Histiocytosis mimicking a pineal gland tumour

Abstract We report an unusual case of isolated Langerhans cell histiocytosis of the central nervous system. A 19-year-old man presented with an incomplete ocular palsy. MRI revealed a solitary mass in the pineal gland with marked contrast enhancement. Complete microsurgical excision was followed by local radiotherapy. Histological examination revealed histiocytosis. Unifocal brain involvement by histiocytosis X is rare with few cases in the literature; the most commonly involved areas are the hypothalamus and the pituitary gland.

Keywords Pineal gland tumour · Histiocytosis · Magnetic resonance imaging

Introduction

Langerhans cell histiocytosis is characterised by abnormal proliferation of lymphocytes in tissue or blood with a granulomatous and inflammatory component [1]. Isolated central nervous system involvement is uncommon and few cases have been reported, in most of which the intracranial manifestations were detected with CT or MRI [2, 3, 4, 5, 6, 7, 8]. Involvement of the hypothalamus [3, 9], optic chiasm [4], cerebellum [10, 11, 12], brain stem [10, 13], meninges [14] and cerebral hemispheres [5, 6, 15, 16] has been described in isolated histiocytosis.

Case report

A 19-year-old man was submitted with an incomplete ocular palsy (Parinaud syndrome). The past medical history was noncontributory, as were clinical investigations. There was no skin lesion or lymphadenopathy. Sonography of the abdomen, bone scintigraphy and chest radiography were normal. MRI with thin (3 mm) T1- and T2-weighted images revealed a solitary mass of the pineal gland (Fig. 1). Its signal and the marked contrast enhancement suggested a pineal teratocarcinoma. The patient underwent resection of the mass, after which MRI showed no residual tumour (Fig. 2). Histological examination of the tumour revealed a cellular pattern of histiocytosis X. The patient therefore underwent radiotherapy with a total dose of 10 Gy and was discharged asymptomatic.

Discussion

Differential diagnosis of pineal region masses is extensive and includes a number of tumours, plus vascular lesions. More than a third of pineal region masses are neoplastic derivatives of pluripotential embryonic germ cells. They are often found during the second decade, beyond which tumours such as pineocytomas, arising from parenchymal cells can be found [14]. The other tu-
malignancy from parenchymal cells, the pineoblastoma, is found in the second decade [17]. MRI of these various tumours can be very similar, with marked contrast enhancement.

Involvement of the central nervous system is not uncommon in disseminated forms of histiocytosis, but isolated lesions within the brain are rare. The most frequently affected region is the suprasellar part of the brain [3, 4, 18], but the pons, cerebral lobes and cerebellum can be affected [2, 6, 18, 19, 20]. Haddad et al. [14] described intracranial disease presenting as lymphocytic meningitis. The lesions typically give high signal on T2- and are isointense in T1-weighted images [5, 6], and usually show marked contrast enhancement. However, George et al. [7] described a lesion giving low signal on T2-weighted images. On MRI lesions in the most commonly involved regions are described as having similar signal changes, with thickening of the pituitary stalk [21], which may be confused with sarcoidosis or tuberculosis [14, 16]. The lesion in the pons led to the diagnosis of a glioma or demyelinating disease [2] and cerebral lobar masses to diagnoses such as sarcoidosis, tuberculosis or parasitic disease [16]. Dural lesions should be differentiated from meningiomas and meningitis [22, 23].

According to the common management for solitary central nervous lesions, our patient underwent radiotherapy after microsurgical excision [24]. The outcome is usually good [6, 9], as in our case, but recurrence can lead to a fatal outcome [5]. This case demonstrates that, despite its atypical site, a solitary intracranial mass with marked contrast enhancement can be a manifestation of histiocytosis.

References