferation suggest the low proliferative nature of these tumors and favorable prognosis (Akhaddar et al., 2003).

4. Conclusion

Cerebellar liponeurocytoma is to be considered as a rare but a distinct variant of medulloblastoma. Surgery to establish the diagnosis and remove the lesion should be the initial therapeutic step. The most common dilemma is whether to administer postoperative radiotherapy. The available follow-up data suggest a substantial rate of local recurrence, justifying its classification as a WHO grade II tumor. Further studies regarding the natural history of this lesion are warranted.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


