Extramedullary plasmacytoma

Extramedullary plasmacytoma (EMP) is an uncommon disorder characterized by the development of abnormal plasma cell tumors outside the bone marrow. These tumors are typically observed in various locations, including the upper respiratory tract, gastrointestinal tract, and other soft tissues. Among the less explored manifestations of EMP is intracranial EMP, which remains poorly understood due to the limited literature available on the subject. The objective was to comprehend the population characteristics, localization, type, treatment, and outcomes of intracranial EMP. A systematic literature review for EMPs followed the Preferred Reporting Items for Systematic Reviews and Meta-Analysis guidelines. The strategy “extramedullary plasmacytoma AND multiple myeloma” was used for the search. The search terms were queried using PubMed, Embase, Scopus, Cochrane, and Web of Science databases. We included only those studies that presented clinical studies with patients diagnosed with intracranial plasmacytomas. In this study, a total of 84 patients from 25 studies were analyzed. The average age of diagnosis was 57.25 years, with a slightly higher proportion of females (57%) compared to male patients (43%). The most common locations of intracranial plasmacytomas were the clivus (29.7%), frontal lobe (18.9%), parietal lobe (8.1%), occipital lobe (6.7%), temporal lobe (6.7%), and sphenoid (4%). Chordoma and meningioma were the most common differential diagnoses encountered during clinical investigations. Treatment modalities included radiotherapy (RT), chemotherapy (QT), surgical resection (SR), and conservative approaches. The most frequent treatment combinations were SR + RT (19%) and RT only (17.8%). Mortality was reported in 48% of the cases, with complete resolution observed in 10 cases and partial resolution in 3 cases. The average follow-up duration was 37.5 months. The clivus is the most frequently reported site of extramedullary intracranial plasmacytoma (EMIP) occurrence, representing 29.7% of cases. Chordomas were commonly observed alongside EMIPs and emerged as the primary differential diagnosis. RT was the predominant treatment modality, with SR considered when feasible. RT alone demonstrated the highest effectiveness in managing EMIPs (30%), while QT as a sole intervention showed lower efficacy. However, a combination of dexamethasone, lenalidomide, and targeted RT displayed promising results, offering improved tumor response and increased safety [1)].

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Palavani LB, Bapat A, Batista S, Mendes JP, Oliveira LB, Bertani R. Extramedullary Intracranial Plasmacytomas: A Systematic Review of Literature. Asian J Neurosurg. 2024 Jun 6;19(2):137-144. doi: 10.1055/s-0044-1787535. PMID: 38974431; PMCID: PMC11226265.