Plasma cell granulomas of the brain: pediatric case presentation and review of the literature

Abstract  Plasma cell granulomas (inflammatory pseudotumors) are benign inflammatory masses that have been observed in virtually every organ system but are most often described in the lung. Rare cases have been reported in which the brain and spinal cord are affected. We present the case of a 5-year-old girl with personality and behavioral changes, discovered to be harboring an intracerebral plasma cell granuloma. The literature on plasma cell granuloma of the central nervous system is discussed with emphasis on the clinical, radiological, and pathological features of these lesions.

Key words  Plasma cell granulomas • Brain • Childhood • Inflammatory pseudotumor • Meninges

Introduction

Plasma cell granulomas (inflammatory pseudotumors) were first recognized in the lung by Bahadori and Liebow in 1973 [4]. Since then they have been identified in virtually every organ. In the brain they present as a meningeal-based mass, with symptoms and signs referable to their location. In the neuropathological literature they had been diagnosed as “inflammatory meningiomas” prior to the general acceptance of plasma cell granuloma as a histopathological entity. To date, 23 cases of intracranial masses and 2 cases of masses involving the spinal cord have been reported [2, 6–9, 13–19, 22, 24, 30, 31, 35, 37, 40, 41, 43].

We present the case of a 5-year-old girl with a plasma cell granuloma (PCG) of the cerebral convexity, apparently not attached to the dura, and review cases of PCGs involving the central nervous system to examine their general features and associated systemic findings.

Case report

The patient is a 5-year-old black girl who was in her normal state of health until February 1991, when she was noticed to have subtle personality and behavioral changes. In March of 1991 she had a seizure at home, described by her mother as “jerking her head to the left and becoming unresponsive.” The girl lost bowel control and remained unresponsive for 5 min. This was followed by a 1-h period of confusion, headache, and neck pain. She was brought to the emergency room by her parents.

Her examination was normal with no focal neurological findings. Visual field testing was normal. Laboratory examinations showed a slight increase in white blood count with a left shift (14.5x10⁹/79% polymorphonuclear neutrophils, 3% band neutrophils, 17% lymphocytes, 1% monocytes), but were otherwise normal. The remainder of the laboratory work, including liver function tests, additional chemistry tests, urinalysis, and culture, showed no abnormalities. Computed tomography (CT) demonstrated an area of decreased attenuation and surrounding edema in the right parietal occipital area. The lesion enhanced with contrast medium, and measured 2x2 cm (Fig. 1). Magnetic resonance imaging (MRI) showed an intra-axial mass with homogenous dense enhancement. She was placed on a regime of Decadron (dexamethasone).
The patient underwent a right parietal craniotomy for resection of tumor. The bone overlying the lesion appeared normal. A firm nodular mass was found just beneath normal appearing gray within the white matter. A glial plane was dissected from the mass. A gross total resection was achieved after biopsies were sent for intraoperative frozen sections. Serum immunoglobulin drawn on postoperative day 6 were IgG 650 mg/dl (normal range 550–1450), IgA 57 mg/dl (28–140), and IgM 322 mg/dl (40–220). The patient was discharged on a tapering dosage of Decadron and Dilantin (phenytoin). Six months after discharge, laboratory studies revealed normalization of the white blood count. Serum immunoelectrophoresis has not been repeated. The patient reported her last seizure 3 months after the operation. She is a good student but continues to have behavioral problems in school and has occasional headaches that are relieved by rest. A CT scan performed 2 years postoperatively showed no evidence of residual tumor. The patient is now lost to follow-up.

The tumor specimen submitted measured 1.5x1.5x1.0 cm. Light microscopy of the frozen section stained with hematoxylin and eosin revealed an abundant lymphoplasmacytic infiltrate. Fixed tissue revealed a well-circumscribed, unencapsulated mass with abundant myxoid stroma. Two cell types predominated: evenly spaced spindle- to stellate-shaped cells with oval to angular nuclei and long tapering processes, and plasma cells in various stages of differentiation, with some perivascular accumulation (Fig. 2). Occasional germinal centers were identified. Extracellular eosinophilic granules were seen throughout the specimen; rare intact eosinophils were also present.

Immunohistochemical staining revealed the lymphocyte population to be composed of both T (UCHL-1 +) and B cells (L26 +). The B cells were polyclonal (IgG, IgM, IgA, IgE, k, l). Staining for infective organisms (Warthin-Starry, Brown & Brenn, acid-fast bacilli, Dieterle, Gomori methenamine silver) failed to reveal the presence of such. Immunohistochemical staining of the spindle-shaped cells showed positive reaction with antibodies to vimentin, desmin, and muscle specific actin. Staining with epithelial membrane antigen was nonreactive.

Ultrastructural studies of the spindle cells revealed fine cytoplasmic filaments, dense bodies, and pinocytotic vesicles. Desmosomes and interdigitating junctions were not found. The combination of immunohistochemical and ultrastructural features of the spindle cells allowed us to conclude that the cells were myofibroblasts. Ultrastructural analysis of the eosinophils revealed lipid droplets and granule core lucencies, suggesting degranulated or hypodense eosinophils.

Discussion

In the original article by Bahadori and Liebow [4] it was suggested that PCGs are the most common isolated primary tumor-like lesion of the lung in children less than 16 years of age. The lung is the most frequent site of presentation, with the central nervous system (CNS) being the second most common, followed by the liver. Other organs with infrequent involvement include the skin, adrenal gland, thyroid gland, orbit, breast, skeletal muscle (psoas), upper respiratory tract, tonsil and gingiva, middle ear and mastoid, heart, spleen, lymph nodes, genitourinary tract, esophagus, stomach, small intestine and appendix [4, 5,