Clivus plasmacytoma

http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4520983/

The mean age at symptom onset among the reported cases of clival plasmacytoma is 57 years with a female preponderance (female-to-male ratio: 2:1). The most common presenting symptoms are headache, visual deficits due to cranial nerve palsies, and endocrinologic abnormalities [1) 2)].

On MRI, intracranial plasmacytomas enhance with gadolinium contrast and demonstrate an iso- to hyperintense signal on T1-weighted images and an iso- to hypointense signal on T2-weighted images.1 On CT scan, the lesions are osseo-destructive and avidly enhance with intravenous contrast. Solitary clival lesions are rare. The most frequent tumor at this site is chordoma (40% of all cases) [3)].

The radiologic differential should also include sarcoma, lymphoma, carcinoma, pituitary neuroendocrine tumor, meningioma, and metastatic disease [4)].

On microscopic examination, plasmacytomas consist of a proliferation of sheets of plasma cells with a few intervening cells of other types. Cytologic atypia is variable. The tumors may be composed predominantly of small mature plasma cells with small round nuclei with peripherally clumped (“clock-face”) chromatin and inconspicuous nucleoli, or of larger cells with plasmablastic morphology, with large atypical nuclei with vesicular chromatin and prominent central nucleoli. Cytoplasm is typically moderately abundant and eccentrically oriented and may contain inclusions of condensed or crystallized immunoglobulin. A pale paranuclear Golgi region may be recognizable. Binucleated and multinucleated cells may be seen. Intranuclear protrusions of cytoplasm containing immunoglobulin called Dutcher bodies may occasionally be present [5)].

Immunohistochemical staining shows expression of CD138 and CD38.5 6 Approximately 70% of the neoplastic plasma cells coexpress CD56 as well, a surface antigen that is not usually present on nonneoplastic plasma cells. A hallmark of plasmacytomas is the demonstration of immunoglobulin light-chain restriction, indicating that the proliferation is clonal [6)]. [7) 8)].

In addition to histologic evidence of a monoclonal plasma cell infiltrate, diagnosis of an isolated plasmacytoma requires demonstration of a lack of systemic involvement.10 This includes absent or low serum/urinary levels of monoclonal immunoglobulin, a normal skeletal survey, and a normal bone marrow biopsy and aspirate.2 3 5 7 10 Some authors also recommend a PET scan to rule out other sites of disease. Given the high rate of progression to multiple myeloma, patients with intramedullary plasmacytoma require careful follow-up.10 In the case of plasmacytoma of the skull base, conversion has been reported as late as 1 year after diagnosis [9)].

The recommended treatment for solitary clival plasmacytoma is radiation therapy given at a dose of 40 to 50 Gy over a 4-week period .1 2 6

Surgery serves primarily a diagnostic role because the benefit of extensive resection has not been determined. Surgical debulking is indicated when there is neurologic dysfunction secondary to the mass effect of the tumor [10)].

Five-year disease-free and overall survival rates are 50% and 74%, respectively. Five-year local control following radiation therapy is relatively good at 86%; treatment failure is more often due to systemic relapse [11)].

If multiple myeloma is diagnosed, chemotherapy or stem cell transplantation may be appropriate [12)].

Case series

2016

Of 47 patients with plasmacytoma of the skull base, the tumor originated from the clivus and sphenoclival region in 28 patients (59.5%), the nasopharynx in 10 patients (21.2%), the petrous apex in 5 patients (10.6%), and the orbital roof in 4 patients (8.5%). The chief complaints at presentation included recurrent epistaxis and cranial nerve palsy, according to the site of tumor. Twenty-two patients (46.8%) had surgical treatment; 25 (53.2%) received radiation therapy. Adjuvant therapy was administered in 11 cases (50%) with concurrent multiple myeloma. The 2-year and 5-year overall survival rates were 78% and 59%, respectively. Clear margin resection was achieved in a similar proportion of patients who underwent endoscopic surgery and open surgery ($p = 0.83$). A multivariate analysis of outcome showed a similar survival rate of patients treated surgically or with radiotherapy [13]

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